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HÆMANGIOMA OF THE SPINAL CORD

ASSOCIATED WITH SKIN NÆVI OF THE SAME METAMERE*

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HÆMANGIOMATA of the spinal cord and its membranes are rare, and in the published lists of cord tumors no record is found of their occurrence (viz., Schlesinger's list [15] of 400 cases and Flateau's [14] of 213). A thorough review of the literature, however, brings to light seven cases beside the one here reported.

GAUPP,[1] in 1887, describes two cases in an inaugural dissertation embodied in *Ziegler's Beiträge* for that year. The first has no clinical history, and tells of the finding at autopsy of an angioma on the cauda equina measuring three by two cm.; above this point in the cord were two neurofibromata, and a glioma with syringomyelia in which was a fresh hemorrhage. He considers that these three different tumors each arose from an embryonal defect in the cord.

His second case [2] is more interesting and gives the history of a woman of forty-five who had a condition diagnosed as "spinal meningitis" at twenty-one, followed by weakness of her legs, and paræsthesia slowly increasing for ten years; during the next four years she could limp about, and then both legs suddenly became spastic and anæsthetic with motor and vasomotor paralysis, and incontinence of urine and fæces. This condition improved a little during the next few months, but for ten years more she was a cripple and finally died of decubitus infection. The post-mortem examination showed a widening of the spinal canal in the lower dorsal and upper lumbar vertebræ, with a large varix of veins arising from the pia which pressed upon and flattened the spinal cord.

The next case reported is one by BERENBRUCH,[3] in 1890, in another inaugural dissertation, at Tübingen. This patient, a boy of sixteen, was born with lipomata showing on his left side over the pectoralis muscle, on the right scapula, and along the latissimus dorsi on the right; these increased in size as he grew older, and at the time of the onset of the paralysis were quite deforming, the one on the right scapula being as large as a boy's head, and the one over the latissimus dorsi, though less prominent, made that whole side of his back appear enlarged. One day after severe exercise he felt weak and tired in his legs; this persisted, and he had lancinating pains down both legs; at the end of two weeks he was entirely paralyzed in the right leg and partially in the left, with increased tendon reflexes and clonus. Sensory perception was dulled, but not lost, up to the middle of the abdomen. A diagnosis of lipoma in the spinal canal was made, and he was operated on two and a half months after the onset of the disease,

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but the three-hour operation exhausted him, and death followed in four hours. Autopsy showed that beside the three superficial lipomata, there was a large cavernous plexus of veins along the spinal column connecting with the two tumors of the back outwardly, and with an angioma of the dura by way of the intervertebral foramina. Furthermore, there was an angioma of the cord substance itself extending from the fifth cervical to the third dorsal segment, and in parts of this were extravasations of blood. Lastly the capsule of the right kidney showed a small angioma. All of these vascular tumors were made up of large and small blood-vessels; the external ones were mixed with fat, the dural with connective tissues, while the cord tumor was a pure hæmangioma.

In 1895 GERHARDT[4] reported a case in great detail. The patient was a man of eighteen; his first symptoms were weakness of the right foot and lower leg, followed eight days later by weakness of the left; "cramps" were then felt in both legs for a few days, while the weakness spread upwards until the whole of both legs was involved and walking became impossible. It was then found (four weeks after the onset) that anæsthesia was present up to the knees. Two months later the paralysis was total in both legs with loss of vesicle and sphincter control, and six months after the onset, hypæsthesia and paræsthesia extended up to the level of the navel; the knee and ankle jerks became exaggerated, and contractures developed in the leg muscles, with a thoracic scoliosis and lumbar kyphosis. For the next two years there was little change except that the hypæsthesia became total anæsthesia and the contractures increased. During the third year the joints became ankylosed, and in the fourth year the deep reflexes were lost. Finally erysipelas developed from a decubital ulcer, and he died in the sixth year of the disease. At autopsy a vascular tumor, diagnosed by von Recklinghausen as angioma, was found pressing on the cord at the level of the fifth and sixth thoracic vertebræ.

HARMAN AND BALK[5] reported a case in 1900. This was another long drawn-out story of a man who began by having pain in his legs, this lasted four or five months and then the pain suddenly increased, he lost motor power in his legs and opisthotonus developed. These symptoms gradually passed away in six months. Two years later he had a similar attack, but at the end of five months he was able to walk, though lame. His third attack came two and a half years later, and was at first slight, only interfering with locomotion for two weeks, but then, while in bed one day, he had a sudden great pain with retraction of the head, followed by difficulty in swallowing and articulation; later his sphincters became paralyzed, and in ten days he was dead. Autopsy showed a large blood clot in the spinal canal overlying the lumbar cord where there was found a cavernous angioma of the pia.

A year later LORENZ[6] had a case in Jena: a woman of twenty-seven, who first noticed weakness of her left hand. Two days later she suddenly collapsed with a sensory and motor paralysis of both legs, partial in both arms, and in a few hours her breathing became labored and she died two and a half days after the onset of the first symptom. The post-mortem examination disclosed an angioma of the pia, about the size of a cherry, at the level of the seventh cervical root, and a hemorrhage into the arachnoid space and cord substance from the first cervical to the fifth dorsal root.

The last case is one reported in 1903 by HADLICH.[7] This, like the first of the series, was an accidental autopsy finding. A woman of thirty-five, dwarfed and with a narrowed pelvis, died after Cæsarean section. At autopsy an angioma of

the pia was found in the lumbar region; it invaded the cord substance somewhat and caused marked distortion, but no hemorrhage was present.

CASE HISTORY.—E. A. P., an eight-year-old boy of Swedish extraction, entered the medical service in the Children's Hospital on May 23, 1914. Previous to the onset of the present illness, he had had a rather varied medical career; he was a full term, normally delivered child, well and vigorous during his first year. He then had measles, followed six months later by typhoid with bloody diarrhœa; during his convalescence, he sprained his right knee and was lame for a month, and then, at two years of age, he developed pertussis. During his third year he was well except for an enlarged lymph-node behind the angle of his right mandible, which disappeared without operation after several weeks. When he was four years old, he suddenly screamed with pain, complaining of his lower abdomen, and on examination a hernia was found in the left inguinal region, which was reduced with chloroform; for this he wore a truss for a year, and there has been no recurrence. From this time until he was eight years old, he enjoyed a healthy, normal boyhood.

Present Illness.—On May 15, 1914, he was suddenly stricken, while playing ball, with a sharp pain in his lower back, causing him to cry out. He was taken home feeling weak and nauseated, but without fever. He vomited once during the night after taking some medicine, but the main symptom was pain in the back, radiating into the lower abdomen. In a few hours this had subsided, but he did not feel well for the next four days, and was kept out of school. He then returned to school, apparently well, but three days later a similar sudden pain in the back attacked him, more severely than the first time, and for several hours he screeched and rolled about in agony. As this severe attack subsided, he said that the left foot felt numb, "as if it was asleep," and this gradually spread up the leg to the thigh, and then "down the other leg," till at the end of fourteen hours both lower extremities were numb and paralyzed. It was noted definitely that the left leg first became numb, but in which leg the paralysis first appeared is not known. The next morning it was found that the anal and vesicle sphincters were also paralyzed, there being incontinence of fæces and retention of urine, necessitating catheterization. On this day, May 23, eight days after the onset, he entered the Children's Hospital, with a presumptive diagnosis of anterior poliomyelitis.

Physical Examination.—This showed a flaxen-haired, blue-eyed boy well nourished and well developed, with a deep depression at the lower end of the sternum, and a birth-mark of the "port wine" variety on the left back. The neck was held stiffly and was perhaps slightly tender. The abdomen was also held

stiffly, but the liver edge was palpable, and a mass above the pubes, evidently a distended bladder. Both legs showed a complete flaccid paralysis, with absent knee-jerks, and no ankle clonus, Babinski, or Kernig.

Sensory examination revealed analgesia up to the level of the umbilicus, and anæsthesia except for the soles of the feet, where a sharp pin prick was felt dully. The abdominal reflexes were absent, and the cremasteric were sluggish. There was incontinence of urine. A lumbar puncture was done, and 40 c.c. of spinal fluid were withdrawn under increased pressure, showing twenty cells per cubic millimetre, 85 per cent. of which were

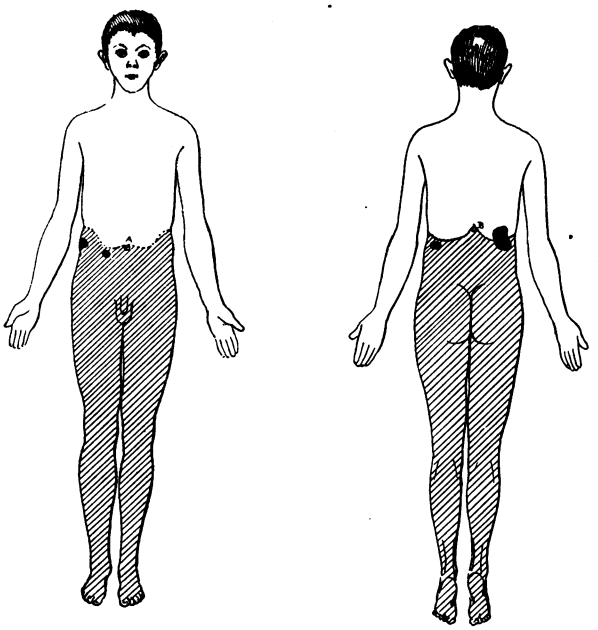

FIG. 2.—Diagram showing area of anæsthesia and position of nævi. *A*, umbilicus; *B*, spinous process of first lumbar vertebra; ■, nævi; ///, anæsthesia area.

mononuclear, and 15 per cent. polymorphonuclear. A von Pirquet skin test was positive in twenty-four hours; the leucocyte count was 15,000, and hæmoglobin (Sahli test) was 82 per cent.

Course of Symptoms.—For the first six weeks the paralysis was of a flaccid type, so the supposition that the disease was poliomyelitis was a natural one. Priapism was first noted on June 1, and persisted until after the operation. Then on July 3, a slight return of the knee-jerk in the right leg was noted; this leg was less flaccid and did not seem to fall so limply as previously. On July 4, stimulation on the soles of the feet caused flexion of the knees and ankles, and on the sixth the right knee jerk was ex-

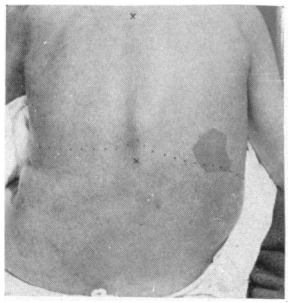

FIG. 1.—Photograph six weeks after operation, showing upper level of anæsthesia (dotted line) crossing lower portion of nævus, and operative incision (between crosses).

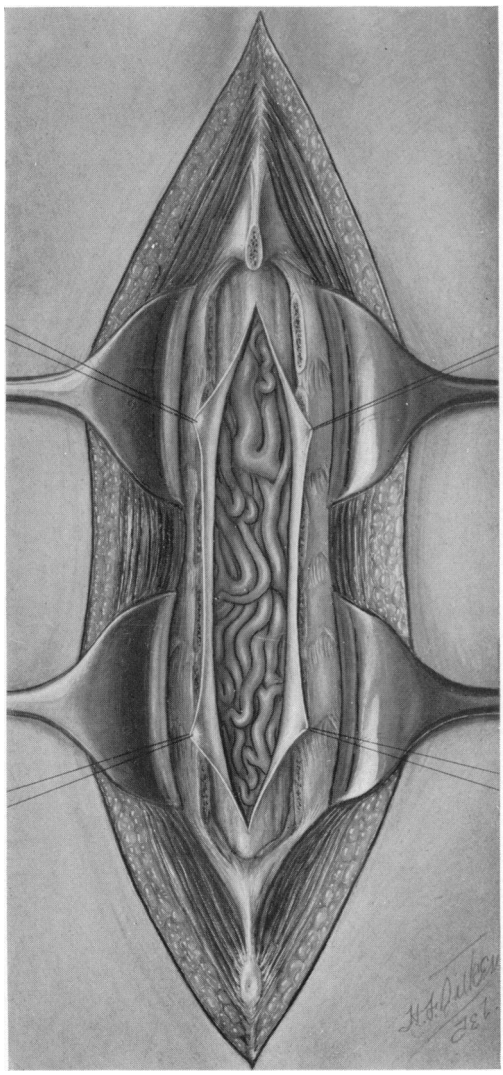

FIG. 3.—From sketch made at operation, showing incised dura with hæmangioma beneath (about one-half natural size).

aggerated, and ankle clonus was obtainable on the right, appearing on the left two days later.

On August 4, Oppenheim and Babinski reactions were found in both legs, the paralysis thus becoming completely and typically spastic ten and a half weeks after the first paralysis developed. During this time there was no return of voluntary muscle power or sensation, the anæsthetic area still extending up to the umbilicus. A swelling along the right side of the vertebral column from the fourth to the eighth dorsal vertebra was noted on September 17, but an X-ray of the chest was negative.

He was transferred to Professor Lovett's service on September 21 with a diagnosis of spinal cord tumor, and the following notes were recorded: "As he lies in bed the spine shows quite a marked right dorsa-lumbar scoliosis, and the ribs on the right side, posteriorly, between the scapula and spine, are more prominent than on the left. The patient is unable to sit or stand on account of loss of power of the supporting muscles of the back and abdomen. The trochanters are very prominent, with atrophy of the gluteal muscles. The Achilles tendons on both sides are contracted, holding the feet in moderate equinus. The Wassermann reaction on the blood is negative." On an exchange visit, soon after this, the patient was seen by Dr. Cushing, who ventured a diagnosis of congenital dermoid or hæmangioma; he based this on the fact that the skin nævus on the child's back lay in the same metamere as the presumed tumor of the spinal cord (Fig. 1).

On October 16, the patient was transferred to the surgical service at the Peter Bent Brigham Hospital. The neurological and general physical examinations here revealed nothing new, except that three more small nævi were noted about the trunk at the level of the umbilicus. The epigastric reflexes were present, but the abdominal and cremasteric were absent. The knee-jerks were hyperactive, ankle clonus was present on both sides, and Babinski, Oppenheim, and Gordon reactions were all obtained in both legs, but more markedly in the left. The sensory disturbances showed no change, anæsthesia extending up to the umbilicus anteriorly, to the tenth rib laterally, and posteriorly to the level of the first lumbar vertebra (Fig. 2). It was, however, noted that the toes showed some sense of position, more in the left foot than in the right. The urine was cloudy and showed many pus cells and a slight trace of albumen; the leucocyte count was 19,000; the hæmoglobin was 80 per cent.; the pulse ranged from 80 to 120, and the temperature from 98° to 100.2° Fahrenheit.

Dr. Cushing's note on the day of operation follows:

"This child shows a transverse paraplegia with the upper level of anæsthesia about at the tenth thoracic level. It has been presumed, owing to the acute onset of symptoms, that the case was

one of poliomyelitis of an unusual type. However, the definite upper level of anæsthesia, the complete lower limb paralysis with exaggerated reflexes, visceral paralysis, and priapism, make it fairly definite that there must be pressure against the cord. Examination shows a slight scoliosis with prominence of the spinal muscles at the scapula level, and over the lower portion of the right scapula there is a nævus about 5 inches in diameter. The presumptive diagnosis is of a congenital lesion pressing upon the cord, either a dermoid or an angioma. Similar meningeal angiomata have been observed by myself in association with facial nævi."

Operative Note.—October 30, 1914. *Exploratory laminectomy. Angioma.* "A long median incision was made with removal of the laminæ, possibly from T6 to T10 inclusive. The exploration was simple and dry. The exposed dura was bulging, and it was the operator's impression that the canal was much wider than usual, particularly for a thoracic region, and that the laminæ were unusually broad, flat, and thin.

"The exposed dura was tense, bulging, and transmitted an unusually dark subdural coloration. Fortunately great pains were taken to enter the dura without injuring the arachnoid, for a careless entrance would certainly have injured some of the enormous vessels which were disclosed on opening the meninx. Such of the arachnoid membrane as could be identified was more or less adherent to the dura by fine adhesions which readily broke down as the dura was drawn to each side. This exposed an extraordinary tangle of huge pulsating vessels filling the canal (see Fig. 3). It was possible in a few places to see the normal coloration of a very flattened spinal cord, through and below the meshwork of vessels.

"It seemed futile to attempt to ligate any of the vessels, and the dura was therefore left widely open, and the wound closed as usual in layers, without a drain."

The patient made a good ether recovery, but had a very uncomfortable convalescence. Careful watch was kept of the sensory disturbances, but no definite improvement was noted at any time. Three days after operation the priapism was still present, but on the fourth it disappeared, and has not returned. This is, unfortunately, the only symptom relieved by operation, the spastic paralysis, incontinence, and anæsthesia remaining. Some slight sensory changes appeared from time to time. At first the level of response to a heavy pin prick seemed to spread down the right leg as far as the groin, anæsthesia for light touch remaining at the level of the umbilicus; the muscle sense in the toes seemed to be more acute, and he could tell quite well in what position his toes were placed. On November 22 it was noticed that the right foot

was definitely colder than the left, and this condition continued until about December 14. On January 26 a spontaneous fracture of the femur occurred, and an X-ray showed extreme atrophy of the bones. So as time wore on, and no recovery seemed possible, it became a struggle against decubitus and cystitis.

A neurological examination on April 16 showed no change, except that there was a band of hyperæsthesia at the upper limit of the anæsthesia, and on the right a segment within the area of protopathic anæsthesia, where light touch could be felt. At the present writing (June 15) the patient is at home in unchanged condition.

Discussion.—This series of seven cases, though small, permits of a comparison which brings out some points of interest. The onset is usually slow, the paralysis not reaching its maximum for days or even months. In two instances, weakness of the feet was the first symptom, preceding paralysis of the legs by two weeks and by six months; in another, weakness of the right hand preceded the paralysis by two days. In three of the cases pain came on suddenly but was not followed by the paralysis for five days in one, two weeks in another, and five months in the third. A history of recurrent attacks after partial recovery appeared twice, while in four of the cases a single attack seemed sufficient to cause a permanent paralysis. Four of the histories note an onset with flaccid paralysis, described as "weakness," the spastic symptoms not coming on for a considerable time—two months, six months, and fourteen years in three of these, while in the fourth the patient died in the flaccid state. In short the symptomatology appears to be that of hemorrhage into the spinal cord or canal, generally recurrent if the patient survives the first paralysis. Death in two of the cases was due to respiratory failure, in two to decubitus, and in two others to operation, while one has no clinical history, and the other patient is alive at the present writing.

Pathologically it is striking that in all of the cases the tumor involved the pia; in two the dura was also involved, and in two the cord substance was invaded. The level of the angiomata on the cord is variable; three were lumbar, one cervical, one cervicodorsal, two dorsal, and one was found on the corda equina. The tendency seems, therefore, to be towards a low position. In two of the cases there was widening of the vertebral canal in the region of the tumor.

One of the most interesting features of the case here reported is the occurrence of nævi on the skin in the same metamere as the angioma of the cord; that is to say, the membranes of the spinal cord in the vicinity of the tumor are supplied by the same nerve roots as the skin on

which the nævi are found. In none of the other cases was a similar observation made, unless the lipomata and deep angiomata in Berenbruch's [3] case may be so considered, but nævi may have been present and escaped notice, or may have been noticed and not thought worthy of record. In searching through the literature on nævi, one case [8] is found where a large angioma of the right flank, in the sensory distribution of the eleventh dorsal segment, was associated with atrophy of the right leg; since this is the opposite of the usual condition of hypertrophy found with angiomata of the skin, it is possible that here is a case where an associated tumor was pressing on the lower dorsal cord; this is only hypothesis, however, as no autopsy was performed. Although no literature is obtainable on the association of nævi with cord tumors, an article by Cushing,[9] in 1906, entitled " Cases of Spontaneous Intracranial Hemorrhage Associated with Trigeminal Nævi," throws much light on the subject. He cites three cases observed by himself, and three other cases found in the literature, where facial nævi were associated with vascular tumors of the cerebral meninges; another case reported by Kalischer,[10] in 1901, brings the total up to seven. Skin nævi have also been found in association with angioma formation in the liver,[11] and in Berenbruch's case,[3] an angioma of the kidney capsule was present.

The subject of the metameric distribution of nævi has been taken up by numerous writers of the French School. Bärensprung,[12] in 1863, made the original communication, describing the topographical distribution of nævi and drawing attention to their development in definite connection with territories of cutaneous innervation. He believed the lesion to be a congenital one of the spinal ganglia because the cutaneous alteration consists in a hypertrophy of the elements in which the peripheral nerves terminate, *i.e.*, the pigmented layer of the skin where the free nerve endings lie, and the blood-vessels of the papillary layer which also receive nerve filaments. Etienne, in 1897,[13] took up the subject thoroughly, and came to a similar conclusion: that a nævus represents an intra-uterine nerve lesion. No one suggested, however, that organs other than the skin, but in the same segment of nerve distribution, could be similarly affected, until the publication of Cushing's article,[9] in 1906. The case in hand seems to substantiate this view, since it shows its application to nerve distributions other than the trigeminal, for in this case, four nævi were found on the skin, three on the right and one on the left side, in the cutaneous area supplied by the seventh, eighth, and ninth dorsal segments, and at operation the vascular tumor was seen to overlie the cord from its fourth to ninth

segment, while the upper limit of anæsthesia is at the level of the ninth dorsal segment. As only an operative incision was made, it is impossible to tell whether or not the angioma extended farther up and down the cord, but from the symptoms it is evident that the main lesion—probably a hemorrhage—was at the ninth dorsal level.

Conclusions.—(1) Skin nævi are at times of diagnostic value when segmental phenomena referable to the central nervous system are present.

(2) Congenital blood-vascular tumors apparently arise from a developmental fault of the central nervous system, so these lesions may occur in any of the organs innervated by filaments from that neuromere.

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