

# A CONTRIBUTION TO THE STUDY OF MYOSITIS OSSIFICANS PROGRESSIVA

BY JULIUS ROSENSTIRN, M.D.

OF SAN FRANCISCO

SURGEON TO MT. ZION HOSPITAL

(From Mt. Zion Hospital, San Francisco, Calif.)

WITH the first case of myositis ossificans progressiva described in medical literature over two hundred and twenty years ago, and for sixty years the course and pathology of this strange and rare malady well studied, its character and etiology are still unsettled and the subject of vivid controversy.

The fact that another resemblant disease, the myositis ossificans circumscripta, produces identical pathological anatomical tissue changes in isolated muscles has not helped to clear up the mystery of its etiology and certain features in the symptom picture. The myositis ossificans progressiva is characterized by such a sharply outlined group of symptoms that it is marked as distinctly etiologically separate from its near namesake and not sharing anything with it in common save a relationship in local products of the pathological process.

These characteristic features are:

1. The ossification of muscles without any apparent cause, traumatic or otherwise.
2. The manifestation of the disease as a congenital one or appearing early in life.
3. The progressive course of the malady unaffected by therapeutical efforts, embracing in some of the most advanced cases nearly the entire voluntary muscular system.
4. The association with symptoms of defective anatomical and physiological formation, mostly of fingers and toes but not infrequently also of stature, habitus and sex differentiation.

These are the earmarks of typical cases of myositis ossificans progressiva, and, strange to say, there are very few exceptions, if any, of genuine cases of this malady which do not show them.

It is true that since Helferich first drew attention to the presence of microdactylia in patients suffering from progressive ossifying myositis, quite a number of histories have been published that do not mention this symptom in cases undoubtedly belonging to this malady, but I have ascertained through personal correspondence with the authors of a number of such publications that the reason for this has been a lack of attention in examining for this symptom, mostly due to an oversight in the outdoor department and also in a few hospital cases.

There are, however, a few cases where the authors have especially mentioned the absence of microdactylia, or similar anatomical defects, and these

are the observations which deserve special attention. Under the heading "remarks" of the collected case histories, the reader will remember this difference between *microdactylia not mentioned* and *microdactylia noted as absent*. The later definite statement should, in future, accompany all histories of such cases verified by the X-ray pictures of the hands and feet.

I have taken the trouble in compiling in chronological sequence all the accessible cases of myositis ossificans progressiva. It seemed to me to be an effort well worth while to assemble in brief all the known data of this rare and mysterious disease, as my studies of its literature disclosed many errors and omissions in the two or three fractional prior attempts.

The reason for undertaking this work was my own observation of a most interesting typical case, presenting among its symptoms the unique feature of several isolated and independent foci of ossification in the skin, a tissue hitherto believed to be exempt from this pathological change. It afforded unprecedented opportunity to observe and study the entire course of ossification from its first beginning to the completed bone-formation, a study which led me to advocate a change of view as to the primary factor of ossification in this disease.

In the arrangement of this paper I shall first give abstracts of all cases of myositis ossificans progressiva published to date, then present my own case and finally discuss the views of the pathology and etiology in the light of knowledge obtained by the study of the existing literature and of the findings of my patient.

ABSTRACTS OF CASES RECORDED IN LITERATURE

CASE I.—Reported by Guy Patin in 1692. Female. Subsequent course of disease unknown. Writes to A. F. (?) referring to a previous letter sent him, wherein he also mentioned the case of a woman who finally became as hard as wood all over.

CASE II.—Reported by John Freke, in 1743. Male, aged 14 years. First symptoms appeared at 11 years of age—swellings on back, muscles and vertebræ of back. Began with swellings three years ago, then growths spread over entire back from cervical vertebræ to os sacrum, and arising also from every rib, forming coral-like ramifications. Third year, from nape of neck to os sacrum and the lateral muscles of back; ossifications like coral branches, especially on back. No congenital abnormalities mentioned.

CASE III.—Reported by Rev. Dean Copping and Robert, R. R., Lord Bishop of Corke, and Charles Smith, in 1744. Skeleton of man, aged 60 years. First symptoms appeared at (?) 18 years of age. Subsequent course of disease unknown. Entire skeleton one mass. Could not open jaws, fed through gap in teeth. Vertebræ consolidated by lateral outgrowths with scapula. One bone, very thin, about four inches long, found in fleshy part of arm, *quite distinct and disengaged from any other bone in his body*. Ridges and reefs of bone through fleshy parts of both thighs like shoots of coral, eight to nine inches long. From back of calcaneum grow spurs of bone. No congenital abnormalities mentioned.

CASE IV.—Reported by William Henry, in 1759. Male, aged 19 years. First symptoms appeared at 17 years of age—right arm, swellings and pains in wrist. Both arms, ankles, and legs up to knees attacked. Both arms from elbow to wrists just like a solid mass of bone; later pain, swellings and ossifications attacked ankles, creeping up fleshy part of calves to knees. Was treated with mercurials to salivation. (Observation covering two years.)

## MYOSITIS OSSIFICANS PROGRESSIVA

CASE V.—Reported by Abernethy, in 1830. Male, aged 14 years. First symptoms and age at their appearance unknown. Various bony masses had formed at different times and disappeared again. Back greatly deformed by irregular hillocks of earthy matter over *processi spinosi*. Head immovably fixed backward and to one side. Exostosis on both *ossa brachii*, and tendinous margins of *axillæ* ossified so that arms were pinioned closely to his sides. Exostosis on pelvis between *os sacrum* and *coccyx*. No congenital abnormalities mentioned. Abernethy found urine contained less lime than normal. Two years later he gave patient phosphoric acid, 4.0 per diem in divided doses, with the result that normal quantities of phosphates of lime were excreted in the urine. When phosph. acid was discontinued deficiency of lime occurred again.

CASE VI.—Reported by David L. Rogers, in 1833. Male, aged 13 years. First symptoms appeared when at the age of 12 years and 6 months—stiffness in arms and neck. Very rapid course, attacking muscles of back and chest. Superior portion of *pectoralis major* and *sternocleido* from sternum to middle portion ossified. A number of osseous swellings in back. Scapula fixed to ribs, studded with bony excrescences; all muscles connected with scapula—viz., *trapezius*, *rhomboideus*, *subscapularis*—more or less ossified; *latissimus* and *longissimus dorsi* formed large bony plates from their origin to angle of scapula. No congenital abnormalities mentioned.

CASE VII.—Reported by Testelin, and Charles Dambesi, in 1839. Male, aged 26 years; mother suffered from rheumatism; patient fell on right thigh. First symptoms appeared at 18 years of age—pains and limping after trauma. Right thigh attacked. Five years later pains in all extremities, movements began to be difficult, particularly scapulo-humeral articulation. Continued to move around seven years longer till he was unable to move either jaws, arms or legs. Became completely unable to move limbs or jaws. Twenty-one years later died, 39 years old. Autopsy showed ossification of right temporal and portion of left pterygoid, left deltoid, both *pectorales major*, left minor right *coracobrachialis*. Both *biceps brachii*, left *triceps*, right *latissimus*, right *long dorsalis*, right *glutæus med.* and left *med. and minor*, and most of thigh muscles on both sides.

CASE VIII.—Reported by Caesar Hawkins, in 1844. Male, aged 22 years. First symptoms appeared at 22 years of age (June, 1843)—lumbar and dorsal regions attacked. Violently painful swellings in lumbar and dorsal regions, which disappeared, leaving in places some small bony masses. Disease later attacks neck and scapular regions. New attacks began in October, 1843, and persisted during the next six months of observation. (One year after symptoms of disease.) Lumbar *vertebræ* immovably bound together by osseous masses; unable to use shoulders freely, particularly on left side, owing to swelling under angle of scapula and ossification of tendon of *pectoralis major*. Small exostosis on rib on either side (which rib?). No congenital abnormalities mentioned.

CASE IX.—Reported by G. Wilkinson, in 1846. Female, aged 21 years. First symptoms appeared at 8 months of age—stiffness in arms and lumps on back. Could move about and work till 11 years old, then lost motion in elbow-joints; other ossifying processes progressed rapidly. Chest cavity much diminished by curvature of dorsal *vertebræ* and acute angularity of ribs. Numerous thin ossific plates in anterior and posterior chest muscles, as *pectorales*, first *dorsi* and *erectorides spinæ*, and one plate ten and a half inches long connected to crest of left ilium and short branches running from it into *rhomboidei* to base of left scapula into *infraspinatus*; similar band from right crest of ilium to various *vertebræ* up to transverse process of third dorsal. Other irregular plates of bone connected spinal processes of dorsal and lumbar *vertebræ* and ribs connecting with sacrum by short tendinous fibres. Numerous bands of osseous matter in depressions of lower jaw; portion of bone two and one-half inches long in left *biceps brachii*. Two irregular bony processes, one from lower end of left humerus three inches downward into pronator *teres* and flexor muscles of

## JULIUS ROSENSTIRN

forearm; other from outer edge posteriorly extending into triceps and extensor carpi rad. longior et brevior. No congenital abnormalities mentioned.

CASE X.—Reported by King Kelburne, in 1854. Female. First symptoms appeared at 3 weeks of age—defective power of suckling and swallowing. Nourishment had to be brought into posterior part of mouth with spoon. Author discovered tumor on right side of tongue, size of large bean, judged to be of vascular nature, and therefore injected a few drops of perchloride of iron. In spite of a supposed improvement following this measure the child did not mend and died about five months later from inanition. Subsequent course of disease unknown. The post-mortem showed ankylosis (bony) between inferior maxilla and temporal bones, musc. masseter replaced by a large osseous plate descending from the inferior margin of the zygomatic process and malar bone. The osseous deposit extended as far forward as the anterior process of the malar bone, incorporated itself in its downward passage with the tuberosity of the superior maxilla, and finally amalgamated with the angle base and ramus of the inferior maxilla. These changes were on both sides. Vascular tumor on tongue had nearly entirely disappeared, leaving a fibrinous remnant of about the size of a split pea. No account given of other parts.

CASE XI.—Reported by Jonathan Hutchinson, in 1860 and 1892. Male, aged 12 years. First symptoms congenital. Midwife noticed deformities; patient put in London Hospital on their account. Subsequent course of disease unknown. Left shoulder, wrist, and carpal joints ankylosed. Left humerus shorter than right and bony ridge running down outer side. Large plates of bony substance in fascia of forearm, palm and back of hand. On right side small areas of bony structure in fascia above and below elbow-joint. No congenital abnormalities mentioned. At the age of 37 years the ossifications have largely increased in size and extent, and over right scapula and in posterior fold of right axilla new bony plates have developed. Movement of thorax during respiration extremely limited.

CASE XII.—Reported by William Skinner, in 1861. Male, aged 13 years. Family history negative. First symptoms appeared at 7 years of age—arms, back and right shoulder attacked. Stiffenings in arms, swellings in nape of neck and behind right shoulder. Stiffness at first was so bad that arms could not be brought to mouth. This improved later so that he could with difficulty feed himself, but hard nodules formed on different parts of body, especially breast and vertebral column. If patient receives blow or knock a hard swelling follows, preceded by pain and slight fever. Patient somewhat bent forward; slight motion in left shoulder, none in right. Arms cannot be extended and lie half bent across abdomen. Spine and scapula immovable. Small bony protuberance on ribs. Right pectorales changed into bony mass—continued into biceps until insertion at radius. Hard nodules at various places. No congenital abnormalities mentioned.

CASE XIII.—Reported by Ivan Minklewitsch, in 1864. Female, aged 15 years. Family history negative. First symptoms appeared at 5 years of age—swelling and stiffness in lower part of neck. A few years later osseous tumors in back and upper extremities began to develop, then attacked lower extremities. Three years ago pain and swelling in right knee disappeared and a week later reappeared. Exostoses in left temporal region at insert of temporal muscle; similar ones over left temporal and over acromion. Muscles forming posterior wall of right axilla ossified. Pectorales show same change, also small rough exostoses over cond. int. humeri. Left arm a similar ossification of axillary muscles. Right lower extremities: over articular tibia tarsalis stalactite-formed exostosis, similar one over fibula; another below lower margin of patella; in soleus muscle osseous growth size of hen's egg; in fossa poplitea narrow one extending upward to thigh and in stalactite formation reaching nearly plica glutealis and inguinalis. Left lower extremities: similar growth below patella as on right side, extending upward and inward to fossa poplitea and ending at upper end of middle third of thigh; sharp-pointed exostoses on outer side of astragalus and

## MYOSITIS OSSIFICANS PROGRESSIVA

outer malleolus. No congenital abnormalities mentioned. Report of autopsy, in 1871: New ossifications had formed at the right upper extremity on the upper posterior third of humerus and over inner part of that bone near elbow. The exostoses over cervical vertebrae and over second to fourth ribs were considerably enlarged.

CASE XIV.—Reported by Johanne Zollinger, in 1867, and Theodor Billroth, in 1869. Male, aged 24 years. Believed to have been caused by fall (?). First symptoms appeared at 3 years of age—arms first showed stiffness in movements. Movements of arms grew more and more difficult, and after a few years present state developed. Bent forward in lumbar position of vertebral column. Head bent forward; mouth can only be slightly opened for introduction of little finger. Both sternocleidii muscles changed to rigid cords. Arms in slight abduction and flexion in elbow-joints; tendons of pectorales and deltoids hard and very tense. Back shows many protuberances of hard osseous consistency, situated mostly around the lower part of lumbar region; they branch off in many different directions. Especially large is a ridge going on either side into glutæus maximus; others in regions of sacrospinalis, irregular and asymmetrical. Both latiss. dorsi nearly entirely converted into bone. First interphalangeal joint of left middle finger thickened by exostoses, and small roundish exostosis on left index finger. Small exostosis at inner side of left tibia and on outer side of r. cond. ext. femur. No congenital abnormalities mentioned.

CASE XV.—Reported by Muenchmeyer, in 1868, and A. Jurasz, in 1873. Female, aged 22 years. Family history negative. Fall when 4 or 5 years of age. First symptoms appeared between 4 and 5 years of age—neck and right arm; nodules on nape of neck. Development of right-sided scoliosis; stiffness of neck and right arm. After first nodules on nape of neck had disappeared others came in different parts of body. Brain fever at 12 years of age. In May, 1864, had scarlatina. Patient does not recollect ever having been able to turn her head freely or move arms away from her sides. Asserts never to have been able to dress herself or to do up her hair. Cervical column and head completely immovable; position of head in right-sided caput obstipum. Dorsal spine right-sided scoliotic and immovable; both scapulae firmly fixed. Left arm nearly completely immovable in elbow-joint; a nodulous osseous mass between cond. ext. hum. and olecr.; left biceps brachii one solid mass and long bony stalactite extending from middle toward axilla. Hard ossified parts also in deltoid. Both latiss. dorsi and trapezii, teres major and minor are changed to osseous masses. Also the enormously thickened lig. nuchæ. No congenital abnormalities mentioned. Latest observation in 1869; all changes have remained, and in addition the whole back one solid mass, protuberances, nodules and ridges, with some softer parts intermixed. A six-centimeter broad osseous mass leading from vertebral column to scapula fixing the latter completely. On the supraclavicular fossa everything except the omohyoideus seems to be ossified. In right leg whole gluteal mass ossified; from thigh down to knee several broad bony masses. During time of observation various places all over the body showed swellings, which disappeared again. Immovability of jaws increased so that patient could take only liquid food. In 1869 disease got worse and continued from bad to worse. In 1872 m. gracilis involved. In November mental trouble began. February 26, 1873, she fell and broke her right arm. Plaster-of-Paris dressing, cured in two weeks. No excessive callus. Died in thirty-second year of her life. Mays' autopsy report in 1874.\* The autopsy report by Mays is too lengthy to be given here. It adds some exostoses to the ossifications found during lifetime, but shows the cerv. muscles not to have been ossified, only hard and infiltrated with a connective tissue hyperplasia. No examination of fingers or toes. The spinal cord and periph. nerves were examined by Doctor Schultze† and found normal.

\* Virchow's Arch. f. patholog. Anat. und Phys., bd. 74, p. 145.

† Erb and Schultze; ein Fall v. progress. Muskelatrophie, etc. Arch. f. Psychiatric und Nervkrkhtn, 1879, bd. 60, h. 2, pp. 385-6.

## JULIUS ROSENSTIRN

CASE XVI.—Reported by Breschet, in 1869. Skeleton. Subsequent course of disease unknown. In addition to real exostoses, also multiple and symmetrical ossifications of muscular attachments. Skeleton in Musée Dupuytren (Paris).

CASE XVII.—Reported by William Byers, in 1870. Male, aged 17 years. First symptoms appeared at 8 months of age—small tumors size of small marbles appearing in different parts of body. Various parts of body attacked. Stiffness of joints, hips and shoulder-joints. The first nodules disappeared, but after twelve to eighteen months stiffness of joints set in again. Gradual ankylosis followed, and at the age of 10 years left hip and shoulder joints were completely ankylosed. Later the muscular system began to show ossification. Chest became as though enclosed in a complete sheet of bone, leaving no trace of outline of ribs. Head was immovably fixed by ossification of both sternocleidides. Up to his seventeenth year muscles of mastication remained unaffected. Could at first move slowly and cautiously over smooth surface; with ankylosis progressing this became impossible. No congenital abnormalities mentioned. Remained small; sexual infantilism. Died at the age of 21 years; muscular system almost completely ossified.

CASE XVIII.—Reported by Florschuetz, in 1873, and Gerber, in 1875. Male, aged 12 years. Family history negative. First symptoms appeared at 5 years of age—hard inflamed nodules in right latiss. dorsi below right scapula. Latiss. of left side, upper extremities and back attacked later. After having attacked shoulders and upper extremities it progressed to muscles of back, trunk, neck and mastication (tempor. and masseter) and upper parts of lower extremities. Frequent nocturnal epileptic attacks. Absolute stiffness of large muscular groups. The greater part of muscles of neck, all muscles of mastication, causing complete lockjaw (nutrition through gap in teeth), muscles of back, breast and arms (arms are immovably fixed to sides of body, only left forearm functions slightly), and finally those of left thigh and hip and part of right thigh are ossified. Many muscles which are not ossified are atrophic, evidently in transitory stage, previous to ossification. No congenital abnormalities mentioned. Penis normal in size relative to age. Scrotum atrophied and testes quite undeveloped. Autopsy report, Mays,\* 1874. Very extensive autopsy in which several exostoses were found and most of the ossification diagnosed during life confirmed except in the cerv. muscles, especially in sternocleid mast. and the delt. and adjoining extensor musc. of the arm, which only showed a firm infiltration, together with development of the connect. tissue hyperplasia. Fingers and toes not examined. Spinal cord and periph. nerves found normal by Schultze.†

CASE XIX.—Reported by Edward F. Hamilton, in 1874. Female, aged 30 years (?). No clinical history obtainable. Subsequent course of disease unknown. Body brought to Medical School of St. Stephen's Hospital. Body inclined to left side; head bent forward, arms closely applied to sides; forearms flexed and pronated. No motion of shoulder-joints nor of scapula upon trunk. Left side of thorax contracted; right ilium nearly in contact with ribs. Lower limbs flexed and adducted, right leg and foot much inverted; ankle joints stiff. Skeleton showed: Along anterior border of left masseter a ridge of bone preventing any movement of jaw. Head immovably joined to upper cervical vertebræ. Spinal column forms one solid mass; cervical, dorsal and lumbar vertebræ firmly joined by copious bone deposits; left shoulder-joint perfectly stiff and large plate of bone fixed to humerus near insert. of pector. maj. reached up to coracoid proc., then down and inward till identified with costal cartilage. Connecting the shaft of right femur and trochanter immense osseous growths resembling stalactites had formed, branched into the fibres of extensor muscles. Similar growths were upon dorsum of ilium. Osseous deposits at patellas, insert. of quadriceps, extensor, tendon and many ligaments of soles of feet had ossified. Between muscles of back from occipital bone to inferior angles of scapula and down to crest of

\* Op. cit.

† Op. cit.

## MYOSITIS OSSIFICANS PROGRESSIVA

ilium. Consolidating hip-joints were enormous plates of ossific matter; in front of left femur osseous deposits bearing resemblance to a second femur. This deposit had been formed in cellular tissue between rectus femoris and cruraeus.

CASE XX.—Reported by Bennett, in 1874. Female, aged 11 years. Family history negative. First symptoms appeared in infancy—swellings over shoulder and back of neck. Irregular attacks over various parts of body. On right side from protuberantia occipit. to supraspinous fossa a bony growth is stretched, apparently situated in trapezius, irregular masses of bone are deposited in location of rhomboid muscles, and bony processes branch upward and inward from latiss. dorsi, portions covering scapular angles, other branches going down from those to crest of os ilium. Across middle and upper parts of loin transverse osseous bars have developed following the course and fibres of the muscle. A spur has commenced to grow from either ulna near attachment of pronator quadratus. No congenital abnormalities mentioned. Cast of girl demonstrated.

CASE XXI.—Reported by Dittmeyer and Gerber, in 1875. Female, aged 8 years. Family history negative. First symptoms appeared at 6 years of age—complained of pains in right side of nape of neck and shoulder. Stiffness of neck and right shoulder; some difficulty in breathing. Right shoulder grew immovable, breathing became more difficult. The left shoulder stood higher, head had been drawn toward left side. Right trapezius muscle stiff and hard, so that the head could not be moved to either side. The supraspinati and teres minor grew as hard as a board and arms were tightly drawn to sides of thorax. In serratus post. super. one could feel the daily spread of the ossifications along the separate muscles. All other muscles were free. No congenital abnormalities mentioned. General health, with exception of the difficult respiration, was good; all organs functioned well. Autopsy report, Mays.

CASE XXII.—Reported by V. P. Gibney, in 1875. Female, aged 10 years. Family history negative. Attack of diphtheria (?). First symptoms appeared at 10 years of age—had been perfectly well up to attack of diphtheria. Subsequent course of disease unknown. Muscles involved were latiss. dorsi, scaleni and erector spinæ. Right arm was held down by tendon of latiss. dorsi. Also lateral curvature. No congenital abnormalities mentioned. No mention of microdactylia. At suggestion of a colleague acid. lact. was given, with no amelioration of symptoms; but there was no further progress during the next year and a half of observation. Seen again in 1893. Had been working in a millinery shop since 1884. No change of status. (Doubtful case.)

CASE XXIII.—Reported by Huth, in 1876. Male, aged 4 years. Family history negative. First symptoms appeared at about 2 years of age—hard spots under chin. Gradual extensions toward angles of lower jaw; then chest and other parts of body. Hard swelling size of egg developed over middle of sternum at about 2 years of age. It disappeared again. On shoulder and both upper arms swelling and hardness developed. In the spring of 1874 a swelling appeared on forehead, extending over eyelids, eyes being entirely closed. This disappeared again. Upper arms lying closely to trunk and fixed; lower arms movable. Head, neck and thorax muscles hard and stiff like armor. Both glutæi, especially of left thigh and leg, ossified. In abdominal muscles a cord thick as finger running from angle of ribs to os pubis. No congenital abnormalities recorded.

CASE XXIV.—Reported by Nicoladoni, in 1878. Female, aged 7 years. First symptoms appeared the first year of life—stiffness in muscles of neck. Later stiffness in muscles of back. At present two hard ridges corresponding to sacrolumbales. The scapular muscles are ossified so that the shoulder-blades are immovably fixed to thorax. A# upper part of biceps brachii are small bony plates in various parts of the muscle, its tendons changed to hard fibrous cords. The same changes present in sternocleidomastoidei. The contracted right knee-joint can only

## JULIUS ROSENSTIRN

be extended to 120°. Semitend. and membranos. changed to two hard lumps. Pectorales also ossified and the axillary fossæ are bounded in front and posteriorly by rigid walls. The muscles of the lower jaw are also involved. No congenital abnormalities recorded.

CASE XXV.—Reported by H. Helferich, in 1879 and 1883; Carl Mannz, in 1893, and Arthur Manneberg, in 1896. Male, aged 16 years. Family history neg. First symptoms appeared at 6 years of age—numerous hard nodules appeared upon head and gradually disappeared again. Later similar painful nodules appeared on back; arms and back became stiff. Between the twelfth and thirteenth years slow development of closure of jaws; first right molar had to be extracted to allow feeding. When 15 years old could still push out tongue between teeth. Jaws now firmly closed. Arms gradually became stiff and nodules developed all over them. Axillary fold hard; masseteric region hard ridges from the process. zygomatic to lower jaws. Jaws locked. Neck in region of hyoid bone hard, but bone and larynx movable. Thorax, pector. maj. shows ossified swellings. Back, swellings of osseous hardness spreading like antlers from pelvis upward. Right hip immovable; left hip normal; left gluteal region, flat resistant bone-hard mass; abscess developed in right inguinal region. First case where attention was drawn to microdactylia of thumbs and big toes in this disease, although anatomical defect was erroneously thought to be defect of an entire phalanx instead of smallness of metacarpal or metatarsal bone. In later observation (1887) it is reported that case had remained fairly stationary since first observation. In latest observation (1896) by Manneberg, quite an additional progress is noted, comprising old and new muscular groups.

CASE XXVI.—Reported by Partsch, in 1882. Male, aged 17½ years. Family history neg. First symptoms appeared at 10 years of age—swelling after fall on right shoulder; disappeared again, but hardness remained in right side of neck. Developed stiffness of muscles, which affected right shoulder and upper arm; later left arm; after fall on and wounding forehead a swelling of os frontale followed. Two years ago muscles of left side of neck hardened and drew head over to left side. About same time ossification of masticating muscles set in so that mouth can only be opened 1 cm. Head bent to left and fixed forward; both arms flexed in elbow-joints, right cannot be approached to body, left only with great exertion. Both shoulders fixed. Cervical column rigid. Sternocleido fibrous, but no bony enclosures. Scaleni show osseous changes. Right scapula carries a small roundish osseous tumor 4 cm. long, 2 cm. broad; lower border of left latiss. dorsi entirely osseous. Left brach. ant. contains osseous growth. Right latiss. dorsi shows osseous growths extending to upper arm and triceps. In back: two osseous plates (11 cm. long), one in each fascia lumbodorsalis. Exostoses 1½ cm. long near right spin. ant. os. il. Broad bony plate in right tens. fas. lat. and narrower one in right sartorius. Similar one 3 cm. broad and 13 cm. long in left sartorius. Microdactylia of thumbs and big toes.

CASE XXVII.—Reported by Herm. Kuemmel, in 1883. Male, aged 13 years. Family history neg. First symptoms appeared fourteen days after birth (congenital)—foster-mother then observed striking deformity of spine and back and restricted mobility of both arms, which could only be slightly abducted from thorax. In second year various fluctuating swellings were observed upon back. They shrank and hardened into exostoses. Except some hardening of more swellings no particular change occurred until twelfth year, when gradual contraction of left knee-joint began. Head inclined forward and slightly to right side; movements very restricted. Small exostosis on occiput. Arms hardly movable from sides of thorax; upper part of body bent forward and curved. Floor of oral cavity forms callous mass, including os hyoid, and thyroid cartilage. Both sternocleidi and sterno-hyoid fibrous. Entire cervical spine bent forward and forms one continuous solid mass. Ligam. nuchæ much thickened. Costal cartilage forms pectus carinat. In both



## MYOSITIS OSSIFICANS PROGRESSIVA

pectorales major, bony bars extending in right to coracobrachialis. Both axillary posterior walls contain bony bars. In right rect. abdomin. tough fibrous and thin bony band. Both psoas tough callous masses. Dorsal scoliosis to right, lumbar to left. From seventh dorsal vertebra downward processi spinos. form one solid osseous mass. Both large trochanters broadened and prominent. Muscles of left thigh posteriorly changed into tough callous mass, with freely movable bony protuberance. Left leg flexed, angle of 45 degrees. Both thumbs nearly (?) normal; show bony ankylosis in interphalangeal joints. Microdactylia of both big toes ascribed to absence of first phalanx. (Real cause is probably smallness of metatarsal bone.)

CASE XXVIII.—Reported by Uhde and Pinter, in 1883. Male, aged 16 years. Family history neg. First symptoms appeared first year of life—swellings on head, which disappeared after some time. Patient broke arm in 1876. Fracture healed perfectly and shows *hardly any trace* of callus. Head and shoulder joints bent forward; arms ankylotic in shoulder joints; movements in elbow joints limited only. Ankylosis of interphalangeal joints in both thumbs. Microdactylia of both great toes (with same probable error as above).

CASE XXIX.—Reported by Krause and Pinter, in 1883. Female, aged 11 years. Family history neg. First symptoms appeared soon after birth—swellings in several places on scalp, which disappeared again after several months. At age of 4 years swelling in some of the deeper muscles of that region. It spread rapidly, then decreased, but left board-like hardness, fixing scapula firmly to thorax. Short time after same process occurred on right side. Disease progressed to left latiss. dorsi and biceps brachii, part of intercostals and serratus. Two years ago right mylohyoid, geniohyoid and part of biventer followed with swellings, which, disappearing, always left board-like hardness with spinous growths 1.38 cm. tall; circumference at mammilla 62 cm. Slight kyphosis, convexity at seventh dorsal. Left shoulder completely stiffened. Deltoid atrophic. Biceps ossified, fixing elbow in slight flexion. Broad band from spine to angle of scapula, two larger on right. Over scapula and thorax flat exostoses. Left latiss. dorsi totally ossified. Ribs below fourth rigid and unyielding. No mention of microdactylia.

CASE XXX.—Reported by Gyula Pinter, in 1884. Female, aged 20 years. Family history neg. First symptoms appeared at 4 years of age—hard nodules on back and disturbances of mobility of both shoulder joints. Patient does not remember ever having been able to lift arms above shoulders. At 6 years abduction of arms became so restricted that she could not write when seated at school desk. Other joints remained free until twelfth year. About the twelfth year, during formation and healing of suppurating swelling over right tibia, nearly all her joints lost greater part of mobility. In 1881 another suppurating swelling appeared on left leg, with a similar injurious effect on the various joints, rendering them still more rigid. When lying in bed rigidity of body like that in tetanus. Head; caput obstipum nearly immovable; chin held out and downward; ankylosis of lower jaw-bone; only lateral excursion of about 2 mm. in horizontal plane possible. Both temporales and masseters of bony hardness. Thorax absolutely immovable during respiration, owing to complete ossification of both pectorales major and minor. Axillary fossæ rigid walls; both cucularis ossified. A bony ridge 2 cm. broad, extending in middle line of back from prominentia occipit to seventh cerv. vert. Back and arms, two bony ones on either side of ninth and tenth dorsal vert. Left 10 cm. long, 2½ cm. wide, sends branches toward left scapular angle and down to crista ilei. Right, 6 cm. long and 2 cm. wide, sends similar branches anastomosing with other side. All firmly united with underlying parts of skeleton. Shoulder-blades immovably fixed to trunk. Stalactites of bone in ant. and post. muscles of both upper arms, and in left pronator teres and radial ext. Pelvis and legs, exostoses 3 cm. long on both crist. near ant. sup. spine. Hip joints immovable; muscles origins from both tuber. and ram. asc. ischii and trochant.

## JULIUS ROSENSTIRN

maj. all ossified. Bony ridges in lower halves of bic. fem. and semitend. and membr.; muscles of lower legs and feet free. Microdactylia of both big toes.

CASE XXXI.—Reported by O. Kohts, in 1884. Male, aged 23 years. Family history neg. First symptoms congenital. Could not move since his ninth year, having then made his first attempt to walk. Severe pains ensued after he first tried to walk, which made him take to his bed until his fourteenth year, when he entered the hospital. Patient 141 cm. tall. In bed could not change his position. Head and trunk slightly bent forward and to right. Lower jaws ankylosed. Masseter and pterygoidei apparently ossified. Exostosis 2 cm. long on inframaxillary bone. Ligam. nuchæ and muscles cucullar. and splen. partly ossified. No thoracic respir. movement. Both clavicles show large exostoses. Shoulder joints ankylosed. Both maj. and minor pectorales contain bony plates; entire lower margins ossified. Both arms firmly fixed to thorax. Both coracoid muscles and left brach. int. ossified. Right elbow-joint ankylotic. At fourth rib exostosis 2 cm. long. Left elbow limited mobility. Muscles coming from right condyl. int. humeri ossified in upper parts. Both hip-joints ankylotic. Deep glutæi and insert. of right quadrat. ossified. Left tens. fasciæ and tendon of right biceps femur and insertion of right gastrocnemius contain bony plates. Spinal column scoliotic; vertebr. column ankylotic; muscles along both sides atrophic and ossified. No mention of microdactylia.

CASE XXXII.—Reported by T. Sympson, in 1886, and Stonham, in 1892. Male, aged 6 years. Father rheumatic and presents same congenital deformity of toes as son. Fell on shoulder when 5 years old. First symptoms appeared at 5 years of age—painful swelling formed on right shoulder after fall one week earlier, then shrank gradually; three months later a similar swelling appeared on left scapula. Various swellings formed and disappeared again on back and chest. Jan. 16 (at demonstration) saddle of bone over loins, making stooping impossible. Margin of each latiss. dorsi, of each teres major and long head of r. triceps were occupied by a series of lumps apparently bony; similar lumps present a little below occiput in left trapezius, the middle of neck and on right side, and on supra-spinous fossæ. Six years later (1892) two bones size of filberts found on inner side of post. sup. spine of r. ilium. Patient fell Aug., 1887, and broke both bones of left forearm. Fracture united well. A firm hard swelling involving left vastus ext. and int., extending from just above patella upward for about four inches, consequent to a fall upon the knee. A similar swelling posteriorly in the popliteal space, biceps and semitendin. Motion of knee limited. A nodule size of filbert in middle of inner side of left thigh, situated in sartorius muscle. Feet show head of each metatarsal bone large and prominent; big toe on either side small and apparently consists of only one phalanx (?) and is directed toward outer border of foot. (No X-ray.)

CASE XXXIII.—Reported by Willett, in 1886, and Stonham, in 1892. Male, aged 4 years. Family history neg. First symptoms appeared at 6 months of age—swelling at superior angle of right scapula. When 1 year old swelling along vertebral border and inferior angle of right scapula. Six months later prominence noted in left erector spinæ and a little later a swelling near lower angle of left scapula. Right frontal eminence enlarged. Hard growth over left transverse processes of fourth and fifth cervical vertebræ. Right lateral dorsal curvature of spine, with slight compensat. curvature in lumbar region to left. Slight kyphosis of upper dorsal spine. Both scapulæ fixed. Hard growth along vertebral border of right scapula. Latiss. dorsi at scapular angle and in its tendinous portion ossified. A similar, less extensive hardness on left scapula and latiss. dorsi. A bony nodule on front angle of eighth rib, similar on left. Both erectores spinæ ossified. On right knee just above head of fibula hard mass 1 inch in diameter, with nodule size of pea. Inner tuberosity of right tibia slightly thickened. Both big toes hallux valgus position, displaced outward and under second toe. Shortened microdactylia, only ungual phalanx present

## MYOSITIS OSSIFICANS PROGRESSIVA

on each side (?). In Stonham's subsequent report six years later no correction of this statement of a very unusual condition is made.

CASE XXXIV.—Reported by Rickman J. Godlee, in 1886. Male, aged three years. Family history neg. First symptoms were noted when 1 year old, but may have been present long before, as nothing was learned of previous history from parents—bony plates in right latiss. dorsi and a nodule near right knee. Was not seen again till two years later, when present status was noted. In r. latiss. dorsi along outer border and post. fold of axilla, irregular bony mass; arm cannot be abducted more than sixty degrees, but can be completely adducted. Flexion of shoulders considerably limited; extension less; rotation inward more than outward. On left side post. a large elastic mass extending medially to nearly within the middle line and considerably below scapula, and upwards over shoulder to near clavicle. Scapula appears fixed in mass below angle. This mass had disappeared on March 23, 1886, but there is a hard mobile mass below left angle. Similar hard bodies, apparently bony, not movable, on free edge of left latiss. Seen again May 20, 1886; over the fourth rib, behind in post. axillary, ossifications. Over third rib in posterior axillary line, similar ossifications. Shoulder cannot be raised to right angle. Head drawn down to left shoulder. Some hard masses in muscles on outer side of neck (trapez. and right mast. excepted). Left arm cannot be raised higher than 45 degrees from trunk; scapula firm. In post. axillary fold hard nodulous mass. From angle of scapula a hard crest runs over to first lumbar vert. Back strongly curved; lower left ribs down on crest of ilium. Pollex valgus and microdactylia of thumbs. Hallux valgus and microdactylia of toes.

CASE XXXV.—Reported by R. v. Volkmann, in 1887. Male, age (?) Skeleton. Subsequent course of disease unknown. Various parts of his muscular system had been partially substituted by bony masses, which had grown into them. V. Volkmann had made attempts by removing some of them to restore a certain movability of ankylotic and immovable parts of the skeleton. The only published mention of this patient is made in a brief and vague reference in a discussion of Helferich's case. The extirpated bony masses showed at one end an epiphysal layer of hyaline cartilage.

CASE XXXVI.—Reported by Alfred Austin London, in 1887. Male, aged 43½ years. Family history neg. Accuses slight blows when 10 years old. First symptoms appeared at 10 years of age—joints of legs and shoulders stiffened. Gradually until his thirtieth year grew quite stiff and helpless. Jaws could only be opened very little. Over sacrum a bedsore formed from which pieces of bone were constantly exfoliating. Ossification of deltoid from acromial origin to insertion; also part of coracobrachialis. Scapulæ immovable on trunk, their inferior angles soldered by dorsal buttress of bone attached by seventh to ninth ribs. Latiss. dorsi ossified nearly entire lengths. Ossification of left brachialis anticus, and bony rims around both elbow-joints. Sacro-iliac ligament ossified. Both hip-joints ossified. Bone rods developed in lower part of glut. max. and extending from sacrum to femoral insertion. Huge bony projection on back of femur. Dorsal lumbar curve, with slight rotation to right. Very rigid, due to ossification of the capsular ligaments of the articular processes and many of the supraspinous ligaments. Died at age of 46. Microdactylia of both big toes. (No mention of thumbs.) General synostosis of the neural arches. All the ribs except the eleventh and twelfth are ankylosed to vertebral column. Bones have undergone eccentric atrophy and are consequently rather light.

CASE XXXVII (?).—Reported by E. Schwarz and Cl. Eichhorst, in 1888. Male, aged 40 years. Family history neg. First symptoms appeared at 39 years of age—a hard mass in posterior muscles of thigh. Subsequent course of disease unknown. Exostoses also on right humerus and on right femur in the region of the trochanter minor. Hyperostosis of right fibula. Suffers from tabes dorsalis. This case, as well as another from Eichhorst's clinic, both affected with spinal diseases. Do not belong to myositis ossificans progressiva. No microdactylia.

CASE XXXVIII.—Reported by Kronecker, in 1889. Male, age (?) First symptoms

## JULIUS ROSENSTIRN

appeared in (?) later years—first pains in nape of neck and in chest, two years previously. In November of the previous year the *scaleni* became affected. Since one year dyspnoea increasing; the *cucullares* are involved and the process is evidently progressing. Patient has pains in neck and one feels distinctly the increasing ossification. No *microdactylia*. (Doubtful, atypical case.)

CASE XXXIX.—Reported by J. v. Bokai, in 1899. Male, aged 5 years. Family history negative. Rickets. First symptoms appeared at  $4\frac{1}{2}$  years of age—nape of neck and back. Subsequent course of disease unknown. Muscles of nape of neck, back and *serrat. anticus maj., pectorales maj., cucullaris* and *latiss. dorsi* are affected. No congenital abnormalities recorded.

CASE XL.—Reported by Ivar Svensson (Sabbatsberg Hospital), in 1891. Male, aged 14 years. Family history neg. Fell when 4 years old (?). Age of first symptoms unknown—stiffness of body, arms and back. Stiffness has been gradually increasing. Stiffness of head and back and arms. Scoliosis on nape of neck; these are hardened and flattened and atrophied about the level of the *levator anguli scapulæ*, with fixed shoulder-blades; arms cannot be raised more than one-fifth of right angle from thorax. Otherwise arms can be lifted to horizontal plane, but not higher. Passive efforts to overstep this give crepitation and pain. In the height of axillary fossa a flat piece of bone of about 7 cm. length and 2 cm. width intimately connected with the tendons of *latiss. dorsi* and *teres major* and to the edge of the scapula. At the inner sides of both *scapulæ* parallel with spine one can palpate a round bony formation of 5 cm. length and hardly 1 cm. width. No mention of *microdactylia* in either fingers or toes.

CASE XLI.—Reported by C. Studsgaard, in 1891. Female, aged 4 years. Family history neg. Measles and whooping cough when 2 years of age. First symptoms appeared at 2 years of age—noticed swelling after measles and whooping cough. (Where?) First swelling (where?) disappeared again during use of cod-liver oil. Growth of bony consistency appeared after another year without any cause on the left side of the neck, rendering movements of the neck, especially downward, very difficult. Diffuse osseous swelling appeared on left lower jaw like convex continuation, cylindrical, size of finger, from base of that bone in region of premolar to *incisura semilunaris sterni*, and firmly attached to left horn of the *corpus ossi hyoidei* with one end, with the lower movably connected with sternum. Slight movements of maxilla are still possible, also small side motions of the head. The mass is situated anatomically as if it were as ossification of the *sternohyoid* muscle, wherefrom the bony part continues to the lower jaw. Bone of neck extirpated February 4, 1891. Dismissed March 20, 1891. New bone formation on location of extirpated bone from neck. *Microdactylia* of both thumbs and large toes and ankyloses of their phalanges.

CASE XLII.—Reported by R. Gordon McDonald, in 1891. Female, aged 4 years. Family history negative. First symptoms appeared at 2 years of age—lumps appeared on left side of neck about middle of *sternocleidoid*. Later on lumps appeared on forehead and back (*dorsal vertebræ*). All lumps disappeared after some time. Head fixed; unable to move it; *sternomastoid*, *trapezius*, *stylohyoid*, *omohyoid* and *sternohyoid* ossified. Over left frontal eminence a large node; smaller one over left temporal bone and right border of occipital. Both elbows can be moved only eight inches from trunk; *teres major*, *latiss. dorsi*, edge of *pectorales major* becoming gradually ossified. *Scapulæ* fixed to ribs, right less than left. At inferior angle of both *scapulæ* large nodes; on left superior angle a similar one; right side free. Various-sized nodes along the vertebral spine from head to sacrum, also along lateral parts of several ribs and over crest of ilium. The superficial muscles of back are gradually ossifying. Ribs are also becoming fixed and she is unable to take deep inspiration. *Teres major* was, *experimentis causa*, exsected during hospital stay. It was totally ossified. Result as to mobility—none. No congenital deformity mentioned.

CASE XLIII.—Reported by Bilton Pollard, in 1892. Male, aged 9 years. Family

## MYOSITIS OSSIFICANS PROGRESSIVA

history neg. First symptoms appeared 6 months of age—lumps formed on lower angle of right scapula and nodules appeared on boy's ribs. In second year neck was getting stiff. In third year lumps formed between right iliac crest and the last rib. In his fourth year hard nodule formed a little below patella after fall on right knee. New lumps formed, his left arm stiffened and bony bands appeared in his neck up to his seventh year, but none during his seventh, eighth and ninth years. Neck quite stiff; rotation and raising of head barely possible. Bony band between chin and sternum extending from the lower border and from the right of lower jaw to right side of cricoid cartilage, there dividing into two, the right ending just above sternoclavicular articulation, left above clavicle between the two heads of left sternocleid. R. arm can be raised to angle of 45 degrees only together with scapula. Axillary folds contracted. Scar there from bony operation five years ago. Beneath it bony plate slightly movable, apparently in latiss. dorsi. Pectorales and deltoid hardened; hard mass on border of radii two inches above styloid process. L. arm equally stiff. In course of teres major runs thick band of bone, extending free from inferior scapula angle almost to humerus. Bony projection at left-elbow bend connected with biceps tend.; another mass a little deeper down involving brach. antic. Spine very rigid, and on both its sides bony nodules and plates in dorsal and lumbar regions. On outer and post. part of r. thigh flat osseous plate; smaller plate on left. On each thigh strong bony spiculæ, extending from adductors tubers. upward to tend. of adduct. magnus for about two inches. Irregular-shaped movable plate between lower fold of left condyle of femur and patella and a smaller one near outer tuberosity of tibia. Inner borders of both tibiæ show bony growths just below tuberosities. No congenital abnormalities mentioned. Pieces of bone were excised to improve mobility of arm and neck, but they soon formed again, and the slight improvement gained by the operation was lost.

CASE XLIV.—Reported by Luigi Bernacchi, in 1892. Male, aged 7 years. Family history neg. First symptoms appeared at 3 years of age—muscles stiffened in nape of neck without inflammatory symptoms. Small hard tumors formed in right cucullaris over tuberosities of frontal and parietal bones. About a year ago stiffness of humero-scapular joint appeared. Left arm could not be raised above horizontal line. New hard osseous growths have formed along the spinous processes of dorsal vertebræ, and growths in lumbar muscle so as to form line from neck to os sacrum. Two cervical projections over frontal tubera. Head slightly inclined forward and to the left; movements restricted. *Neck and back*; from the occipital insertion of right trapezius near middle, irregularly formed osseous tumors forming one solid mass of muscles. *Thorax and abdomen normal*; small exostoses above styloid process of radius. Posterior wall of axilla shows fibrous consistency and is shorter than normal. The scapulæ are united by bony ridge about their middle; an osseous tumor within the dorsalis major can be felt, size of thumb and somewhat movable. No mention of microdactylia. (Photograph of boy shows indication of microdactylia of thumbs.) No mention made in text.

CASE XLV.—Reported by Ludwig Rabek, in 1892. Female, aged 3½ years. Family history neg. First symptoms appeared at 6 months of age—hard nodules in region of shoulder-blades. Parents noticed movements of upper extremities were restricted; movements of lower jaw have been impeded since one year before. Difficulty and restriction of movements of lower jaw and both upper extremities. The distance ad maximum between the front rows of teeth is only 0.5 cm. Right masseter hardened and hypertrophied. The arms cannot be raised to horizontal line. Right elbow-joint contracted, extension incomplete, flexion normal. Shoulder-blades only slightly movable. Osseous tumors arise on many parts of the body, mostly on back. Large one in right axilla extends to latiss. dorsi as far as scapula line of tenth rib. Another on left side reaches eighth rib. There are four more in the scapula and one in lumbar region, all in the muscles. Right biceps changed into hard movable

## JULIUS ROSENSTIRN

mass united with shoulder muscles. Small pea-sized nodules in right pectoral. Big toes of both feet contracted outward and downward so that each lies under the second toe. (Microdactylia and hallux valgus.) Congenital deformity.

CASE XLVI.—Reported by J. Brennsohn, in 1892. Male, aged 20 years. Family history neg. First symptoms appeared in early childhood—a certain clumsiness of motion and elevation of right shoulder. Later a scoliosis, and three years ago right arm grew stiff, then left arm, then the trunk and subsequently the legs. Had to give up work in factory. The muscles of the posterior cervical region are completely ossified; all the anterior muscles feel rigid, especially the scaleni and right sternocleido; below the right lower jaw the mylohyoid forms a bony growth like a stalactite. Mobility of head nominal; shoulders quite stiff; arms cannot be abducted from side of thorax. Left elbow-joint ankylotic; right only partly movable. Both anterior and poster. walls of axillæ ossified; also deltoids, serrati antici maj. and intercostales. The lower parts of left biceps and brachialis indurated and tense. Hand- and finger-joints free. Ramified osseous growths from both cristæ ossis ilii into both glutæi max. Some mushroom-like exostoses from left crista ossis ilii. Both big toes microdactylic (metatarsus primus?) and in valgus position. Brennsohn ascribes it to missing of basal phalanx. No X-ray taken.

CASE XLVII.—Reported by V. P. Gibney, in 1893. Male, aged 10 years. Family history neg. First symptoms appeared at 5 years of age—was admitted to the hospital at this time to sever a strip of ossified muscle, but was removed by parents before operation. Subsequent course of disease unknown. Head tilted to right; upper extremities bowed so that thumbs touch elbows of opposite side; elbows abducted from trunk; shoulders stiff; right clavicle has extra curve at outer half; greatest convexity posteriorly. Left clavicle curved entire extent; convexity toward neck. From its middle springs irregular bony mass elevated half an inch. Base of this mass spreads in clavicular portion of pectoral. major and continues in its pectoral part, terminating in anterior wall of axillary space. Over sternal articulation of second rib small exostoses tapering off into pectorales mass. Similar ones over sternal end of third and fifth ribs. None on right; about middle of fifth rib, just in front of axilla, a bony enlargement extending back and downward, involving the whole area of latiss. dorsi and serratus magnus. March, 1893; the bony tendon of the right latiss. dorsi was divided and a piece of bone about one inch wide was excised, but new bone was thrown out and rendered void the effect of the operation. An osseous tumor the size of a peanut over tendo achillis which was removed did not return. No mention of microdactylia. Private advice kindly given me says that author failed to take notes about presence or absence of microdactylia.

CASE XLVIII.—Reported by R. Virchow, in 1894, and Bollinger, Linsmeyer, Kraske, Wollenberg, A. Weil, Nissim and Weil, Faulkner, Lyon, De la Camp, Ponfick, Birch-Hirschfeld. Male, aged 29 years. Family history neg. First symptoms appeared at 18 years of age—swelling and pain in maxillary articulation. Pain, swelling and final ankylosis of maxillary and various joints of extremities. Osseous masses on both sides of median line of neck. Bony elongation from right process coracoideus extending along tendon of that muscle and of the short head of the biceps for about 5 cm.; somewhat less on the left side. Ankylosis of right shoulder-joint. Ossification of right triceps extending upward to teres and dorsalis muscles, also some ossification in anconeus. Small cartilaginous nodules on the tendons of abductor longus and extensor brevis pollicis. The left arm shows the same changes as the right and a cartilaginous, fork-like formation over elbow-joint. Irregular osseous masses in upper part of trapezius and over lumbar region projecting at various angles and of varying sizes. Bony formations project from the sacro-iliac articulation to the right and left sides, reappearing in the iliac fossæ and extending to the coxofemoral joints, which they immobilize, reaching to the great trochanter and into the muscular insertions thereon. Thickening of tuberosity of right tibia; exostosis of head of astragalus.

## MYOSITIS OSSIFICANS PROGRESSIVA

Heads of metatarsal bones thickened as in arthritis deformans. Left leg shows similar changes as right one; osseous mass extending from popliteal fossa down between the two bones and immobilizing knee-joints. Thickening and ankyloses of ankle-joints. Ankyloses of big toes; from the tip a long bony appendix is branching off, size of a walnut, like a supplementary toe. In the discussion of this case Gerhard drew attention to the microdactylia of a big right toe. Case began rather late in life, but except this one symptom, has all the earmarks of the disease. The epiphyses of the long bones are so soft that pins can be pushed into them.

CASE XLIX.—Reported by A. A. Kisel, in 1893, 1906-1909. Male, aged 1 year and 7 months (13 years?—16 years?). Family history negative. First symptoms appeared at 1 year of age—swelling in nape of neck size of small walnut; no accompanying pain or inflammatory symptoms. After three or four weeks most of tumors disappeared without leaving any trace. Some others seemed to soften and discharged a puriform liquid. All without symptoms of inflammation. *Later*, head has been drawn forward and posterior part of neck has become very rigid and quite immovable. Impossible to turn head backward. Head drawn forward. Chin touches thorax. Motions of head very limited. The entire posterior part of neck appears considerably swollen; the borders of the muscles very much thickened. The sternomastoid and pectoral muscles present some changes. Movements of right shoulder very limited, especially abduction and elevation. April 25, 1893, a new tumor appears in the right flank, with the same character as the others and reaching size of a small walnut. When 16 years old bone-like hardening of ant. and post. cerv. muscles, muscles of thorax, scapulæ, shoulders, abdomen, osteal attachment between twelfth rib and crista ilii; exostoses on lower jaw and scapular spine; head inclined forward and to the left, and immovable; head, vertebræ, neck, thorax and upper extremities in shoulders all one mass of bone; limited movement in elbow-joint. Improvement at first under K. J. At 13 years walked well, but now unable to leave bed. Extirpation of small tumors; tumor shows muscle cells much swollen, reddish-yellow and oedematous. Microscopical examination; very young embryonic tissue cells large star-shaped cells with various-formed processes; very few striated muscle fibres, very much modified. Microdactylia of both big toes, with aplasia of first phalanx.(?)

CASE L.—Reported by Weldon Carter, in 1894. Male, aged five years. Family history neg. First symptoms appeared at 4 years of age—lumps in back. Never any complaint of pain or tenderness or inflammatory symptoms. Back and shoulders affected, especially latissimi dorsi. Both big toes in hallux valgus position; the phalanges of fourth and fifth toes were very short and slight webbing between second and third toes of each foot.

CASE LI.—Reported by Carl Hochhalt, in 1894. Male. Family history neg. First symptoms appeared at 19 years of age—pains and swelling in right triceps brachii. Pains and swelling gradually disappeared after three months, leaving osseous hardening; similar attacks repeated annually, involving new groups of muscles. Spinal column is rigid and ankylotic in all its articulations. The muscles of the back are changed into hard bony plates, especially both cucullaris and latissimi dorsi, exostoses trunci; mimic muscles of face intact, masticating muscles rigid. In order to allow nutrition patient had several teeth broken out. Pushes nourishment through gap. Both shoulder-joints stiff, both triceps changed into hard bone masses. Under left gastrocnemius large bony mass size of one and a half fists reaching partly up into popliteal fossa, contracting knee-joint. Left glutæus changed entirely into shapeless bony mass. No congenital abnormalities mentioned.

CASE LII.—Reported by E. Lexer, in 1895. Male, aged 50 years. Family history neg. First symptoms appeared at 35 years of age—pains in left side of chest. Pains disappeared after three weeks, leaving soft-covered nodules the size of hazelnuts at the painful places. These nodules also disappeared after a short time. In 1884 again pains in left side of chest and back. Soft swelling in left lumbar region size

## JULIUS ROSENSTIRN

of palm of hand. Later grew smaller and harder. Till 1888 recurrence of symptoms every summer. The lumbar swelling grew larger and harder and spread in front and upward to the axillary line. He again fell sick in 1890, then remained free until 1894. New nodules had formed and a very severe recurrence took place, with pain in both shoulders and in muscles of both upper arms. In the lower part of the pectoralis major a cartilaginous nodulous tumor, size of a hen's egg. At the margin of the trapezius muscle, level of sixth cerv. a similar but smaller tumor. On the left deltoid a growth size of a man's fist, of fibrous consistency, covered by muscle and slightly movable over the underlying bone. Both upper arms show under atrophic biceps a cord-like, bone-hard mass, movable on humerus and occupying the place of the brachialis internus. The right forearm shows in its middle a bone-hard growth about 5 cm. long and wide, movable sideways under the supinator longus. Some of the other muscles show peculiar indurations. In the back the left latiss. dorsi nearly entirely pronated by a tumor formed by the amalgamation of a number of smaller ones, which reach from the crista ilii to the vertebral column and up to the scapula. In the region of the left infrascapular a very large free tumor from spina scapulae reaching to the acromion. In right serratus, over fifth and sixth ribs, a very hard flat tumor. No deformities of fingers or toes (specially mentioned). Excision of deltoid tumor. Ten days later the tumor over fifth and sixth ribs was excised. Patient left clinic December 6, 1894. Letter of March, 1895, mentions several muscles of neck and lower extremities involved. Microscopically the sections show acute and chronic inflammation and induration of the inter- and intra-muscular connective-tissue (perimysium) hypertroph. changes from which the formation of bone tissue starts. Atypical case. Commencement of disease very late. Ossification not shown.

CASE LIII.—Reported by Stephen Paget, in 1895. Male, aged 7½ years. Family history negative. First symptom appeared at 4½ years of age—swelling behind left ear. Since that time various swellings and growths have appeared, some disappeared again, while others remained in different parts of his body. Holds head in wry-neck position from contraction of left sternocleid. A small nodule of bone in the anterior edge one inch above clavicle. In each pectoral muscle irregular bony nodules moving with the muscles. In the right pectoral they are dispersed all through the substance of the muscle and continuous with plates of bone under the deltoid and along the latiss. dorsi. Latiss. dorsi contains a sharp bony ridge in the axilla; the serratus magnus shows hard nodules at its origin. The bony masses in the l. pectoral. are also continuous with bone in the fascia beneath deltoid. Along each side of spinal column a hard bony ridge, more marked on left than right side, seemingly part of latiss. dorsi. From ant. fold of each axilla a narrow hard cord, thickness of about No. 5 English catheter, runs right down the level of iliac crest; they are not bony. Both big toes are shortened and turned outward in metatarsal phalangeal joint, and the phalanges turn under the second toes. (Microdactylia-hallux valgus.)

CASE LIV.—Reported by Fuerstner, in 1895. Female, aged 15 years. Family history neg. First symptoms appeared at eight years of age—nodules size of pigeon eggs appeared and disappeared again after five or six weeks. Since the age of eight or ten months hindrance in motions of neck and arms from nodules appearing in these regions. Position of head and vertebral column stiff; rigid walk. All movements of head very limited through tension of hardened but nowhere nodulated muscles of the neck. In the long muscles of the back, however, there are several hard bony thickenings. At the inner margin of the scapula there are several osseous protuberances and bony plates in the latiss. dorsi. The arms are flexed most of the time; cannot be abducted, as tendons of pectorales major are hard cords. Both biceps muscles are hard as well as presenting a peculiarly sharp tendon; the lower parts of triceps feel ossified. On the left side the same changes are present in a lesser degree. Abdominal muscles and lower extremities show but slight changes.



## MYOSITIS OSSIFICANS PROGRESSIVA

During observation of this case a swelling occurred in the left biceps muscle within a few days. It was of a doughy consistency, the skin was slightly reddened and there was considerable pain upon pressure. A small piece of the biceps muscle was carefully excised and the *microscopical examination showed a very pronounced and intense hypertrophy of the interfibrillary connective tissue*, especially in the neighborhood of the blood-vessels, while the muscular tissue itself remained completely unchanged. Both thumbs and little fingers considerably smaller than normal. Microdactylia of both big toes.

CASE LV (?).—Reported by Eichhorst, in 1895. Male, aged 24 years. Family history neg. First symptoms appeared during time of observation in hospital—swelling and hardening of muscles of left calf. Subsequent course of disease unknown. The swelling extended from the upper end of Achilles tendon 5 cm. into the muscles of the calf. Skin at first red and inflamed, afterwards resumed its natural state. While the affected parts of the muscles decreased somewhat in size they so increased in hardness that they felt like bone; the skin was movable over them, and in so doing crepitation was distinctly felt. Patient had spina bifida. No change of condition was felt when patient left hospital or was observed later; no progressive ossifications in other muscles. No deformity of fingers or toes. This, as well as the former case reported by Eichhorst's assistant (No. 37), is a doubtful, atypical case.

CASE LVI.—Reported by O. Paget, in 1895, and W. P. Herringham, in 1899. Female, aged 5 years. Family history neg. Said to have suffered very intensely from rickets. First symptoms appeared three weeks before date of examination—lump in back, which gradually increased in size more rapidly since the last week. Subsequent course of disease unknown. Head fixed by two indurated nodules and hard sternocleido. Right pectoral, left pectoral and latiss. dorsi all in same condition. General health fair (pigeon-breasted); two growths, smooth, oval and firm, about 4 x 2" symmetrically on each side of vertebral column just below inferior angle of scapula. Similar growth, size of large walnut, attached to inferior angle of each scapula. Ossified node on external edge of right bicipital groove. Ossified node on forehead, the latter due partly to inability of child to protect itself when falling forward. Bilateral microdactylia of big toes ascribed to absence of first phalanx. Observation in 1899 shows marked progress of disease on skull, abdominal walls and both ulnæ; movements of shoulder very restricted. Skiagram of big toes shows microdactylia and hallux valgus, due to: (1) Irregularity and shortness of metatarsal bones; (2) thickening of outer side of first phalanx, tilting bone outward; (3) synostosis of the first and unguinal phalanx at oblique angle. Exostoses on first phalanx of the right middle finger.

CASE LVII.—Reported by Harry Lockwood, in 1896, and Raymond Crawford, in 1899. Male, aged 4½ years. Family history neg. First symptoms appeared at 2½ years of age—German measles shortly before malady showed itself. Subsequent course of disease unknown. Body and head bent forward. Masseters hardened; right and left pectorales major ossified and fixed. Large masses of hard material on both sides of spine. Microdactylia of thumbs and toes.

CASE LVIII.—Reported by Zoege v. Manteuffel, in 1896. Subsequent course of disease unknown. Demonstration of a skeleton of a case of myositis ossificans progressiva, where the products of the disease could be seen from head to foot. The osseous neoformations correspond to the course of the muscles. They had developed in their connective tissue nearly everywhere independently of the skeleton, so that they could not be ascribed an exostoses. A synostosis of the shortened phalanges of the big toes. The thickening of the mandibulæ seemed to be caused by an ossification of the muscular insertions. Microdactylia.

CASE LIX.—Reported by Ludwig Pincus, in 1897. Male, aged 25 years. Family history neg. When 13 years old fell on back from a height of two metres. First symptoms appeared at 14 years of age—observed in nape of neck two little

## JULIUS ROSENSTIRN

nodule-like glands, but they disappeared later spontaneously. Later back, shoulders and left arm again attacked after trauma. Causes of attacks are generally traumatic. After a prolonged quiet an attack of violent pains and swelling, first on the left, then on the right side. Disease particularly pronounced in neck, back and surroundings of neck, surroundings of axillary fossæ, masseters, upper arms and thighs. Scoliosis, caput obstipum, exostoses and hyperostoses. The right carpo-metacarpo articulation pollicis ankylotic; thumb smaller than normal. Microdactylia of both big toes, with hallux valgus. Faradic contractibility diminished. Strong fibrillary spasmodic contractions spontaneously, but more pronounced when touched. Venous stasis, especially in legs.

CASE LX.—Reported by Von Kryger, in 1898. Female, aged  $4\frac{1}{2}$  years. Family history negative. First symptoms appeared at  $2\frac{1}{2}$  years of age—with inflammatory symptoms, a painful bluish-red nodule formed on back a little to the left of twelfth dorsal vertebra. The first tumor decreased greatly and in its place a round osseous protuberance appeared. At various times this process, nearly always accompanied by inflammatory symptoms, spread over back, nape of neck and vertebral column, abdomen and hips. At last face and neck were involved. From the rigid vertebral column emanate long irregular bony ridges; shoulders drawn up, lower arms flexed at right angles, only slightly movable. Hands crossed across the abdomen; legs slightly flexed at hips; right one abducted, immovable. Head inclined toward front and left; face turned somewhat to right; muscles of the cheek and mouth are very hard and mouth can be opened only half an inch. Everywhere in muscles one finds tough cords or bony ridges, some of which are connected with the skeleton, others appear to lie free in the tissues and nearly always follow the course of the fibres, rarely crossing them. From the ribs, the vertebræ and the pelvis real exostoses project vertically to the skin. Microdactylia of both big toes. They appear to have only one phalanx, which stands in valgo position upon the metatarsus. (No X-ray picture.)

CASE LXI.—Reported by Walter Stempel, in 1898. Female, aged 7 years (?). Family history neg. First symptoms appeared at three and a half years of age—swelling in sternocleidomastoidei; later in cucullaris and various other muscles of thorax and body. Swellings felt hard at first, got gradually softer and disappeared, as did many other hard swellings in various groups of muscles during the course of three and a half years of observation. Last examination, January, 1898, showed ossification of both cucullares at occiput. Ossification of scaleni. Spine quite stiff. Fixation of lower jaw through hardening of masseters. Microscopical examination of muscle in first stages of hard swelling showed a hypertrophy of the outer fibrillary tissue, which by its rapid growth into the muscular tissue, separating and pulling the muscle fibres, causes an intramuscular hemorrhage, which, according to Stempel, represents the first cause of the swelling which is being followed by the fibrous tissue growth and later by ossification.

CASE LXII.—Reported by A. Roth, in 1898. Female, age (?). Family history neg. First symptoms appeared between 1 year and 9 months and 2 years of age—swelling in left side of middle of back. Another swelling at the left side of the neck followed closely; both disappeared after inflammatory symptoms. A similar process developed on the right side of the back, followed by swellings of both arms from shoulders to elbows and of various other places, like chest, abdomen and thigh. After a recedence of the inflammatory symptoms the afflicted parts showed hard exostoses-like masses. Arms grew stiff and lately the face became swollen. Patient perfectly stiff, as if in tetanic rigidity. Two small exostoses, size of peas, above left ear. Maxillary joints ankylotic. Mouth can be opened only  $\frac{1}{2}$  cm. The entire cervical muscular group forms one hard mass. The upper arms are bound together by a hard bony ridge in the anterior and posterior height of the axillary folds, comprising latiss. dorsi and pectorales major. Both scapulæ are fixed. Superior

## MYOSITIS OSSIFICANS PROGRESSIVA

and inferior scapularis of osseous hardness. In both upper and lower arm muscles various ossifications and enclosures of bone. Muscles of hands free, narrow strip of bone in musc. obliq. abdom. ext. On both sides, at origin of sartorius, ossification. Microdactylia of both big toes. Valgus position; only one phalanx reaching to first interphalangeal joint of second toe. Microscopical examination of a piece of new-formed osseous tissue shows the change of connective tissue to fibrous, cartilaginous and bony tissue. He believes in passive and active participation of periosteum, as he found hardly any bony pieces free in the muscular substance; also in a congenital diathesis brought into action by traumatism.

CASE LXIII.—Reported by Mr. Jacoby, in 1898. Male, aged 27 years. Family history neg. First symptoms appeared at 19 years of age—pains in ankle-joints and legs. Four years later pains occurred in hips and neck, and only two and a half years ago induration and ossification in the neck, involving all the contour of the neck. Another ossified mass between pelvis and lower edge of arch of ribs. Slight hardness in masseters, but patient can still open mouth well. July 14th: diagnosis confirmed by X-ray pictures characterized it as a bone-muscle disease, which leads to the formation of exostoses and direct ossification of the muscular connective tissue. No mention of congenital abnormalities.

CASE LXIV.—Reported by A. Salomoni, in 1898. Male, aged 12 years. Sound, healthy parents. First symptoms since birth—deformities in neck and arms. Gradual and steady progression. At the time of first observation, in 1897, deformities had greatly progressed. They occupied the entire neck and the two upper limbs in a very characteristic manner. A personal letter from Piacenza, Italy, where Salomoni is now head of a military hospital, explains that on account of being so taken up with his present duties he is unable to go further into the history of this case, and merely gives me what data he remembers. He says: "I extirpated from the neck and from the arm two pieces of the muscles where ossification had gone on. They were histologically demonstrated to be derived from the fibro-connective tissue and from the new-formed bone near the periosteum. The little patient underwent various pharmaceutical treatments, more for the moral effect than for any other purpose, and died at the age of 14 without my being able to continue the observation of his case." This case was reported in the record of the congress of the Italian Surgical Society of Turin, 1898. Salomoni's letter differs from the above-named report and that of the Italian Pathological Society in 1898, which reads: "A girl, 7 years; when 2 years old ossif. of l. sternocleido noticed; later r. side; then gradual swelling and ossif. of other muscles, sometimes with fever. Microdact. of thumbs and big toes."

CASE LXV.—Reported by Hendrick Burgerhout, in 1898. Male, aged 40 years. Family history neg. First symptoms appeared at birth—left shoulder-blade and left shoulder-joint stiff. During schooling years hip- and knee-joints became stiff. Later, after twentieth year, both elbow and wrist-joints stiffened. Ten years ago both maxillary articulations became stiffened so that he could not open mouth. Sits nearly immovable; head bent forward upon chest. Lower jaws very slightly movable. Distinct movement of temporales and masseters. Sternocleido and cucullaris feel hardened, but not osseous. Arms stand in strong adduction; scapulæ fixed; at lower outer angle not separable from the stone-hard mass which goes in the direction of teres majores and latiss. dorsi. The long muscles of the neck are connected from neck to os sacrum by irregular ridges of bone. Left arm, shoulder and elbow stiffened; left thigh flexed in hip-joint; left knee-joint and ankle-joint immovable. Large exostoses above patella in location of vast. externus. Right leg articulations resemble left side, but are freer. On inside of femur in place of adduct. hard exostoses extending from pelvis to insertion of adductor magnus. Microdactylia of thumbs and big toes due to smallness of metacarpal and metatarsal bones. Very exact examination of electric irritability of muscles and their nerves; both normal. For very thorough metabolical research work and conclusions of this case see text.

## JULIUS ROSENSTIRN

CASE LXVI.—Reported by Robert Jones, in 1899. Male, aged 16 years. Family history neg. Fell and injured his back.(?) First symptoms appeared at 13 years of age—growths on back started one year after his fall. Subsequent course of disease unknown. Growths on inner aspect of right jaw; small lump below right olecranon. Hard mass attached to upper two-thirds of femur; thickening at insertion of right ligamentum patellæ and at each tarsometatarsal joint. Over spinous processes a continuous line of hard cartilaginous or osseous material, extending from level of fifth dorsal to fourth lumbar spine. Strips projected from the upper and lower ends of this mass two and three inches long. They were movable in the surrounding tissues. No deformities mentioned.

CASE LXVII.—Reported by Morian, in 1899. Male, aged 4½ years. Family history neg. Fell twice down a flight of fourteen steps. First symptoms appeared at 2½ years of age—soon after second fall swellings in various muscular groups. Fell again after commencement of disease and struck on a stone with his neck. Ossification in right frontalis muscle, in both sternocleid., in the group of muscles from sternum to hyoid and from there to mandibula, in both pectorales major and deltoids, in both sacrolumbales and in both bicipites; in both cucullares and right altissimi dorsi. Both big toes show microdactylia.

CASE LXVIII.—Reported by Lyder Nicolaysen, in 1899. Female, aged 4½ years. Family history neg. First symptoms appeared at 2 years of age—stiffness of shoulder, neck and back. Subsequent course of disease unknown. Height 99 cm. Circumference of chest 45 cm. Posture: head bent forward; nape of neck, back and shoulder muscles completely ossified. In both axillary fossæ anterior and posterior folds are hard; greater part of pectoral. maj. ossified. Both scapulæ fixed to thorax. Trapezius bone-hard. Several nodular exostoses on back. Arms stiff and rigid. Hardened parts in musculature of upper arms. Excursion angles of both forearms about 45 degrees. Microdactylia of both thumbs and big toes.

CASE LXIX.—Reported by George Wilkinson, in 1900. Male, aged 13 years. Family history neg. First symptoms appeared at 2½ years—swelling in right side of back. Swelling in back subsided. Later isolated hard lumps appeared on back, remaining and spreading. Later stiffness in back. Lower jaw fixed, mouth can only be opened one-fourth inch, due to hard temporal fascia. Bony bars in right occipital bone. Thin plate of bone in occipital tendons and left trapezius, and hardening in right. Hard nodules in both sternocleidii and scaleni muscles. Big bony mass extending from left to right scapula. Two large hard masses in right lat., shoulders raised, scapulæ fixed. Nodule in left pectoral. maj.; two small ones on both sixth ribs. Respiration wholly diaphragmatic; shoulder movements, especially rotation, limited. On each femur large irregular mass of bone fixed to the upper end near great trochanter. A spike of bone extends on each side of glutæus maximus near its sacral origin. Another one down along outer side in tendon of fascia lata to below middle of thigh. Hard nodule in right rectus femor. Sharp nodule of bone projects below each tuberosity ishium. Angular bony nodule on posterior border of right tibia, three inches above int. malleolus. Movements of hips limited. Trunk bent somewhat forward, can sit only on corner of chair. Both thumbs microdactylic. Their metacarpals and phalanges are shorter than those of other digits and interphalangeal joints stiff. Both big toes microdactylic and with hallux valgus. Their two phalanges very short, their interphalangeal joints synostotic; nodules have formed without pain. Some masses that have appeared during the nine months of observation have disappeared again.

CASE LXX.—Reported by Morley Fletcher, in 1900. Male, aged 9 years. Family history neg. First symptoms appeared at 7 years of age—location (?). Subsequent course of disease unknown. Numerous exostoses on bones of head, one on the hand and one on each tibia. Thickening of bridge of nose. Both big toes small and terminal phalangeal joints ankylosed. Interesting association of numerous exostoses with

## MYOSITIS OSSIFICANS PROGRESSIVA

multiple neuro-fibromata; looks upon it as abnormal condition of mesoblast from which the mesoblastic structures were formed and as related to the condition of multiple exostoses and multiple neuro-fibromata.

CASE LXXI.—Reported by Theodore Schwickerath, in 1901. Female, aged 14 years. Family history neg. First symptoms appeared at 9 years of age—violent toothache. Two molars were extracted, whole left side of face swelled greatly after extraction of teeth and became permanently stiffened. Patient's jaws became so stiff that she could not open her mouth sufficiently for mastication. A piece of an osseous ridge 1 cm. broad and  $\frac{1}{2}$  cm. thick was removed from the left masseter to increase the possible excursion of the lower jaw. After the operation mouth could be opened 3 cm., but later it returned to almost its pre-operative status. Patient holds herself bent forward. Scar on left masseter where excision was made. Near upper insertion of left masseter a firm nodule, size of cherry pit; in its lower part an osseous ridge  $1\frac{1}{2}$  to 2 cm. broad. Lower jaw can only be opened 2 mm. Lateral movements entirely inhibited. Left clavicle enormously thickened. Left sternocleido a thick cord, shows osseous ridges in sternal and clavicular portion of the pectoral. maj., is as hard as bone, running toward axilla and ending at spine of tuberosity maj. In the middle of right scapula spine is an osseous nodule size of a walnut. Back scoliotic to left and shows partly flat, partly osseous protuberances. On both sides of vertebral column osseous plates; in latiss. dorsi and teres osseous nodules extending right down to region of sacrolumbalis. Right arm: very few excursions possible; adduction, abduction and rotation impossible; in the axilla a hard nodule. Left arm nearly normal; right thigh exostotic thickenings. Microdactylia of both big toes. No X-ray picture.

CASE LXXII.—Reported by Vaughan and Burton Flanning, in 1901. Male, aged 33 years. Father had same disease, dating from a fall in his fifteenth year on his right arm, where it began. The disease had steadily progressed, but had not reached the same degree as with his parent, when he died at the age of 33 from an accident. He had congenital deformity of both thumbs, there being no joint beyond metacarpal ones. Feet were not observed. First symptoms appeared at 8 years of age—left shoulder gradually became stiff without any previous injury or pain. At 12 years right knee began to stiffen and gradually became ankylosed. Following this came stiffness of left hip, sides of chest and lower part of back. At 26 years right arm from shoulder downward became swollen, red and tender. This disappeared, but left arm is fixed extended in shoulder and elbow. Wrist remained free. Two years later stiffening of right hip was noticed. During winter of 1898 difficulty in opening mouth began; passed off during summer, but returned worse the following winter. The following summer got slightly better, but had gradually increased since. Both erector spinæ nearly completely replaced by bone, preventing any movement of that portion of the spine covered by them. Deep muscles of back of neck similarly affected, only possible movement of head is slight rotation and flexion. Bony plates between several ribs; on each side and along ant. border of l. quadrat. lumb. was hard band fixing twelfth rib to crest of ilium. Movement of chest almost prevented by these growths except over first two ribs. The distal halves of both teres maj. and pect. maj. were bony. Close over the right fossa olecrani a bony mass, the size of a walnut, involved the whole thickness of the triceps. A similar growth over the upper ends of the ulnæ and radii. Both extens. quadriceps were almost replaced by bone; osseous growths round head of right fibula and enlargement of both int. malleoli. Both big toes had hallux valgus, with shortening and broadening of phalanges, which were fused together. No X-ray.

CASE LXXIII.—Reported by Wilhelm Rager, in 1901. Female, aged 14 years. Family history neg. First symptoms appeared at 2 years of age—swelling over os frontal. After that many painful inflammatory swellings in various parts of the body, leaving osseous hardenings. Mouth can only be opened  $1\frac{1}{2}$  cm. Both shoulder-

## JULIUS ROSENSTIRN

joints and right elbow-joint ankylotic. Ossification in both masseters, right sternocleido and platysma, both deltoids, pectoral. maj. and latiss. dorsi, quadriceps femoris, right soleus and insertions of both gastrocnemii and solei. Similar ossifications are distributed all over back of neck and the back proper down to lumbar vertebræ. Both thumbs and big toes are deformed, only half as long as normal. In the thumbs, due to two shortened and synostotically united phalanges; in the toes to stunted metatarsal bones.

CASE LXXIV.—Reported by Christopher Graham, in 1901. Female, aged 6 years. Family history neg. First symptoms appeared at 4 years of age—small lump between shoulders. Gradual but steady progression. Rarely any pain except during last three or four months. Ligamentum nuchæ solid throughout. Both pectorales and sternocleidi are more or less solidly ossified. Right biceps affected in its entire course; the left only in its upper part. Right trapezius is completely changed into bony mass; the left less so. Both latiss. dorsi are ossified. The deeper trunk muscles seem to be also similarly affected. Right external oblique ossified along the line of the linea semilunaris, below and externally from Poupart's ligament, a hardness and nodules can be felt. No microdactylia mentioned. Upon my inquiry, Doctor Graham kindly answered that he had no recollection of having examined the feet, the boy having been seen in the policlinical department only.

CASE LXXV.—Reported by H. D. Rolleston, in 1901. Male, aged 8 years. Subsequent course of disease unknown. Ossification in both sternomastoids and posterior scaleni muscles and in the scapular attachments of the rhomboids. Bony plates in lower parts of latiss. dorsi. Hard masses in region of erectores spinæ over lumbar spine. Both biceps and brachialis ant. muscles extensively ossified. Both recti abd. are affected. Exostoses on both tibiæ near insertion of sartorius muscle and on both frontal bones. Microdactylia of both thumbs and toes, due to dwarfing of metacarpal bones. Commencing ankyloses of interphalangeal joint. Both hallux valgus, with exostoses. Exostoses on first phalanx of right middle finger. (Same peculiarity in Doctor Herringham's case, No. 56.)

CASE LXXVI.—Reported by George Carpenter and Walter Edmunds, in 1901, and Cyril Nitch, in 1908. Female, aged 4 years. Family history neg. First symptoms appeared at 3½ years of age—two weeks after both tonsils were removed a hard and tender swelling appeared on right side of neck. The first swelling subsided after a couple of weeks. Two weeks later a swelling appeared on left side, which also disappeared again. A short time afterward the mother noticed a hard mass on right side of neck, and a few months later similar bumps in left pectoral, scapular and lumbar region. *Condition in 1901*: Spicule of bone in left sternocleido. The neck muscles of same side infiltrated and hard. Below chin bony infiltration size of pea in geniohyoid muscle. Plates of bone can be felt in right sternohyoid, right coracobrachialis, left pectoralis major and erectores spinæ. February, 1905: back and neck quite rigid; movements of shoulders very limited. The early pea-like prominence of bone in geniohyoid has now become a long spinous process. Osteoid tissue in form of plaques, bosses and spicules can be felt in both erectores spinæ, latiss. dorsi, trapezii and pectorales, right rhomboideus major and minor, the sternomastoids, right vastus externus and in popliteal space. Microdactylia of both big toes. 1901: excision from a muscle that showed gray infiltration revealed under microscope fibrocellular tissue infiltration; no micro-organisms.

CASE LXXVII.—Reported by Menard and Tillaye, in 1902, and Comby and Davél, in 1904. Female, aged 5½ years. Family history neg. First symptoms appeared at 17 months of age—swelling (where?). Swelling always preceded hardenings and ossifications (where?). Osseous hardenings in post. cerv. regions and down the back to right axilla, extending toward thorax and to the inner border of humerus; and also to the dorsalis maj. On the left side similar changes in the trapezius. Two and a half years later: slow, but steady development of new osseous hardenings.

## MYOSITIS OSSIFICANS PROGRESSIVA

Stalactite formations in axilla, reaching down along humerus to elbow. Head, arms and back stiff, and movements extremely limited. The thumbs and little fingers of both hands as well as the two big toes are undersized. (Microdactylia.) Woodcuts of hands in later observation show microdactylia of thumbs depends on shortness of metacarpal bones. No X-ray in either communication.

CASE LXXVIII.—Reported by Fabio Rialta, in 1902. Female, aged 16 years. Family history neg. Father suffered for thirty years from arthritis of the left hand. Vaccination (?). First symptoms appeared at 2 years. After vaccination swellings appeared on head, from size of pigeon's to hen's egg. The entire left arm became swollen, then subsided. The same occurred then on right arm. Later both arms contracted, with flexion of forearm and adduction of upper arm and inability to raise them. A gradual, continual increase of all these symptoms. The neck swelled and remained stiff. After swelling had subsided some hard indolent nodules developed over the dorsal spine of the scapula the size of those on the head. From the superior dorsal region these formations descended, increasing in number every spring for about twelve years until two years ago, when they stopped. Height 138 cm. Head flexed rigidly toward right and forward; neither active nor passive rotation, extension or flexion of head possible. Vertebral column equally rigid; muscles of neck atrophic; sternocleido contracted and hard. The posterior cervical ligament ossified, with osseous plate; another one runs from seventh cervical downward; ossification of left levator scapulæ and cucullaris. The tendon of the pectoralis major is ossified to its insertion. Arms crossed over chest; forearms in exaggerated flexion; osseous plates in biceps, the caput brevis completely enveloped with an osseous growth the size of palm of hand. The anterior wall of axilla also ossified and posteriorly is an ossification extending on both sides from dorsal. maj. to scapular angle and to vertebral column. R. biceps has ossification similar to but less pronounced than left. Processi spin. below seventh dorsal are hidden by ossified masses. Column is slightly scoliotic and absolutely rigid. Respiration completely abdominal, slightly thoracic at base. External genitals normal in size, but hairless. Microdactylia of both big toes. Shortness of first metatarsal bone and hallux valgus. Hemidrosis facialis (sinistra).

CASE LXXIX.—Reported by M. Ferraton, in 1903. Male, aged 18 years. Family history neg. First symptoms appeared at 3 years of age—previous to first swelling in scapular region a three months' period of pain, sleeplessness and loss of appetite. After swelling subsided ossification set in. Ossification of right masseter; left more atrophied. Right sternocleido hard; the greater part seems to be ossified. Pect. maj. and deltoids contracted and atrophied. On the right humerus anteriorly two round exostoses, size of hazelnut, firm upon the bone; on the left side similar outgrowth. Another exostosis on the pelvis on crista iliac. post. to its edge, close to ossified insertion of dorsalis maj. Several large ossifications on the inner side of the right thigh, under middle and large adductors. Right foot: second toe much larger than the others, microdactylia of big toe; on left foot a synovial bursa on first phalanx of big toe. Hallux valgus. (No X-ray.) Operation on masseter muscle; exsection of small piece of muscle. Patient can open his mouth.

CASE LXXX.—Reported by Julius Michelson, in 1904. Female, aged 18 years. Family history neg. First symptoms appeared at 7 years of age—both thumbs over both flexor. brev. hot and inflamed. Later thumbs grew stiff and nearly immovable. Kyphosis and scoliosis at cervical and dorsal spine. Cervical spine stiff, only minimal movements possible in rotation, extension and flexion. Same conditions prevail in dorsal and lumbar portions. Muscles of neck hard. (X-ray shows no ossification.) Pars clavicularis of left pectoralis maj. shows bony deposit extending over the shoulder-joint and into the axilla; this mass can be moved to and fro. A similar mass is on the right side. Both thumbs, are ankylotic between first phalanx and metacarpus. The little fingers are shortened. (Second and third phalanges

are shortened.) Both big toes are 2 cm. shorter than normal and their two phalanges in firm osseous union. The little toes also are shortened and their second and third phalanges ankylotic.

CASE LXXXI.—Reported by Clito Salvetti, in 1904. Female, aged 4½ years. Family history neg. First symptoms appeared at 10 to 12 months of age—swelling observed in the left cervical region. This swelling disappeared, but was followed by two elastic swellings on the occiput, which persisted until now. Neck stiff. Hard infiltrations of muscles back of both supra- and infra-spinosi, and tumor-like indurations size of hen's eggs. Left pectorals like an inflexible plate, preventing the shoulder movements entirely and those of the forearm partially. The left Mohrenheim fossa absent. Neither fingers nor toes show microdactylia. (Specially mentioned.)

CASE LXXXII.—Reported by A. P. Beddard, in 1905. Male, aged 37 years. Family history neg. First symptoms appeared at 7 years of age—bony growths began to develop in muscles. Subsequent course of disease unknown. Bony growths can be felt in innumerable parts of his muscular system. Some of them are movable within the muscles; the majority are fixed, having developed in tendons and insertions of muscles. Ankylosis of joints has developed to such an extent that patient is entirely helpless and absolutely bed-ridden. No mention of microdactylia.

CASE LXXXIII.—Reported by A. Gaster, in 1905. Male, age (?). Grandfather. father and three sons were afflicted with same disease. Subsequent course of disease unknown. Doctor Gaster states in the discussion of Doctor Beddard's case (presented by Doctor Russel) that he knows a family in which the father and grandfather had myositis ossificans progressiva. The three sons suffer from the same disease. Mother and two daughters are free from it; two sons have two baby daughters without a trace of the disease.

CASE LXXXIV.—Reported by Nové Josserand and René Horand, in 1905 and 1912. Female, aged 7½ years. Family history neg. First symptoms appeared at about 6½ years of age. Subsequent course of disease unknown. On both sides of scapulæ a tumor size of mandarin, the right a little larger than the left. Osseous indurations on the trapezius and in the upper part of right sternocleidoid. In lumbar region an osseous plaque near the iliac crest. In right and left pectorales major and minor are ossifications. Both big toes are shortened and show hallux valgus. Last phalanx of each little finger is noticeably turned outward. In 1912, on account of progression of disease, patient was induced to submit to X-ray treatments. Had twenty-six treatments, all below five Holzkecht's units. Effect surprising; patient straightened out, can again walk with youthful agility up and down stairs, sit and lie down unaided, and is no more imprisoned in her shell. As to duration of this improvement, author promises watchful waiting.

CASE LXXXV.—Reported by S. Biegel, in 1906. Male, aged 7 years. Family history neg. Born in occipital presentation; was delivered with forceps; large hæmatoma over occiput (?) Head at first drawn out lengthwise, but after few days normal shape restored. First symptoms appeared at 3 months of age—parents observed apparent displacement of parietal and other bones of skull, and a little later swellings on head size of pea to a marble. These swellings disappeared after a few weeks. Previous to appearance of swellings on top of head and on neck there was always stiffness vertebral column and cyanosis of head and of extremities, which again disappeared as soon as swellings were fully developed. This lasted with intervals to his third year. From third to sixth year he was quite well, although a slight knock or a slight fall always produced hard nodules, which disappeared again after a few days. About one and a half years ago he fell upon his head. From this time on he grew stiff. Can now hardly move. At first his neck grew stiff and he could not turn his head. Later maxillary muscles stiffened; mouth could not be opened more than enough to admit flat spoon. This condition improved so that he



## MYOSITIS OSSIFICANS PROGRESSIVA

could open his mouth again, but stiffness developed in the muscles of his mouth. *September, 1905*: a stone-hard thickening of both sternocleidæ; *December*, stiffening of upper arm and thorax muscles of both sides, in left side osseous ridge remained. Head bent to left side and forward; breast and abdomen bent backward and to the right. Both sternocleidæ and platysmæ feel like hard plaques. From left side of axilla a bony ridge goes over pectoralis major, serratus ant. maj., obliquus ext. abd. Both cucullaris are hard, the edge of the scapulæ strongly projecting. The right arm hangs along the side of the trunk and the hand rests on the thigh; the left is abducted in shoulder-joint and flexed in elbow so that the hand rests nearer to Poupart's ligament. Both are limited in their movements, the left more than the right. All the hand and finger-joints are well movable. Both legs are flexed in hip- and knee-joints. Hip-joints are stiff, but below these joints limbs are well movable. Microdactylia of both thumbs and big toes.

CASE LXXXVI.—Reported by E. J. Maxwell, in 1907. Male, aged 10 years. Family history neg. First symptoms appeared at 5 years of age—hardness of spine muscles. Gradually increasing ossification of muscles of spine near the sacrum and back, spreading upward. Spine and head absolutely rigid; growths of bones branched out from spine to scapula. Breathing mainly abdominal. Only liquid could be swallowed; condition of masseters and jaws not given. Bone could be felt on right side of larynx. No mention of microdactylia. No X-ray picture.

CASE LXXXVII.—Reported by A. E. Garrod, in 1907. Male, aged 21 months (weight 23½ pounds). Family history neg. First symptoms appeared at 3 or 4 months of age—lumps appeared on head. Child healthy at birth, but soon began to waste. Improved to normal condition, however, when nursed by colored woman. The lumps disappeared completely three or four months later. At 16 months fresh swellings appeared upon back, increasing in size for some time, then disappeared again, while fresh swellings had appeared in new locations. Swellings upon back, two in front of chest in outer parts of pectoral muscles; one on the head over occipital bone. Swellings disappeared and new ones came during the time of observation, *May 29, 1906, to July 20, 1906*. Patient left without any bone formation having occurred in any of the swellings during this first period of observation; no constitutional disturbance or particular pain seemed to accompany the appearance of the swellings. One year later reported: right biceps is contracted and feels stony hard; stiff elbow and right shoulder-joint. A lump size of mandarin orange over infraspinatus, of ivory hardness. Other lumps forming. Health seems to be perfect. Microdactylia of both big toes in valgus position. Over the inner aspect of the head of each first metatarsal bone was a small linear scar, suggesting that an extra digit had been removed.

CASE LXXXVIII.—Reported by Paul Krause and Max Trappe, in 1907. Female, aged 16½ years. Family history neg. Severe cold (?). First symptoms appeared at 12½ years of age—tired feeling in arms and legs, and back began to feel stiff. Pains in various parts of her body; knees sensitive to pressure; stiffness of legs increased, arms became bent and stiff. About the winter of 1905-06 appeared a swelling in lower sacral region, which disturbed the patient when lying down. A bone-hard nodule was removed from right ant. axillary wall; since then she can move right arm better than left. A swelling of the feet appears now from time to time, but soon disappears again. The swellings when softened evacuated an emulsion of amorphous calcium salts. Later the skin became sclerotic, dry and immovable over its underlying tissues. Patient 155 cm. tall; weight 34.5 kil. Very strongly developed *arci zygomatici* and large pupillary distance (65 mm.). Slight left scoliosis in lower dorsal and lumbar portion. Pect. tendons are hard as board, contain several nodules hard as cartilage. Scapulæ show over spinæ several small tough nodules; over right acromion a hard tumor size of a cherry pit. In upper and lower parts of both biceps are several small nodular imbeddings. In right cubital fossa a

collection of nodules connected with the lacert. fibrosus, continuing to the forearm; on the left only one small nodule. *Hands show nothing abnormal.* Over right sacroiliac joint nodule of bony (?) consistency, size of millet grain, and to the left of the spinous process of fourth and fifth lumb. vert. a soft swelling, size of a silver dollar, sensitive to touch. On both sides of os coccyx three globular swellings about  $2\frac{1}{2}$  cm. In the gluteal muscles small nodules size of cherry pits. The adductors at their origin, at os pubis, as hard as board. At the inner epicondylus of the left thigh two nodules of cherry-pit size adherent to skin and movable with it over the underlying tissue. Toes all well developed. The microscopical examination of the excised axillary nodule showed only a sort of fibrous hyperplasia and infiltration of the muscular tissue. The late beginning ( $12\frac{1}{2}$  years), after a cold, the absence of new pathological cartilage- or bone-formation or even of calcification in the X-ray pictures or in the excised nodule, the softening, liquefaction and discharge of thick, mucous emulsions, the dryness, sclerosis and immovability of the skin, the absence of microdactylia—all these features do not fit into the picture of myositis ossificans progressiva, but stamp the case as belonging to *myositis fibrosa*. Since writing I found that in a later article the authors\* abandoned a diagnosis of this case as myositis ossif. progressiva.

CASE LXXXIX.—Reported by Roberto Solè, in 1908. Female, aged 7 years. Family history neg. First symptoms appeared at 4 years of age—parents detected two cords in abdominal wall extending from thorax to pubes, and according to description of location were the recti muscles. They felt hard to the touch. These two cords persisted in their hardness till child was 4 years old, when they gradually disappeared and abdomen grew normal. Often fell in the first years of her life and at  $2\frac{1}{2}$  years had severe fall, after which and since numerous hard tumors appeared, mostly on neck and thorax. Suffered severe pains at 3 years. These disappeared at 4 years, but tumors remained and increased in number and size. Head bent forward and fixed firmly. In the ant. region of the neck two stiff hard cords in the location and direction of the sternohyoids and mylohyoids, like rosaries on account of their interspersed osseous infiltrations along their course. A similar formation exists posteriorly on both sides of the cervical vertebral column, from the superior part of the thorax to the base of the occiput. Thorax in kyphosis position, but not angular. It shows numerous hard osseous tumors over the ribs, some intimately adherent to them; others simply on their borders. Both pectorales major and latiss. dorsi are ossified, keeping the arms in their rigid posture in the shoulder-joints. Some few nodules of ossified tissue in triceps brachii. No mention of microdactylia, but in one of the photos of this case the presence of microdactylia of the big toes can be plainly seen.

CASE XC.—Reported by Warren Walker, in 1908. Female, aged 7 years. Family history neg. First symptom appeared at 6 years of age—stiffness of neck. Later a swelling appeared in the right pectoral muscle and others in different parts of the body. These swellings were painful to the touch. Marked prominence of occipital tendon of right trapezius, muscle very hard and stands out like a cord. The scaleni and sternocleidii also involved, but not so extensively. Head fixed on sternum, no rotation possible. At lower angle of left scapula exostosis one inch high, another one over ninth rib just below the scapula is covered by a mass of indurated muscles. Right scapula similar to left. Exostoses on eleventh and twelfth left ribs and several on left of lumbar spine in substance of erector spinæ. Spinal column rigid. Arms are held off trunk in abduction, cannot be brought to side of trunk. Left humerus only movable with aid of scapula, over shaft a bony mass size of pigeon egg. Motion of right arm also limited. Bony masses in anterior and post. axillary folds prevent motion. Entire absence of thoracic respiratory movement. Breathing abdominal muscles free. Left thigh 1.75 cc. larger than right. Fingers are all

\* Fortschr. auf d. Geb. d. Röntgen Strahlen, bd. 14, H. 3.

## MYOSITIS OSSIFICANS PROGRESSIVA

shorter than normal; little finger particularly so. Hallux valgus and microdactylia of both big toes. The first metatarsal bones short and stubby and interphalangeal joints of big toes ankylosed.

CASE XCI.—Reported by Charles Adair Dighton, in 1908. Male, aged 11 years. Family history neg. First symptoms appeared between 4 and 5 years of age—following successive attacks of measles, bronchitis and whooping cough; complained of general malaise and weakness—most marked in legs; child unable to walk. Subsequently muscles became stiff and hard, lumps formed in them, increasing steadily in size and numbers up to time of observation. Weak and undergrown. Skin very tender to touch. Walks on balls of toes. Lost almost all movements of shoulders and elbows. Both arms show large bony nodules in biceps, triceps, coracobrachialis and anconæus. The biceps are almost completely converted into bone. Left arm shows greater development of osseous masses. Forearm free, but muscles atrophied. In both legs adductors completely ossified. Extensors and flexors less affected. Left leg more marked than right. In the line of fibres of obliquus abd. ext. on either side where they run toward the pubic bone is an osseous plate the size of a man's hand. No mention made of microdactylia. Doctor Dighton kindly sent me his notes of the case, but no mention was made therein of presence or absence of microdactylia.

CASE XCII.—Reported by Charles F. Painter and John D. Clarke. Male, aged 25 years. Family history neg. First symptom appeared at 6 years of age—torticollis; a symptom pronounced as hereditary in his family. Numerous nodules developed all over the body in various parts and different articulations of spine and limbs became ankylosed. The right hip was mobilized operatively by removal of bone so that the joint could be freely moved after operation, but bony masses soon formed again. An open incision tenotomy of internal and external hamstrings was equally unsuccessful. A hard mass on right side of chest. A mass the size of a lemon situated under the left glenoid cavity. Motion of elbow limited. Numerous nodes can be felt over arms, wrists and fingers near joints. On both sides of the spine over the fourth or fifth ribs are pronounced nodes, and similar ones occupy the right side of spine opposite sixth to eighth ribs. Slight motion in atlanto-axoidean articulation, restricted by nodes. No thoracic respiratory motion. Dorsal and lumbar spine completely ankylosed. Left hip flexed about 40 degrees and adducted, with practically no motion. Nodes on left tibia on the upper epiphysis, limiting both extension and flexion of knee. Left ankle ankylosed, with nodes on dorsum and planta of metatarsus and phalanges. The right quadriceps muscle and the hamstring tendons undergoing ossification along their entire length. Nodes on tibia similar to left side. Right foot similar to left. The second toe of the left and second and third toes of the right foot are mentioned as being enlarged. (The X-ray picture of the left foot shows microdactylia of big toe through smallness of metatarsal bone.) This and a similar condition in the right foot may explain the elongation (?) of second and third toes.

CASE XCIII.—Reported by G. Rizzuto. Female, aged 8 years. First symptoms appeared at 4 years of age—in greater part of muscles of neck and nape of neck. Later attacked sacrolumbar regions. Osseous hardenings in neck, lumbar region, scapula, thorax and arm. Later affected the roots of right last ribs over the corresponding iliopsoas. Made hæmatological observations. (Only this brief, incomplete history as an account of the demonstration in the official report of the congress.)

CASE XCIV.—Reported by H. B. Allen, in 1909. Male, aged 27 years. Family history neg. First symptoms appeared in early youth (?). Subsequent course of disease unknown. Right temporal muscle partly ossified. Plates of bone along main tendon of erector spinæ in dorsolumbar region and spreading in direction of supraspinous ligaments, serrati post. inferior, and from lower ribs to inferior angle of scapulæ, uniting them both and sending off superficial process into ossified liga-

mentum nuchæ. Left arm: thickening of bone at inner bicipital ridge, at posterior edge of ulna and around wrist. Right arm: thick buttress of bone in pectoralis major from clavicle to outer bicipital ridge. Beginning ossification in capsule of shoulder. Osseous plates along shaft of humerus and process from upper part of olecranon. Osseous hypertrophies at upper and lower ends of forearm. Left leg: thick buttress bone from ischium to femur, in line with quadrat. fem. Some osseous hypertrophies near insertion of glutæus maximus and upper part of adductor longus; osseous growth with knee-joint, causing ankylosis, also binding the heads of tibia and fibula; similar growth at lower ends of bones. Right leg: bony ankylosis of hip-joint, with outgrowths from trochanter reaching tuberos. ischii, and others from both trochanters to front of os pubis. Lower ends of bone similar condition as left. Only photographs and skeleton available. Microdactylia of both thumbs and big toes and ankylosis between some carpal and interphalangeal bones; ankylosed and deformed second and third toe-phalanges.

CASE XCV.—Reported by L. R. Krever, in 1910. Female, aged 28 years. Family history neg. First symptoms appeared in twentieth year—very painful swellings on left shoulder, with light chilblains. Subsequent course of disease unknown. Ossification of pectorales, deltoid, supra- and infra-spinati muscles, trapezii, latiss. dorsi, the lumbar muscles, fascia lumbodorsalis. No microdactylia mentioned.

CASE XCVI.—Reported by K. F. Person, in 1910. Male, aged 18 years. Family history neg. First symptoms appeared at 5 years of age—soft, painful tumors on sides of neck. Tumors disappeared later, only to reappear in back of neck. At 13 years of age ossification of posterior muscles of neck, strongly involving ligam. nuchæ, spinal and lumbar muscles, and erectors of left thigh. Head slightly inclined backward and completely immovable. Spinal column has slight scoliosis, is completely immovable in cervical and dorsal portions. Slightly movable in lumbar portion. Exostoses and hyperostoses on scapula, vertebræ and head of left fibula; sharp atrophy of muscles of back, scapula, pelvis, which have not become ossified. Limited movements of both shoulder-joints, especially right; of both knees, especially left; of both ankle-joints and of toes. Scapulæ and ribs completely immovable; respiration abdominal. Microdactylia of both big toes, with aplasia of phalanges; bilateral hallux valgus and bilateral partial syndactylism of second and third toes.

CASE XCVII.—Reported by G. A. Pirie, in 1910. Male, aged 6 years. Family history neg. First symptoms appeared at 2 years of age—could not turn his head freely; had difficulty in moving it upward. Thickening and induration of ligamentum nuchæ. Hardness gradually increased up and downward, and when child was 3 years old the new tissue felt as hard as bone and had extended and adhered to occiput. This osseous growth was removed, and thereby the absence of spines and laminæ of third and fourth cervical vertebræ and defect in the closure of the spinal canal revealed; the spinal cord was exposed. A few months after operation difficulty in raising his arms developed, and examination showed swellings and indurations in latiss. dorsi muscles along both sides of vertebral column. They gradually increased and spread until nearly all the muscles, tendons and ligamentous tissues over back of thorax have become ossified. Osseous formation on back of neck has been formed afresh and is adherent to vertebræ lower down. A similar one is forming on left side. On right side osseous band extends from lower rib to crest of ilium. A few bony nodules at inner sides of both tibiæ and slight induration of adjacent muscles. Microdactylia of both thumbs.

CASE XCVIII.—Reported by Alves de Faria, in 1910, and Jorge de Toledo Dods-worth, in 1912. Sex unknown. First symptoms and age of their appearance unknown. Subsequent course of disease unknown. Entire report based on two radiographs. 1. Taken in left profile: osseous changes in superficial muscles of neck, after severe atrophic changes. Osseous bridge from occipital protuberance to dorsal muscles.

## MYOSITIS OSSIFICANS PROGRESSIVA

Between these and vertebral column some indefinite shadows, denoting second stage of invasion. 2. Anterior view of thorax: from seventh left intercostal space running toward humerus, but not reaching it, was a cross-formed osseous process. On right side a similar bridge joins the bone. From sixth rib a bony elongation runs to the outside and ends about middle of preceding one. The two are connected by a third one. Humerus thickened in upper part; numerous osseous nodules on back. Microdactylia not mentioned. No X-ray picture of hands or feet.

CASE XCIX.—Reported by P. J. Stoyanoff, in 1912. Male, aged 25 years. Family history neg. First symptoms appeared at 10 years of age—when patient had variola. Subsequent course of disease unknown. Signs of former variola all over body. Insertion genioglossus muscles ossified. Neck rigid and moves with great difficulty. Trapezii ossified and joined to scapulæ, which stands out wing-like. Scaleni ossified. The clavicles are bent, but muscles of the supra- and infra-spinous fossæ being ossified. Multiple exostoses; axillæ ossified. Large portion of pectorales ossified. Insertion of deltoids ossified, as also that of left triceps humeri. Parts of biceps and coracobrachialis ossified. Left elbow ankylosed at 180 degrees. Movable 15 to 20 degrees. Spine ossified; vertebræ immovable. Scoliosis toward left 150-160 degrees, in cervical region lordosis almost 90 degrees. Musculature of back ossified and united to scapulæ. Gluteal insertion ossified. Left trochanter major has exostoses 7 cm. in diameter. Adductores femoris ossified and aponeurosis hard. Tensor fasciæ hard. Vastus intern. on condylus intern. ossified. Right knee almost ankylosed at 90 degrees. Exostoses in right popliteal region and in pes anserinus 4 cm. long. Triceps suræ hard as though ossified. No microdactylia mentioned.

CASE C.—Reported by Author R. Elliot, in 1911. Female, aged 17 years. Family history neg. First symptoms appeared shortly after second year—lumps appeared on her head and disappeared again; no apparent cause, no injury. In the succeeding three years more nodules formed. At 5 years torticollis appeared and remained for about six months. Later painful swellings on arms and legs interfered greatly with motion. Since that time patient has never been able to abduct her arms or raise them. A valvular heart lesion was then discovered; no previous acute infection or rheumatism. January, 1907, typhoid; July, 1907, stiffness of jaw; teeth could be separated but slightly; hard bony mass fills submaxillary region. Hard circumscribed swellings free from ribs on both sides of thorax. October, 1907, both arms became stiff and partial fibrous ankylosis developed in right elbow-joint. Left shoulder and arm followed. Head bent forward and downward; stiff. Maxillary movements much restricted. Teeth separable only about one inch. Lower jaw displaced to right side. Anterior and posterior neck muscles firmly indurated. Mastoid part of sternocleido of bony hardness; clavicular origin indurated. Pectoral muscles hard and contracted. Hard nodule on upper border of left scapula. Small exostoses on right eighth and eleventh ribs; also ninth left; also on posterior crest of iliac bone. Body bent forward; rotation of trunk impossible. Spine firmly rigid. On either side of lower dorsal and lumbar spine a broad bony mass, over right inguinal region a firm induration size of a man's palm. Abdominal muscles tender to pressure. Muscles of legs free, but patient states that they feel too short. Both thumbs show bony ankylosis in terminal phalangeal joints. Second joint pliable, but atrophic. Thenar flat and atrophied. Never had voluntary movements of thumbs. They are held flexed on palms (pollex valgus). Microdactylia of both big toes; absence of one phalanx (?). Toes directed outward, lying partly under second toe (hallux valgus). No X-ray of toes. Cause of short-hallux probably erroneous, and shortened metatarsus primus; here the usual explanation—X-ray of hands shows shortened metacarpus primus.

CASE CI.—Reported by Jgn. Peteri and Gust. Singer. Male, aged 4 years. Family history neg. First symptoms appeared at 1½ years of age—swellings on right and left sides of forehead. They disappeared again after a few days. Other

## JULIUS ROSENSTIRN

swellings appeared, which grew harder and finally became as hard as bone. 1906: head bent forward and fixed rigidly, only slight lateral motion. A swelling size of a child's fist developed in submental region; during his hospital stay other inflammatory symptoms. With relief of these symptoms a solid mass remained, impeding movement of lower jaw. Ridges of bony substance in temporal and masseter muscles. Muscular part of nape forms a diffuse, solid bony mass with nodulated surface, causing rigid stiffness of neck. Slender bony ridges in both sternocleidii bound to broader ones in deeper cervical muscles. Pectorales major and minor changed to hard osseous plates. The upper arms are firmly united to shoulder-blades and ribs by broad hard bridges, in anterior and posterior axillary folds. Both biceps and brachialis ant. contain broad bony ridges along entire length. While the right one is partially fixed to humerus, the left runs free from bone to tuberosit. ulnæ. Both scap. fixed to back. Musc. of forearm tough and fibrous. Entire vert. column rigid. The processi spinosi form, together with muscles and ligaments, thick, heavy, protuberating masses of bone tissue. Four years later: patient again admitted. Stiffness and immobility of entire upper part of body. Angular conformation of shoulder girdle and various relief-like elevations throughout body had increased, especially those of obliquus intern. Microdactylia of both thumbs and big toes. (Synostosis between short first metatarsal bone and first phalanx.)

CASE CII.—Reported by Felix Bauer, in 1911. Aged 2½ years. Sex not mentioned or alluded to in entire paper. Family history neg. Scarlet fever when 6 months old (?). First symptom noticed was ossification of neck after scarlet fever. Ossification of neck and shoulder progressed and caused decrease of movability of head and arms. Strained, bent-forward posture of head and upper part of body. Upper extremities fixed. Spinal column stiff. Sternocleidii and the deep neck muscles, the shoulder muscles and both bicipites are ossified and intersected with projecting lumps and spurs. They are mostly fixed to the underlying bones; only those of the biceps are free and movable in the muscles. From costal arch in anterior axillary line extends a round partly fibrous and partly osseous ridge size of penholder toward the symphysis. It continues upward toward the fifth rib on both sides. Over the shoulder-blades and at sides of spine several immovable, semiglobular, osseous tumors. Microdactylia of both big toes; in a lesser degree of both thumbs and little fingers.

CASE CIII.—Reported by Otto Jüngling. Male, aged 6 years. Family history neg. First symptoms appeared before end of first year—lumps developed in nape of neck. At first lumps disappeared again without leaving any traces, but after some time they recurred in the same place, remained and grew hard. Gradually this process spread from the neck to the back and arms, and for some time the upper part of the boy's body has been stiff. Head, neck, shoulders and trunk quite stiff. Mouth can only be opened 1¾ cm. In the neck only the vert. prominence can be felt; from it a bony ridge 4 cm. long, thickness of a small finger, starts downward, divides into two and at level of scapula turns up again. At about the ninth vertebræ another ridge starts obliquely to the angle of the left scapula. On the right side only two lumps. The lumbar portion of the spine shows slight scoliosis convexity to right. The sacrospinalis on the left is stone-hard in its entire length. A small bone plate in the height of the fifth lumbar vertebra. The upper arms are fixed to the thorax in an angle of 35 degrees. From the scapula angles hard ridges run to both humeri. In both triceps are hard bony masses and two stalactites in the right elbow head. Both thumbs very small, due to smallness of first metacarpus. The first and second phalanges of second and third fingers very small and ankylotic. Shortness of right first, third and fourth and left first, fourth and fifth metatarsal bones. The left first, fourth and fifth metatarsal bones show only minute epiphysial nuclei (distal) and the phalanges of all the toes are deficient. The first metacarpal metatarsal bones show

## MYOSITIS OSSIFICANS PROGRESSIVA

only proximal, the other second, third and fifth only distal epiphyses. Hypospadias-urethral orifice at scrotal insertion.

CASE (?).—Reported by Riehl, in 1912. Male, age (?). No family history. First symptoms appeared since 6 years of age—febrile attacks of pain in joints, with redness and swelling of skin. Later lime deposits in muscles and subcutaneous tissue. Skin difficult to fold; thickened in places, in others atrophic. Very little fat in subcutaneous tissue. Irregular and indistinct cloudy or striped shadows and very little pronounced ossification in X-ray picture. Author speaks himself of similarity of his description of this case with severe scleroderma. (Not myositis ossif. progress.) No mention of microdactylia.

CASE CIV.—Reported by C. A. Manjapa, in 1912. Male, aged 25 years. Family history neg. Feverish illness at 6 or 7 years of age. First symptoms appeared at 6 or 7 years of age—had fever for nearly six months, and about that time lumps were discovered on back of neck and chest. Grew gradually unable to stand erect or walk freely and became mentally deficient. His disease, on account of lumps, was erroneously diagnosed as leprosy and he was transferred to the leper asylum; later, in September, 1907, on account of his mental condition, to the lunatic asylum. Patient can only sit and stand in a crooked position. Head fixed, with deflection to left side; incomplete lockjaw, due to stiffness of masticating muscles. Neck stiff and almost all voluntary muscles in slight or greater degree of ossification. Trapezius and erectors spinæ present, bony ridges extending from suboccipital to lumbar region and bony tumors at spine and inferior angle of right scapula. Left pectoralis major hard; bony ridges along sternal fibres, connecting with one from their humeral insertion. Right deltoid also shows bony ridges. Right forearm fixed at right angles to arm by bony buttresses. Left forearm fairly movable, but not to full extension. Right supinator longus entirely ossified and bony swellings in flexors and extensors of left forearm. Left thigh has movable bony plates in region of extensor quadriceps, adductor longus and hamstring muscles. Had short fingers and short toes. About eight months prior to his death (July, 1910), from dysentery, patient began to lose flesh and strength. The joints became more firmly fixed and the osseous growths very prominent and distinct.

CASE CV.—Reported by Fritz Magyar, in 1912. Female, aged 3 years and 9 months. Family history neg. First symptoms appeared at 3 years—hindrance in motion of muscles of upper arm and pains on pressure. Pains disappeared, but hindrance remained and muscles grew stone-hard. Head bent forward. Ossification of both bicipites brachii; on back also ossifications in serrated form adhering to underlying tissues. The movements of arms were interfered with by hardening of upper and lower margins of latiss. dorsi posteriorly and pectorales major anteriorly. No mention made of microdactylia. (Brief notes only in Wien. mediz. Wochenschr.) To personal letter received no reply.

CASE CVI.—Reported by David Rankin, in 1912. Female; a little negro girl. Subsequent course of disease unknown. Casual observation of author during visit to a hospital in Brazil, where he saw a little black girl suffering from myositis ossificans progressiva, with trapezius, latiss. dorsi and greater portion of both pectorales ossified. No mention made of microdactylia.

CASE CVII.—Reported by Joseph Frattin. Female, aged 14 years. Family history neg. First symptoms appeared at 1½ years of age—painful swelling at nape of neck; disappeared again within twenty days without leaving a trace. At 4 years painful swelling again developed in same place and disappeared again, but neck remained stiff; later back and hip, which likewise showed exceedingly hard tumors. She suffered from epilepsy at an early age. The attacks grew rarer, and two months after leaving hospital disappeared entirely. Head slightly inclined toward right; can be turned, but not freely moved from side to side. Hard nodules on both sides of glabella, some adherent to bone. Muscles on both sides of nape of neck have bony-

hard nuclei at occipital insertion, the right sternocleido at mastoid and a bony thickening at clavicular. Lower insertion of scalmi ossified. Upper margin of cucullaris and upper part of left sternocleido ossified. Tendon changed to osseous band. Both arms fixed in adduction. In right biceps and triceps, pectorales, latiss. dorsi, many bony plates; on left side tendons of these muscles ossified; on lower epiphyses of humerus irregular bony mass encircling joint completely. Breathing abdominal. Broad muscles of back completely ossified, left more so than right. Infraspinalis and sacrolumbalis show osseous plates movable in muscular tissue. Near sacroiliac symphysis are osseous tumors; both hips stiffened; soft parts surrounding coxo-femoral joints show many ossifications. At lower part of glutæus major large osseous band and an osteoma on inner side of crista ilii. Similar conditions on left leg. Head, slightly inclined toward right, can be turned but not freely moved from side to side. Both big toes show microdactylia. Left smaller than right. X-ray picture not quite clear.

CASE CVIII.—Reported by S. Goto, in 1912. Male. Family history neg. First symptoms appeared at 1 year and 10 months of age—swelling and hardening in jugular fossa. Later an osseous formation developed in the jugular swelling. A year afterward swellings on occiput and various other parts of the head, which disappeared again. Since January, 1911, parents have noticed stiff posture of body. Weight, 14.525 kils.; height, 93.0 cm. Movements of head very limited. From posterior surface of mandibula comes a hard osseous band to posterior surface of chin. Small tumors at sternal origin of sternocleido and at insertion of trapezius. Both scapulæ fixed to thorax and their lower angles are united in one solid mass with surrounding ossified tissues. On both sides of sixth to eighth dorsal vertebræ bony growths size of thumb. Along both sides of spine broad, symmetrical, flat, bony growths 3 cm. broad, united below to crista ilii, above to masses at sides of vertebræ. The ant. and post. axillary folds are ossified. Arms only very slightly movable in shoulder-joints, free in elbow and other joints. A string-like band in right side of abdomen and a small, hard, osseous thickening on inner condyles of femur and tibia. Both thumbs and big toes abnormally small, owing to smallness of metacarpal and metatarsal bones. (Hallux valgus.) Electrical sensibility; decrease to absence of irritability to faradic or galvanic current in affected muscles. Those apparently free from affection show no disturbance.

CASE CIX.—Reported by Rudolph Jacobi. Male, aged 6 years. Family history neg. First symptoms appeared at 3 years of age—hard nodules formed on back. Nodules were rather flat at first, but grew gradually to their present size. For the last five weeks there has been a disturbance of the free motion of his right arm. Movements of head not free; chin cannot be brought to thorax. Scattered over entire back are numerous osseous protuberances of semiglobulous and flat form, averaging pigeon-egg size; also long ridge or beam-like elevations of about 15 cm. length and thickness of thumb diverging from median line to lateral parts of thorax. Nearly all the muscles of the back are changed into a hard osseous mass, encasing the ribs armor-like and inseparably connected with them. Respiration abdominal; thorax shows only minimal excursion. Arms and legs show no change except exostoses on both condyli interni tibiæ, 2 cm. long. Microdactylia of thumbs and big toes. (Hallux valgus.)

CASE CX.—Reported by S. J. Khaikis, in 1913. Male, aged 4 years. Family history neg. First symptoms appeared when about 3 years of age—old swellings appeared on occiput. Later swellings showed on back and lower extremities (treated by local physicians with inunctions); they would subside, then reappear. These tumors began to appear first on the occipital musculature, then on that of the back and upper ribs. The swellings along the ribs were quickly absorbed, but the others hardened and had the consistency of bone. Patient only complained when new swellings appeared. Boy normally developed. Pain when bent over, but head can be bent



## MYOSITIS OSSIFICANS PROGRESSIVA

forcibly in the different directions without pain. Occipital movements interfered with by tubercles on occipital bone about attachment of cucullaris. Similar tubercles on both sides of spine along lower third dorsal and first lumbar vertebræ. Other nodules appear over middle of upper ribs; also in axillæ, interfering with raising of arms. During observation of one month methodical treatment with potassium iodide, baths and fibrolysin. Condition grew worse, redness and swelling to left side of second lumbar vertebræ. Redness disappeared again, but swelling grew osseous. No mention of microdactylia.

CASE CXI.—Reported by Volhynia Medical Soc., 1913. Female (?). Subsequent course of disease unknown. The foregoing case was before his discharge shown in the Volhynia Medical Society. A similar case of a girl was demonstrated there also. Demonstrator not mentioned in report.

CASE CXII.—Reported by Erich Blenkle, in 1914. Male, aged 21 years. Family history neg. Not much known about early history; lost parents very early in life. Subsequent course of disease unknown. Stature of boy 10 to 12 years; height, 137 cm.; panniculus underdeveloped. All the limbs except the right leg and hand stiff; cannot sit down, has to do his work, eat, etc., standing. Impossible to mount stairs. Jaws cannot be opened more than  $\frac{1}{2}$  cm. Teeth (incisors) have been removed for feeding. Masseters hard, with hard osseous nodules imbedded. Head only movable sideways in very slight degree. Spine completely stiffened. Sterno-cleido and neighboring muscles of neck, entire musculature of back stiff and hard and interspersed with small osseous nodules. Along entire dorsal spine large osseous plates; below tenth and eleventh vertebræ no muscles to be felt; only one continuous osseous plate, extending to crista ossis ilii. Thorax and ribs immovable; breathing entirely abdominal. Pectorales and deltoids stiff and hard. Both arms fixed, left less so. Osseous imbeddings in biceps and triceps and near origin of extensor carpi radialis. Slight volar flexion possible in left wrist. Both hip-joints immovable; hard masses surround both trochanters; knee-joints slightly movable; osseous plates in left adductor. Exostoses of femur; hard mass around left knee muscles of right leg, around hip and knee-joint hard and stiff. Both thumbs small (microdactylia). Third phalanges of fourth and fifth fingers turned toward radial side. No microdactylia in either foot.

CASE CXIII.—Reported by F. Parkes Weber and Alwyn Compton, in 1915. Female, aged  $2\frac{1}{2}$  years. Family history neg. First symptoms appeared at  $7\frac{1}{2}$  months of age (March, 1913)—was shown before the Royal Society of Medicine, with bony projection from left side of neck and thin spicula of bone attached to middle of the back of left clavicle. In March, 1914, various diffuse swellings appeared, involving muscles and superficial fasciæ; skin could be freely moved over them. They disappeared and soon were followed by others in left scapular region and left lower part of thorax. Similar ones appeared also on right side. In May, 1914, a distinct bony formation appeared in left posterior axillary fold, and transient swelling in right pect. maj. In June hard swellings in right biceps and lower part right scapula and corresponding ones on left side. In September and October a bony lump on occipital insertion of trapezius. Very stiff in movements of back and neck, extension grows more and more limited. There also seems to be osseous infiltration in both anterior and posterior axillary folds, limiting abduction of both arms. Microdactylia of both thumbs and big toes; in the thumbs due to smallness of the first metacarpal bones; in the toes to a synostosis and resultant smallness of phalangeal bones. (Hallux valgus.) A small piece of right latiss. dorsi examined microscopically showed invasion of muscle by newly formed fibrocellular connective tissue.

CASE CXIV.—Reported by Angel M. Centeno. Male, aged  $8\frac{1}{2}$  years. Family history neg. At  $3\frac{1}{2}$  years phlegmonous (?) inflammation of left sternocleido. First symptoms appeared at 6 years of age—entered Children's Hospital. Smooth, hard, painless tumors in region of right scapula. (Caught measles in hospital, but recovered

## JULIUS ROSENSTIRN

without complications.) When seven months in hospital first operation was performed on sternocleido tumors. Result not known to padres. Later new, round, hard, painless tumors of various sizes appeared over the entire region of the dorsalis major, the trapezius and the costal walls. Shortly after the first operation a second one was performed, and a little later a third one, for the avowed purpose of removing little pieces of new-formed bones. Wassermann positive. (Author seems to attach much etiological importance to this reaction; in other such cases reaction was negative.) Face looks as if patient has adenoids and prognathism of left maxillary bone. Head inclined forward and downward. The physiological lordosis has disappeared. Thorax slightly inclined forward; arms fixed in the same direction as upper part of thorax, separated from it and in attitude to grasp an object. Osseous tumors in dorsalis longus, trapezius and both sacrolumbales; in fact, entire posterior wall of trunk. In upper movement of left trapezius a perfectly movable, painless, small, hard nodule. Lower extremities slightly flexed, left forming an obtuse angle with thighs. Double hallux valgus turned outward and metatarsophalangeal at nearly right angles with first metatarsal bone, covering with the lateral edge of the phalanges the second metatarsophalangeal articulation. There is a disturbance in the development of the two upper and inner incisors. They show an excrescence, which descends to the point of an implantation from the inner side to the middle of the teeth. The two rows of teeth do not meet together, and in the hard palate is a deep oval impression. The voluntary motions are limited by the described lesions. Left arm passes with difficulty just the horizontal line; right one appears as if glued to the side, its angle of motion not more than 30 degrees. Hands show thinness of little fingers and shortness of thumbs at expense of last phalanx (?). No X-ray of either hands or feet. Both testicles are ectopic, having remained in the inguinal canal near the external opening. Nothing else of note.

CASE CXV.—Reported by Seth Hirsch and Joseph Roth, in 1917. Male. Family history neg. First symptoms appeared at 2 years of age—lump on right side of neck. Lumps on neck disappeared. Other lumps developed below right scapula. Said in orthopedic hospital he had suffered from tuberculosis, applied a brace for nine months. It was then removed. Incision was made upon lump below right scapula. During last year the shoulders were becoming progressively displaced forward. Tonsils and adenoids were removed a year ago. Slimly built; walks about, but is constrained in movements of back and arms; shoulders very narrow, as though cramped forward. Calvarium shows at right parietal region large soft tumor about two inches in diameter, seemingly set into the bone. Both scapulæ are tilted forward and seem to be fixed in relation to the ribs. A bony protuberance below right scapula. Some pain at movements of shoulder beyond difficult limitations. Very rigid spine throughout. (Pokerback.) Normal genitals. Shortenings of both thumbs and big toes. X-ray shows below the angle of the left scapula a large, rounded bony mass, and extending upward and inward a strip of bone located in the teres major. At the right side a large bony mass posteriorly behind the tenth and eleventh ribs, extending upward and outward into a rib-shaped prolongation, bounding the inferior axillary space and divides near the humerus into two small branches. This entire ossification is in the latissimus dorsi. In the right lumbar region opposite third and fourth lumbar vertebræ is an irregular stellate ossific shadow, and extending upward a linear mass merging with the body of the twelfth dorsal. On the left side runs an ossification strand from the lower ribs toward the iliac crest. Knees show a spur-like, bony formation from the posterior surface of the lower and of the left femur diaphysis. Also small ossification strand above its head. Thumbs and big toes show brachydactylia.

CASE CXVI.—Reported by Seth Hirsch and Joseph Roth, in April, 1917. Family history neg. First symptoms appeared at 6 months of age—small nodule on forehead, increasing in size to that of a plum, then remained stationary. Well after lump on

## MYOSITIS OSSIFICANS PROGRESSIVA

forehead till age of 8 years, when it was noticed that he could only raise the right arm with great difficulty. Soon after the left became similarly affected. Patient was removed to post-graduate hospital, where incisions were made over both scapulæ and pieces of a bony growth removed. Child got scarlet fever; returned home. Conditions progressed slowly, involving neck and rest of the spine. Later the lower extremities became also involved; boy still being able to walk with some difficulty. The disease shows signs of progression. Patient very thin, mentally active, able to walk with difficulty. Spine rigid and kyphotic, neck rigid, arms are adducted; he is unable to move them either outward or upward. Scars over both scapulæ from which portions of the osseous growth have been removed. There is a bony growth about the left frontal bone. The posterior muscles of the neck appear hard to the touch. The scapulæ are markedly winged. The upper extremities are fixed to the axillary border of the thorax. There are hard bony masses running from the serratus magnus on each side to the humerus. Posterior spinal muscles appear hard; there is marked rigidity of entire spine. There is marked contraction of the adductors and the *tensores fasciæ latæ*. The thighs are flexed on the pelvis, fixed and adducted; the legs are flexed on the thighs. Hard bony masses in the adductors and muscles of the calves. The genitals are normal. The big toes are considerably shortened. X-ray examination. Numerous strands of ossification in both *latissimi dorsi* and in many of the intercostal muscles of both sides. Examination of the pelvis shows numerous branch-like strands of ossification extending into the left gluteal group. A large spur-like mass, of bone between the lesser trochanter and the right ischium. The knee shows a long spicule of bone springing from the posterior surface of the femur. Both big toes show brachydactylphalangia.

CASE CXVII.—Reported by Eug. L. Opie, in May, 1917. Male, aged 70 years. Nothing mentioned of previous history, description of skeleton only given. Subsequent course of disease unknown. Left internal pterygoid muscle ossified. The anterior edges and articular surfaces of the cervical and lumbar vertebræ show bony projections. Head and neck of seventh and eighth ribs ankylosed by bony union with the corresponding vertebræ. Broad bands about 2 cm. across connect on the right side of the fifth, sixth and seventh ribs about 2.5 cm. from their vertebral origin; the fourth and fifth ribs are similarly united on the left side. The bones of the shoulders and upper extremities show prominent ridges for the insertion of the muscles. No deformity of hand. Pelvis shows similar prominences and projections also on edges of acetabulum. Both femora show irregular ridge of bone 1 cm. high on the *linea aspera* just below the middle of shaft. Tibiæ and fibulæ very irregular in outlines. Rough areas with spiculæ project upward in the direction of the muscles. Bones are considerably thickened by periostitis; membranæ interossei are ossified at irregular places, forming ridges of bone. Right foot: porous texture of bones. Distal end of first metatarsal is enlarged and irregular in contour. Projecting downward and backward from the head of bone is irregular osteophyte 1 cm. long. Small bony projection, 3 cm. long and ankylosed, at the upper inner surface of the articular surface representing the two terminal phalanges of the great toe. These bones do not extend beyond the first interphalangeal point of second toe. Left foot: shows some porous texture of bones. Head of first metatarsal bone absent; end of bone broadened. Appearance of metatarsal bone as if its head, together with big toe, has been amputated. Microdactylia of both big toes (?).

CASE CXVIII.—Reported by Ten. Bokkel Hunink, in June, 1917. Female, aged 7 years. Family history neg. First symptoms appeared at 2½ years of age—swelling under chin. Swelling under chin disappeared; was followed by one in neck, causing stiffness. Had hard nodules on forehead, which also disappeared later. Mouth can only be opened ¾ cm. Flexion in elbow-joint to 90 degrees, extension to 120 degrees. Left shoulder hardly movable. Along the upper ribs many bone-hard swellings. The lower third of the left m. biceps feels like a bone-hard mass. The left axilla is entirely enclosed with bone-hard plates; no muscle to be felt. The muscles of the

## JULIUS ROSENSTIRN

neck, with the exception of the sternocleidomastoidei, are changed to a bone-hard mass. Large bone-hard plates in the long muscles of the back; also in the left latissimus dorsi. The medial edge of the right scapula merges into a bone-hard mass, which is fixed to the boneplates in the long extensor muscles of the back. Returned later with about the same status, only that the mouth can be opened wider and the muscles of the neck, which felt like a bone-hard mass at the first examination, now have grown softer, and though not as soft as ordinary muscles, no bone-hardness can be felt in them, and the movability of the head is accordingly improved. X-ray plates not shown in this publication, but author mentions that only in the chest and back the palpatory findings are confirmed by the X-ray, while the neck muscles show no ossification and the biceps show only a very small piece of bone in the muscular tissue. Bilateral microdactylia of both big and little toes; big toes have large first metatarsus and only one very small phalanx. (Synostoses?) Little toes very small phalanges.

CASE CXIX.—Reported by C. Johannesen, in 1917. Female, aged 2 years and 8 months. Family history neg. First symptoms appeared at 1 year and 8 months of age—swelling over skull and occiput. (Protub. occip.) Swellings were not sensitive. After four months later swelling over left shoulder, spreading to left scapular region and left side of neck to ear. It traveled in four to five days under left arm and across back. Not sensitive; no discoloration of skin. Arms could hardly be moved in shoulder-joints. After a week swellings began to disappear; instead a swelling appeared in left sternocleidomastoideus. After the left-sided swellings had disappeared swellings showed in right scapular region under right arm and lumbar region. About two weeks ago patient became sick; had chills; took to her bed and has lost flesh. Patient well developed. Height 88.5 cm., weight 13.8 kg. Head circumference 52, chest 54 cm.; anæmic; purulent discharge from both ears; nasal discharge and cough; chest and abdomen normal; temp. 37.2; pulse 92, regular; sits rigid; bends head forward and over to right side, with face toward left; opens mouth wide and masticates and swallows without difficulty; right side of neck and right fossa supraclavicularis are swollen; swelling of lower half of sternocleido. The inspection of right side of back shows swellings and grooves like topographic chart. Swellings in the neck in fossa supraclavic. continue over right part of back down to eleventh rib. Swellings in muscles of trapezius, axillary folds and serratus ant. They reach middle axillary line from second rib down to eighth rib. Swellings are bone-hard and immovable over underlying tissue. Skin normal appearance. Right arm stiff; left can be moved freely. Wassermann and v. Pirquet neg. Erythrocytes, 6,000,000. Leucocytes, 125,000. Urine nothing abnormal. Microdactylia of both thumbs and big toes. Hallux valgus bilateralis. According to Roentgen plates the thumbs show a shortened first metacarpus and both big toes a synostosis of a shortened first metatarsus, and first phalanx with a separate small last phalanx outward, resulting in bilateral hallux valgus.

CASE CXX.—Author's own case, in 1916. Male, aged 20 years. First symptoms congenital. Subsequent course of disease reported elsewhere in this text. Status at time of observation reported elsewhere in this text. Congenital abnormalities reported elsewhere in this text.

(TO BE CONTINUED)