OSTEOGENIC TUMOURS OF THE JAWS

Charles Tomes Lecture delivered at the Royal College of Surgeons of England
on
15th July, 1949
by
Kurt H. Thoma, D.M.D., F.D.S.R.C.S.
Professor of Oral Surgery, Emeritus ; Charles A. Brackett, Professor of Oral Pathology, Harvard University

THESE TUMOURS ARE MADE up of tissue derived from osteogenic mesenchymal cells, which when fully differentiated are osteoblasts. So here again we may encounter benign and malignant neoplasms made up of cells, the development of which may have been arrested at various stages of their growth. Osteogenic tumours, therefore, may be made up of simple spindle cells, chondroblasts and osteoblasts. These tumours do not necessarily produce bone; they may form myxomatous tissue, collagen, osteoid, or cartilage. In the benign osteogenic tumours, all cells, as a rule, reach the same stage of differentiation; while in malignancy, because of rapid growth, two or more types of tissue may be seen simultaneously or at different periods of their existence.

On this basis, the following classification may be made:

Osteogenic Tumours

Osteoma :
- osteoma durum
- osteoma spongiosum

Chondroma :
- peripheral
- central

Osteogenic myxoma

Osteogenic fibroma :
- peripheral
- central

Fibrosarcoma :
- peripheral
- central

Fibro-osteoma :
- ossifying fibroma
- osteoid osteoma
- mature osteoma

Osteogenic sarcoma :
- osteolytic
- osteoblastic

OSTEOMA

This is a benign tumour, which generally occurs as an expansive formation growing from the periphery of the cortex of the jaw to which it is attached. The attachment may be in the form of a flat pedicle or occur over a wide base. Osteomata generally develop slowly and produce a gradually increasing tumefaction in the oral cavity or deformity of the face. We distinguish microscopically the osteoma durum with an eburnated, lobulated surface, made up completely of dense cortical bone and scant fibrous stroma, and the osteoma spongiosum in which cancellous bone predominates. The marrow substance in these is replaced by dense fibrous tissue.

Osteoma of the mandibular condyle has been reported, though it is classified by many (Rushton, 1944)[1] as a hyperplasia. A lobulated,

spherical unilateral enlargement of the condyle causes occlusal abnormalities, oblique attrition of the teeth, facial asymmetry (Fig. 12A), and dysfunction of the mandibular joint with limitation of motion. I have seen two such cases ; in each the histological examination showed normal bony structure with red marrow in the marrow spaces. Roentgen examination should be made from a postero-anterior projection as this gives the clearest picture of the enlargement that has occurred (Fig. 12B).

Osteomata are found also in multiple form. Fennel (1938)[2] described the case of a patient who had so many osteomata throughout his entire skeleton that he was colloquially known as " Knobby Willy." I took care of a patient who had numerous osteomata, principally in the facial skeleton (Thoma, 1936)[3] ; multiple at each side of the inferior border of the mandible, two at the right ascending ramus, one in the right maxillary sinus (Fig. 13), one attached to the mastoid process, and a small one forming in the apex of the orbit.

CHONDROMA

Although it is generally assumed that chondroma in membranous bones forms from cell rests of the embryonic cartilaginous skeleton, it is true that it may also form by differentiation of mesenchymal cells into chondroblasts. The tumour is extremely rare in benign form, though cartilage is frequently seen in osteogenic sarcoma. It consists of irregularly arranged, vacuolated cartilage cells producing hyalin intercellular substance, and sometimes osteoblasts which give rise to osteoid or bone.

Chondroma may form as a central tumour or as an outgrowth from the surface of the bone. A globular tumour of the latter type which was attached to the alveolar process in an edentulous area was reported by Dr. Francis McCarthy*. A maxillary chondroma causing great deformity of the face in a patient 52 years old was seen by Dr. C. C. Simmons†, and Dr. M. Jacobs (1942)[4] published a case of a chondroma of the mandible, which, however, may be classified as a fibro-osteoma (Fig. 18F).

MYXOMA

Myxoma and fibromyxoma are, as we have seen, generally of odontogenic origin. Differentiation is not easy, however, and there is a question in my mind whether cases of osteogenic myxoma can always be distinguished by either microscopic or roentgenologic investigation. As a clinical criterion, one may use the rather malignant behaviour of the osteogenic type, which resembles that encountered in other parts of the skeleton. The odontogenic type, as has been pointed out in the previous paper, being benign, recurs seldom if carefully enucleated. The osteogenic type I have encountered in a case confined to the mandibular condyle, far enough away from the region where odontogenic activity

* THOMA, K. H. (1949) *Oral and Dental Diagnosis*, 3rd edition, Philadelphia, W. B. Saunders Co., p. 448.

† THOMA, K. H. (1934) *Clinical Pathology of the Jaws*, Springfield, Ill., Charles C. Thomas, p. 457.

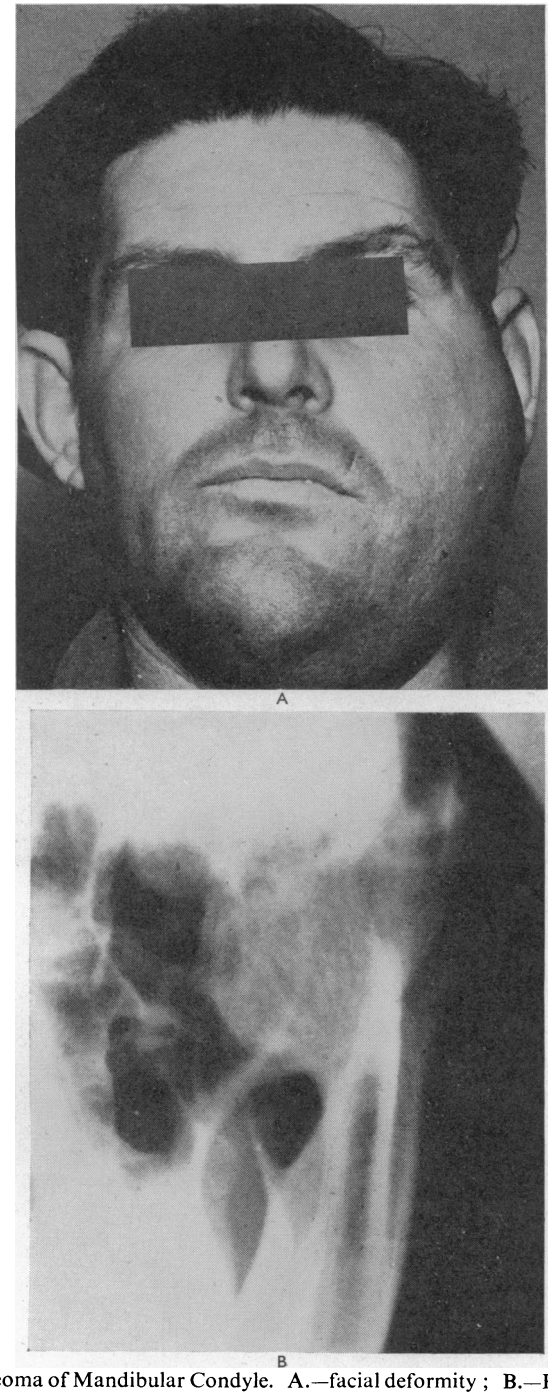

Fig. 12. Osteoma of Mandibular Condyle. A.—facial deformity ; B.—Roentgenogram.

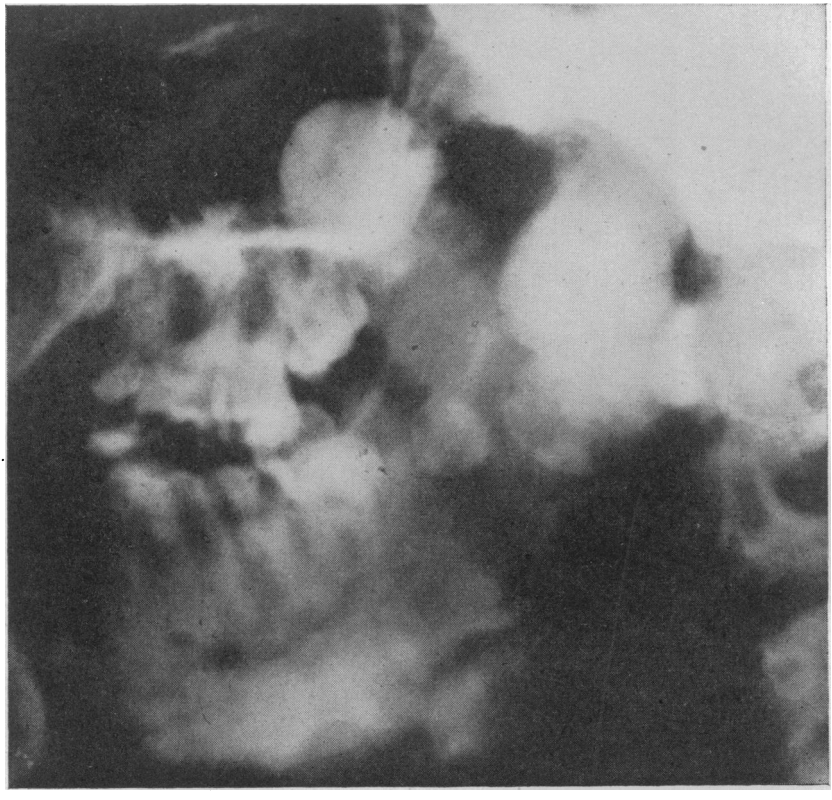

Fig. 13. Multiple Osteomata of Mandible and Maxilla.

occurs to classify it as an osteogenic myxoma (Fig. 14). Resection of the condyle in the healthy bone of the ramus completely cured the patient.

OSTEOGENIC FIBROMA

The osteogenic fibroma may occur as a peripheral and as a central tumour. It is made up of fibroblasts which deposit collagen fibres. These constitute the main part of the avascular tumour in which a few spicules of bone may form.

PERIPHERAL FIBROMA

Peripheral fibroma arises from the periosteum of the jaw and is generally situated on the alveolar process, where it is known as epulis fibromatosa. This tumour is well known and of frequent occurrence.

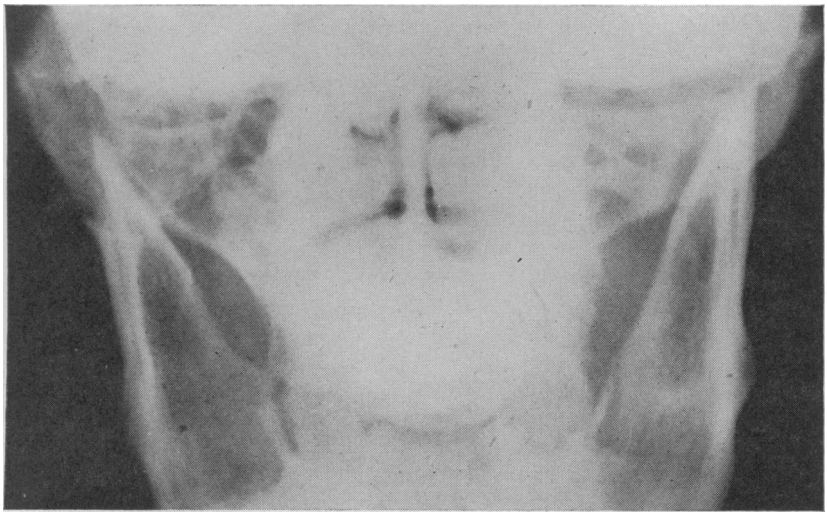

Fig. 14. Myxoma of Mandibular Condyle.

CENTRAL FIBROMA

Intraosseous fibromata are uncommon. They are said to arise in the maxilla from the periosteum of the maxillary sinus, causing a gradual enlargement of its walls by pressure. I have not seen this type. They may form in the mandible from the connective tissue of the perineural sheath of the inferior alveolar nerve (neurofibroma), but mostly they are formed from the dental follicle. These have already been described under odontogenic tumours. A central fibroma appears in the X-ray as a well-circumscribed cystic area. Because of a tendency to form osseous structures this tumour generally develops into an ossifying fibroma or fibro-osteoma, which will be described later.

FIBROSARCOMA

Again we encounter peripheral and central tumours. Those which occur on the oral aspect of the jaws tend to be less malignant than those which arise from the region covered by skin.

Peripheral fibrosarcoma varies in rapidity of growth as well as in malignancy. In the mouth, they form bulky nodular tumours (Fig. 15), sessile in character, bluish red in colour, often presenting an ulcerated surface when injured by the teeth in the opposite jaw. At the cutaneous part of the jaw, the tumour forms from the periosteum and generally grows expansively and with great rapidity. It soon invades and destroys the bone. It occurs at the angle of the jaw, the symphysis, and once in my experience, it arose from the condylar part of the mandible causing a swelling under the skin in the preauricular area.

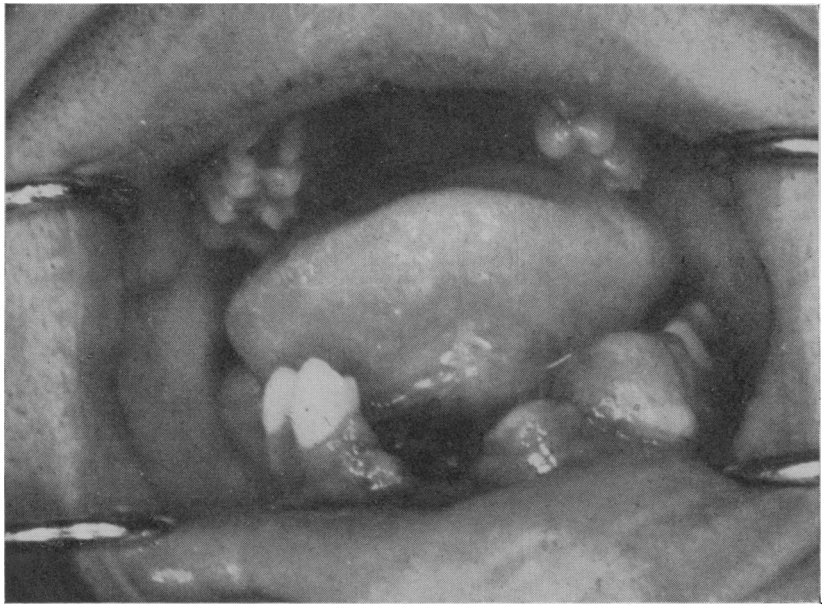

Fig. 15. Peripheral Fibrosarcoma of Mandible.

The tumour may be easily diagnosed from the roentgen picture, as the tissue is quite dense and frequently shows invasion and destruction of bone.

It is likely to recur even after radical removal, and visceral metastases occur early through hematogenous transportation of tumour cells.

Central Fibrosarcoma. The central variety may arise from the connective tissue of the nerve trunks or blood vessels. This tumour may not be recognized when small unless routine X-ray examination discloses it. It appears as an osteolytic area. It is infiltrating in character and may break through the bone. Symptoms occur when the teeth are encroached upon. They become loose and are often extracted without the disease being recognized. Pain and swelling of the expanding jaw are other signs. The maxillary sinus and the nasal cavity may be involved, and the orbit may be encroached upon, causing exophthalmos. Pathological fracture may occur in the mandible (Fig. 16). The histological make-up of fibrosarcoma may show spindle-shaped nuclei, oval cells with vesicular nuclei, or large cells with pleomorphic nuclei, together with tumour giant cells. The number of mitotic figures varies with the rapidity of growth (Fig. 17).

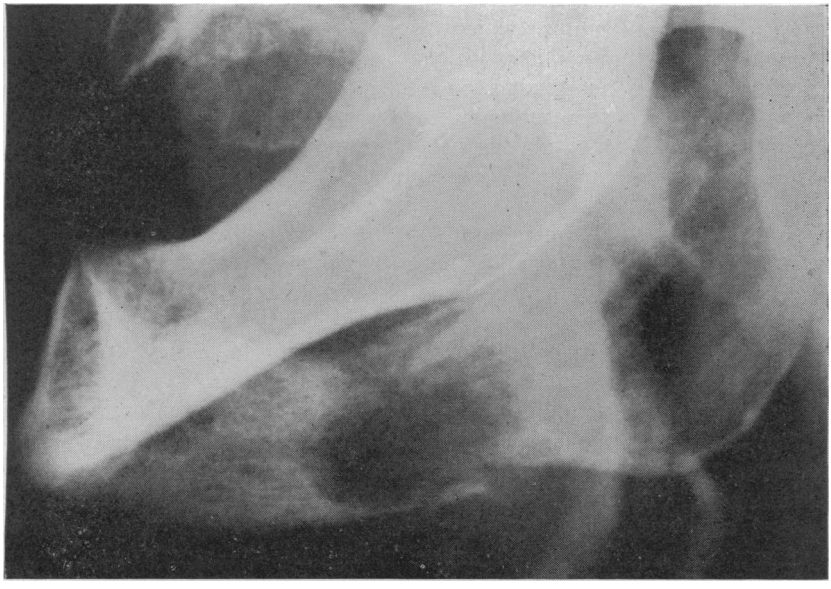

Fig. 16. Central Fibrosarcoma with pathological fracture of mandible.

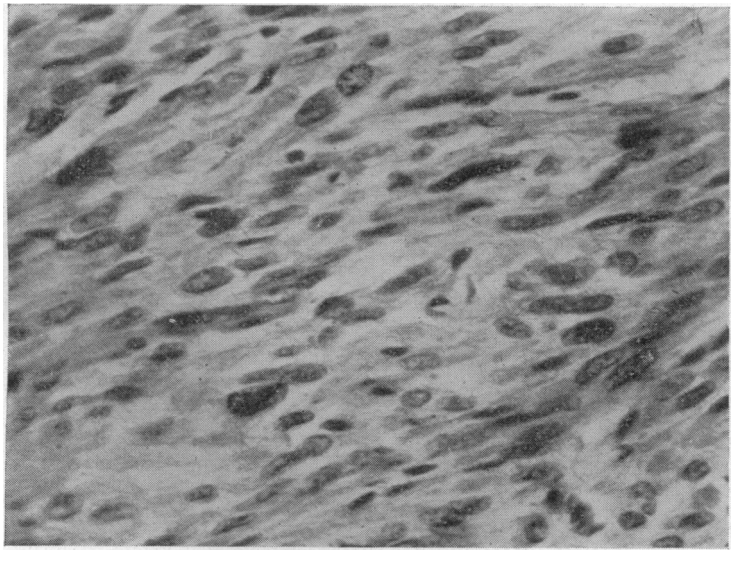

Fig. 17. Photomicrograph of section from Fibrosarcoma shown in Fig. 16.

FIBRO-OSTEOMA

This is a benign tumour which originates in the marrow spaces of the spongiosa. The tumour, because of its great variability in the proportion of fibrous to osseous tissues and the varying degrees of calcification encountered, has been described under many different names, i.e., ossifying fibroma, osteofibroma, fibrous osteoma, by those who concede that it is a tumour, while others who dispute its neoplastic nature prefer the terms localized osteitis fibrosa, localized osteodystrophia, and monostotic fibrous dysplasia. Schlumberger (1946)[5] of the Army Institute of Pathology in Washington, who reported 67 cases of fibrous dysplasia, including five of the maxilla and two of the mandible, feels that the disease represents a non-specific abnormal reaction to injury, producing a connective tissue overgrowth. He points out that similar connective tissue replacement of the marrow cavities and cancellous bone is seen in osteomyelitis, osteomalacia, rickets, and the healing of fractures, as well as in certain systemic diseases such as generalized osteitis fibrosa, and osteitis deformans. The fibro-osteoma, however, has no constant relationship to injury and does not represent, according to my findings, a reparative process of a variety of local or systemic factors. It develops without apparent cause and cannot be associated with regional injury or infection. Nor are vitamin deficiencies or endocrine influences, such as hyperparathyroidism, associated with it, since in none of the cases could an abnormal blood chemistry be discovered. Also, no relationship with neuro-fibromatosis has been established, since none of the facial cases in my experience presented areas of pigmentation, so-called " café au lait " spots, described by Albright, Butler, Hampton and Smith (1937)[6] and lately by Tannhauser (1944)[7].

From my clinical findings and pathological study, I can only deduct that the fibro-osteoma is a tumour which causes the marrow spaces to be filled with actively proliferating young osteogenic connective tissue cells. As the tumour tissue accumulates, resorption of the bone trebeculæ takes place, and later when more tissue is formed, the region of the jaw in which the tumour formed, expands. This process is similar to the development of a tumour formed from hemopoitic tissue, such as a myeloma, except that in the fibro-osteoma the tumour cells also produce intercellular substance which is not the case in myeloma.

The formation by metaplasia of various types of new but abnormal bone is a characteristic of the degree of differentiation and versatility of the osteogenic tumour cells. For this reason various types of fibro-osteomas may be observed.

In some cases the fibrous tissue predominates and grows massively, replacing completely the trabeculæ of the old spongiosa. New immature bone trabeculæ may be laid down in certain places (Fig. 18A). This type we may conceivably call ossifying fibroma if a subdivision is desirable.

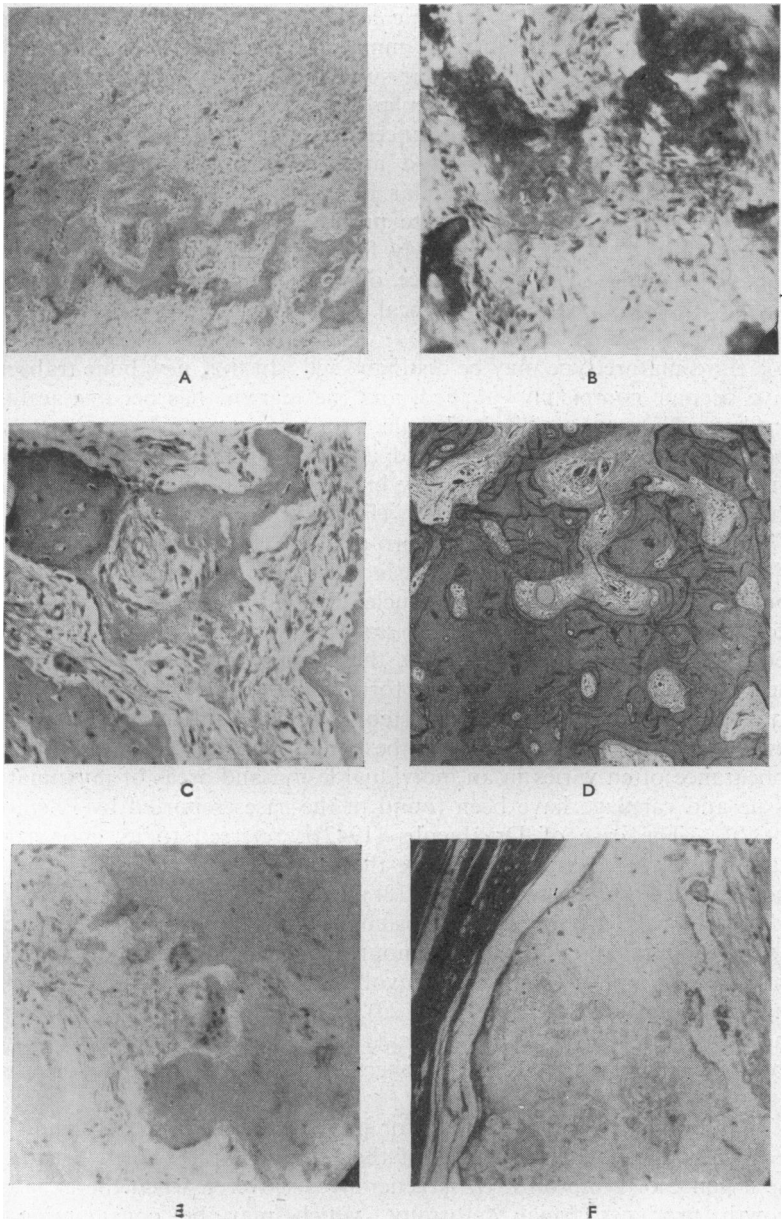

Fig. 18. Fibro-osteoma :—Photomicrographs of : A.—ossifying fibroma ; B.—intercellular deposit of osteoid ; C.—trabeculæ of osteoid; D.—mature bone trabeculæ with cement lines forming mosaic pattern ; E.—focus of giant cells causing bone resorption ; F.—fibro-osteoma with formation of cartilage.

In the second type, the osteogenic connective tissue cells fill the marrow spaces of the spongiosa, forming immature, so-called fibrillar bone from condensed, hyalinized, intercellular substance, which is deposited around them (Fig. 18B), while the trabeculæ of old bone are undergoing osteoclastic resorption. In other instances, the bone is partly or completely replaced by trabeculæ of osteoid and, in some cases, lamellar bone (Fig. 18C). The immature fibrillar bone is formed by fibroblasts of a low level, while the trabeculæ are produced by fibroblasts of a higher level of differentiation. The newly formed bone has irregularly placed osteophytes, and there is evidence of apposition and resorption. The osteoid in these tumours is so typical that the term fibro-osteoid osteoma seems fully justified.

A third mature type may be distinguished. In this, new bone trabeculæ have become completely calcified, and the marrow has become acellular or fibrous in nature. The trabeculæ again are arranged irregularly and show evidence that resorption and apposition had been actively going on previously. They are united by broad cement lines, which stain deep blue and provide a mosaic pattern (Fig. 18D).

The main characteristic of the fibro-osteoma, therefore, is the formation of atypical bone by direct metaplasia of connective tissue. The cells are rounded out, present vesicular nuclei and are surrounded by profuse deposits of osteoid which calcifies slowly but not completely and stains deeply with eosin. In other cases, the cells have applied themselves to the periphery of metaplastic bone, forming more or less calcified atypical bone trabeculæ. In most cases osteoblasts are numerous, and in some cases areas rich in giant cells may be noted (Fig. 18E). The histological appearance often varies in an individual lesion and areas of myxomatous tissue and cartilage have been found in the cases reported by Phemister (1937)[8]. The case of Dr. Jacobs (1942)[4], referred to as chondroma, may fit into this classification, since the greater part of the tissue had the character of a fibro-osteoma (Fig. 18F).

The clinical features of fibro-osteoma are quite as characteristic as the pathological make-up of the tumour. The tumour occurs in the maxilla as well as the mandible. Bilateral involvement and simultaneous maxillary and mandibular occurrence (Fig. 23), has been observed by Phemister and Grimson (1937)[8] and Thoma (1949)[9]. In this respect, therefore, fibro-osteoma resembles the peripheral osteoma, which also occurs in multiple form.

The tumour has a predilection for individuals in the second and third decades of life. The early stage of the tumour is rarely seen because of the absence of symptoms. It is generally discovered when the expansive growth produces facial deformity, which may be considerable, or disturbances in the occlusion of the teeth. The duration of the tumour is, therefore, difficult to ascertain ; patients state it to be from one to 30 years. No cases have been reported that have undergone sarcomatous changes.

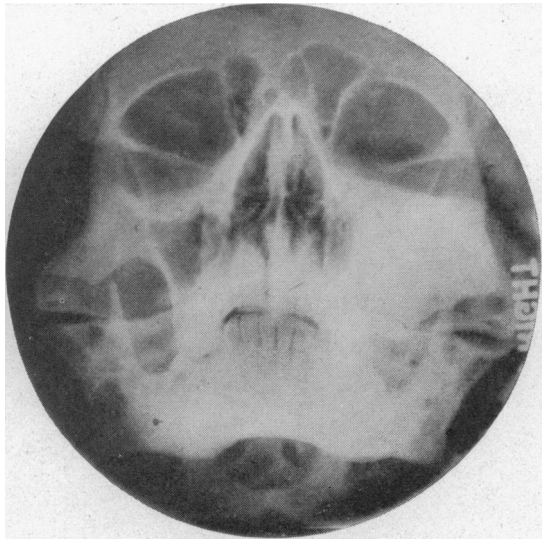

Fig. 19. Fibro-osteoma of maxilla involving maxillary sinus and malar bone. (Photomicrograph shown in Fig. 18C.)

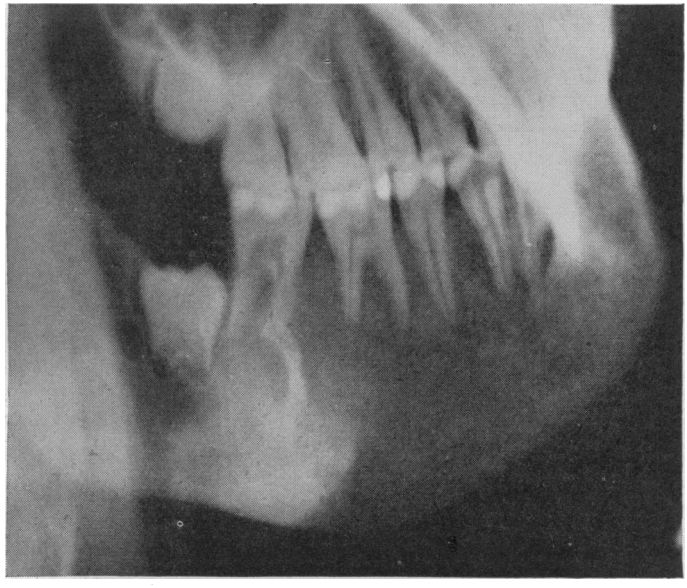

Fig. 20. Fibro-osteoma of mandible with expansion of bone.

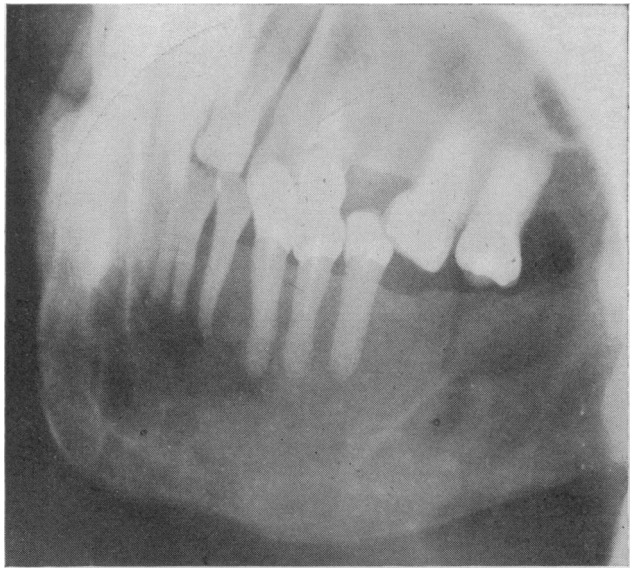

Fig. 21. Fibro-osteoma of mandible, osteoporotic type. (Photomicrograph shown in Fig. 18E.)

In the maxilla the typical case includes obliteration of the maxillary sinus and causes expansion of the canine fossa as well as the palate. The malar bone is generally involved and the eye may be pushed to a higher level causing diplopia. Curiously, the nasal wall is not involved, and the nasal passages remain normal. The roentgen film shows a characteristic picture, a dense mass is seen filling the maxillary sinus and obliterating its walls (Fig. 19). In a side view, the increased density gives a shadow of a typical stippling, particularly if the tuberosity is involved and enlarged. This effect, as Worth (1937)[10] pointed out in an article in the *British Journal of Radiology*, resembles the texture of orange peel.

The radiopacity of the tumour, of course, depends on the amount of osteoid and degree of calcification present.

In the mandible, the lesion may be limited in extent or may involve a large part of the bone, causing expansion of the alveolar process with irregular deformity of the chin or side of the face. Roentgen examination may reveal a circumscribed area of decreased density, which may be cystic in character, or it may show diffuse involvement without distinct demarcation. In some cases the cortex is expanded with convex bulging and thinning of the inferior border of the jaw (Fig. 20). A case of this type, which I would classify as fibro-osteoma, was reported by Rushton (1946)[11]. The radiopacity of the bone varies greatly. In the type for which the term ossifying fibroma has been suggested, the tumour is mostly radiolucent, though there may be islands of greater density due to deposit

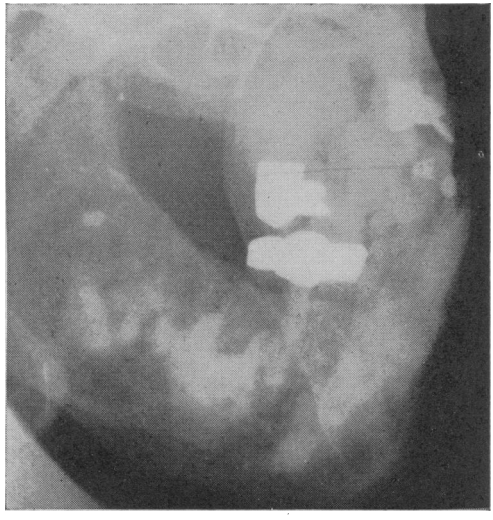

Fig. 22. Fibro-osteoma of mandible, hyperostotic (mature) type in woman 56 years old, involving both maxillæ and mandible. (Photomicrograph shown in Fig. 18D.)

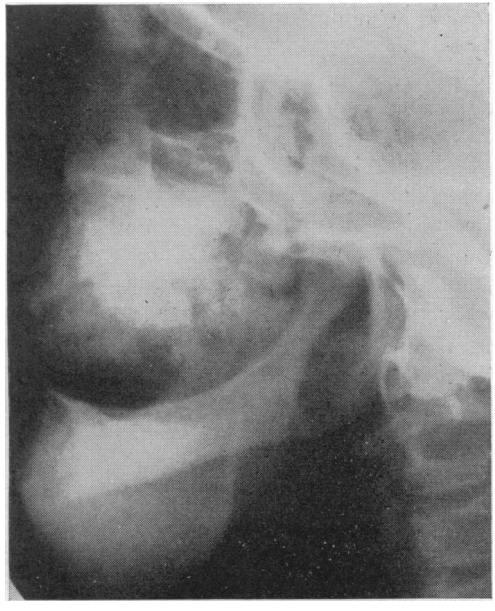

Fig. 23. Fibro-osteoma of mandible-hyperstotic type.

of partly calcified osteoid. Extensive but gradual resorption of the calcified bone trabeculæ and cortex also causes a radiolucent defect (Fig. 21), while the replacement by ossifying fibrous tissue gives an irregular granular appearance. If mature bone is formed, circular or jagged areas of great density result (Fig. 22). The hyperstotic form of the tumour seen in older individuals presents a very radiopaque appearance and gives the picture of an osteoma (Fig. 23).

OSTEOGENIC SARCOMA

The registry of Bone Sarcoma of the American College of Surgeons includes under this name all malignant tumours derived from ancestors of cells, which, when duly differentiated are known as osteoblasts.

Because of the rapidity of growth, we may see all stages of development of the osteogenic mesenchymal cells with production of their respective intercellular substances, fibroblasts, with collagen and myxomatous tissue, chondroblasts and cartilage, and osteoblasts depositing osteoid or bone.

Jaffe (1935)[12] suggested that osteogenic sarcoma may be the malignant counterpart of osteoid-osteoma and indeed there is much that could be said in favour of linking fibro-osteoma with osteogenic sarcoma, except that it would suggest unnecessarily and needlessly a bad connotation on a benign and harmless disease.

Clinically, we find that the osteogenic sarcoma occurs most frequently in young, strong and vigorous individuals and has an extremely poor prognosis. It may involve the maxilla or the mandible, causing pain, a sense of pressure with loosening of the teeth, and paresthesia of the face.

Osteolytic and osteoblastic types are generally recognized, though both conditions may be presented in one tumour. In those predominantly osteolytic, a great deal of bone is destroyed and replaced by fibrous and myxomatous tissue and cartilage, while in the osteoblastic or sclerotic variety, there is a high degree of bone produced, not only in the tumour but also subperiosteally. The latter causes the well-known " sunray " effect. In other cases, osteolytic and osteoblastic formation may be balanced in such a manner that very little change in radiability can be demonstrated.

The roentgen examination is important and gives a great deal of information regarding the part of the jaw involved. Signs which are diagnostic were set down by Codman (1926)[13]. These are combined central and subperiosteal involvement, the presence of the old shaft or cortex in the tumour, the invasive character of the tumour, osteolytic and osteoblastic components being present simultaneously, and involvement of the soft parts which surround the bone.

The clinical course and rapidity of growth vary greatly. Osteoblastic tumours usually grow slowly and are not very malignant, while the soft vascular type which shows cells of great variation in size and shape with many mitotic figures, has a very poor prognosis. Recurrence after even very radical surgery is not uncommon and metastases form in the lungs rather than in the lymph nodes and bones. Most cases end fatally inside of two years.

TREATMENT OF OSTEOGENIC TUMOURS

Most of the benign, highly differentiated osteogenic tumours are treated simply by excision. The mandibular condyle if involved, generally must be resected. The fibro-osteoma presents a greater problem. Only in very small early lesions can the tumour be completely excised. In most cases the jaw is so extensively affected that the operation should be limited to partial removal to eliminate a deformity, keeping in mind that slow recurrence calls for over- rather than for under-correction. I noted that in a report by Dr. Martin Rushton, published in the *Proceedings of the Royal Society of Medicine* (1947)[14], a patient was treated by radiotherapy preparatory to a cosmetic reduction of the bony prominences. The procedures we have experimented with are the reverse; we have followed Phemister and Grimson's advice to treat the patient with irradiation postoperatively. We have administered to three patients 150 R. twice a month for six months, but found that it did not greatly alter the course of the tumour. Phemister and Grimson, however, recommend a total of 3,000 R. given in a similar way over a protracted period of time. In many cases repeated cosmetic surgical procedures are required and with advancing age, no doubt, the tumour attains a state of maturation of the bone with atrophy of the cellular elements, eliminating further growth.

The malignant tumours, especially the osteogenic sarcoma, require prompt radical interference, not only of the affected bone, but of the surrounding tissues as well. Radical resection may occasionally save the life of the patient, although the prognosis is invariably very poor.

REFERENCES (Part II)

1 RUSHTON, M. A. (1944) Growth of the Mandibular Condyle in Relation to Some Deformities. *Brit. Dent. J.* **76**, 57.

2 FENNEL, E. A. (1938) Osteogenic Sarcoma from Exostoses. *Amer. J. Surg.* **39**, 121.

3 THOMA, K. H. (1936) Osteodysplasia with Multiple Mesenchymal Tumors : Fibroma, Exostoses, and Osteomas. *Amer. J. Orthodont.* **22**, 11, 77.

4 JACOBS, M. (1942) Cystic Osteochondroma of the Mandible. *Amer. J. Orthodont.* **28**, 499.

5 SCHLUMBERGER, H. G. (1946) Fibrous Dysplasia of Single Bones. *Milit. Surg.* **99**, 504.

6 ALBRIGHT, F., BUTLER, A. M., HAMPTON, A. O., and SMITH, P. (1937) Syndrome Characterized by Osteitis Fibrosa Disseminata, Areas of Pigmentation and Endocrine Dysfunction with Precocaine Puberty in Females. *New Engl. J. Med.* **216**, 727.

7 TANNHAUSER, S. J. (1944) Neurofibromatosis and Osteitis Fibrosa Cystica Localisata et Disseminata, *Medicine, Baltimore.* **23**, 105.

8 PHEMISTER, D. B., and GRIMSON, K. S. (1937) Fibrous Osteoma of the Jaws, *Ann. Surg.* **105**, 564; reprinted *Amer. J. Orthodont.*

9 THOMA, K. H. (1944) *Oral Pathology*, 2nd edition, St. Louis, C. V. Mosby Co., p. 988.

10 WORTH, H. M. (1937) Tumours of the Jaws. *Brit. J. Radiol.* **10**, 223.

11 RUSHTON, M. A. (1946) Solitary Bone Cysts in the Mandible. *Brit. Dent. J.* **81**, 37.

12 JAFFE, H. L. (1935) Osteoid-Osteoma. *Arch. Surg.* **31**, 709.

13 CODMAN, E. A. (1926) Diagnosis of Osteogenic Sarcoma. *Surg. Gynec. Obstet.* **42**, 381.

14 RUSHTON, M. A. (1947) Regional Osteitis Fibrosa Affecting the Facial Bones. *Proc. Roy. Soc. Med.* **40**, 316.