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MULTIPLE PRIMARY HAEMANGIOMATA IN THE BONES OF THE EXTREMITIES

BY

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Primary haemangioma in the bones of the extremities, particularly in the diaphyses, has been so rarely recorded in the literature that a single case deserves to be described in order to throw light on the clinical course of the tumour.

The patient was a 69-years-old fisherman from Sogn, Norway. No previous case of bone tumour in the family is known. In 1905 he was operated for a duodenal ulcer. For at least 20 years he had noticed a lump in the middle of his left leg, anteriorly. When he first noticed it, it was the size of a hazelnut. During the last 6-7 years, since about the middle of 1940, it has been tender if he knocked it. At the beginning of November 1947, he consulted a surgeon about a lipoma on his head and at the same time showed him the lump on his leg.

A roentgenogram was taken of the leg on November 17th, and the roentgenologist reported: "In the middle of the left tibia anteromedially, there is a depression a little bigger than the size of a fig, with a sclerotic bottom and a slightly raised border. It corresponds to the palpable tumour. It is probably a chondroma, or a slowly growing tumour which is eating into the underlying bone."

Diagnosis: Probably chondroma. Secondary finding: Arteriosclerosis.

The patient was referred for surgical treatment to Höyanger Hospital, where the condition on November 27th was found to be:

"Left leg: Anteromedially in the middle of the tibia is seen a tumour 5 × 3 cm. broad, which makes the skin bulge forward rather more than 2 cm. The skin is movable over the tumour. The tumour has, anteriorly, a central area, the size of the tip of a finger, which is softly elastic and is surrounded by bony-hard tissue, which merges directly with the tibia itself. The patient states that this soft area is moderately tender. The remainder of the tumour is not tender or painful. No pulsation is felt in the tumour.

Examination of the rest of the lower limb showed nothing abnormal."

On questioning, the patient said that when he first noticed the tumour the central soft area was about the size of a pea.

The tumour was excised from the left tibia under spinal anaesthesia on November 27th 1947: It was evident that the tumour had originated in the interior of the tibia, and had expanded, pushing before it a

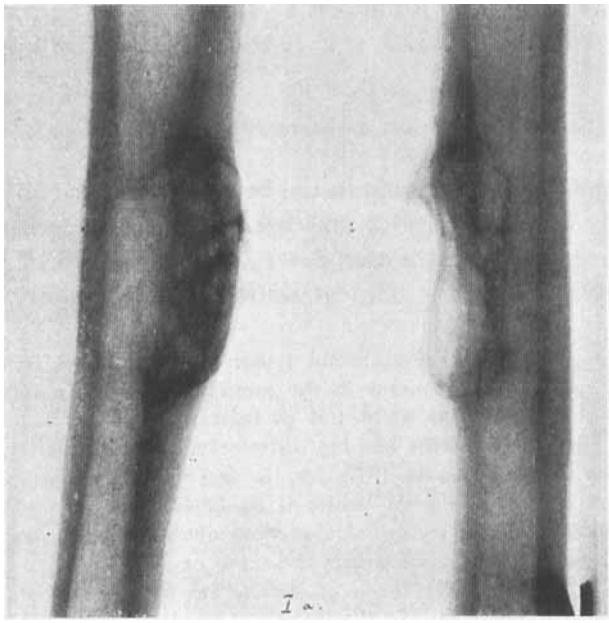

Fig. 1 a.
Hemangioma in the diaphysis of the left tibia. Before operation.

thin lamella of bone. Distally there was a small area without bone covering, and here one could see a soft, bluish-red tumour. The periosteum and the thin bone lamella which covered the remainder of the tumour were removed so that it was exposed. It was easily shelled out, and was found to be the size of a prune, with an obvious capsule. The protruding bone edges were rounded off. On cutting into the tumour it felt soft and was bluish-red with white spots. The surrounding bone was very sclerotic. The periosteum was sutured and the wound closed.

The tumour was sent to Gades Laboratory of Pathological Anatomy, in Bergen. The following report was made:

"Received some irregular bits of tissue, which consist partly of thin-walled small cavities, partly of a more compact, shining substance, with a dark, reddish, spongy tissue amongst the firmer material.

"Microscopically there are a number of small and large endothelium-lined cavities containing red blood corpuscles. Between the cavities there is some degenerated connective tissue, deficient in cells, some connective-tissue with more cells, and some more hyaline connective-tissue. In some places there is a more irregular network of small blood cavities.

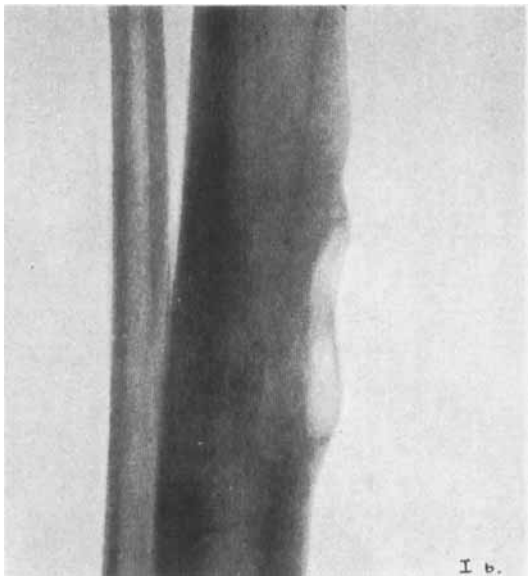

Fig. 1 b.
After operation.

In several places there are blood-clots with early organisation. There is no evidence of malignancy.

Diagnosis: Cavernous haemangioma.

(signed) *Erik Waaler*, Bergen
18.2.48.

The postoperative course was complicated by a left femoral thrombosis, which was treated with heparin.

He was discharged 27.12.47 after primary healing of the wound.

While he was in hospital roentgenograms were taken of the skeleton. They showed a large number of cystic areas in the region of the epiphyses of both radiuses, both ulnae and the right second metacarpal.

Roentgenological diagnosis: Probably haemangiomata.

Cavernous haemangiomata are not very rare in the skin, muscles, vertebrae, flat bones and intestines. Occasionally the tumour is found as a secondary finding at post mortem exam-

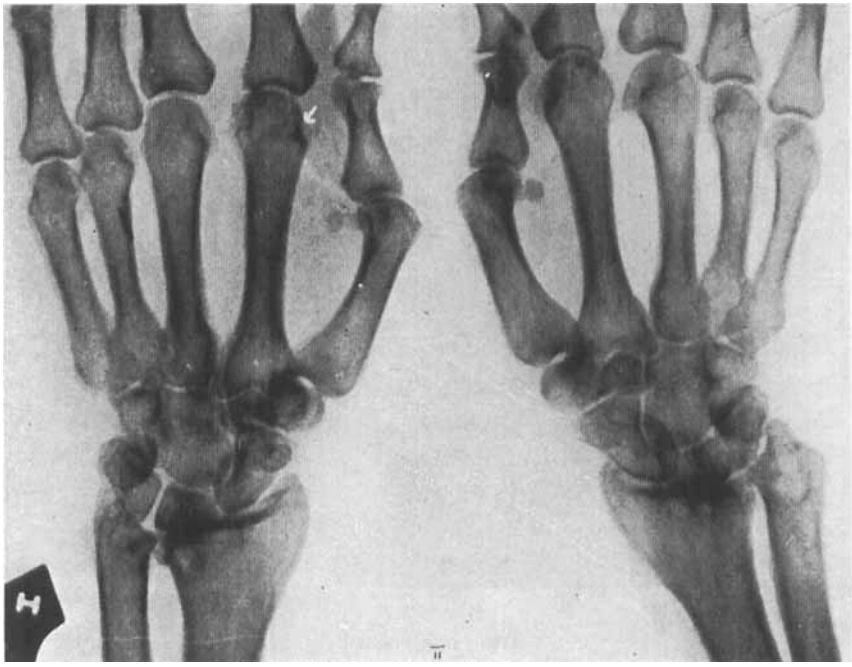

Fig. 2.
Hemangiomata near the epiphyses of both radii, both ulnae and the right 2nd metacarpal.

ination. The literature gives the impression that most workers consider that haemangioma has rarely been proved in the long bones, and very rarely in their diaphyses.

Paul C. Bucy and *Charles S. Capp* (1) examined the literature up to 1930 for cases of primary haemangioma in bone. They mention 11 cases of hemangioma cavernosa in the bones of the extremities. In 4 of these the tumour was in the epiphyseal region of the long bones. They came to the following conclusions on the tumour in the long bones:

"The tumour presents no characteristic clinical signs. The microscopic appearance is that of a benign haemangioma, usually cavernous.

Amputation is never indicated. Excision of the tumour when possible gives the earliest and best results. Roentgen therapy over a considerable period of time is equally effective and satisfactory. Radium is untried as yet, but may prove useful."

In "The Tumour of Bone", 1936, p. 616, there is a picture of a roentgenogram of a haemangioma cavernosa in the femoral diaphysis of a 17-year-old girl.

C. F. Geschickter and *I. R. Maseritz* (2) have examined the literature up to 1938 and write of primary haemongiomata in the bones of the extremities. They mention 3 further cases of cavernous haemangiomata in the epiphyseal regions of the long bones.

In 1939 *Werner Roscher* (3) describes a case of "Haemangiom der Arteria Nutritia der Tibia". It was a tumour the size of a pea, immediately below the middle of the tibial diaphysis, which had enlarged the nutrient canal and caused pain.

John W. Pierson, Georg Farber and *John Howard* (4) have examined the literature up to 1941 and describe a case of "Multiple haemangiomas of bone, probably congenital". In addition to numerous haemangiomata in the pelvis, ribs, vertebrae, skull and clavicle, both the humeri, the right radius and both femora were affected. In the long bones the tumours were situated at either end of the diaphysis in or close to the epiphyseal region. The patient was a 21-years-old man, who consulted a doctor for fracture of the clavicle while playing football. The roentgenogram showed "Fracture through a cystic area in the left clavicle". Subsequently the whole skeleton was examined roentgenologically, and the final diagnosis was verified by biopsy. The authors stress:

1. The excellent general condition of the patient.

2. The diagnosis was only made by biopsy, in spite of clinical and roentgen examination. They add "It is interesting to speculate as to how the patient's disease might have con-

tinued unrecognised had not his fracture necessitated roentgen examination", and, after 5 years of observation: "We have given this patient a sanguine prognosis and urged him to carry out his proposed matrimonial venture".

Ernest A. Pohle and *Elisabeth A. Clark* (5) have examined the literature up to 1945 and write about "Multiple cavernous haemangiomata of the skin, brain and skeleton". In their cases there was a haemangioma in the region of the trochanters of the femur.

Philip D. Wilson (6) writes in his account of experiences with a bone bank about an 11-year-old boy with "Recurrent haemangioma of Right Humerus" at the upper end. (Treatment: Currettage and packing with bone chips after 5 days refrigeration).

Altogether I have found in the literature 20 cases of cavernous haemangioma in the bones of the extremities. 12 of these had tumours in the long bones, 10 in the epiphyseal region, and 2 in the diaphysis. Cavernous haemangioma in the long bones may be classified as a benign primary bone tumour. It consists mainly of blood-channels separated from the vessels of the main circulation (MacCallum). These channels do not behave according to the rules which usually govern the normal development of vessels, but grow more or less irregularly, and form a mass which in varying degree is separate from the general circulation. The tumour grows from its own vascular structure. It grows slowly and expandingly and gives clinical symptoms only after many years. In the present case pressure symptoms only occurred at the end of 20 years and then the tumour was found to have broken through the cortex.

In most cases the tumour gives no clinical symptoms. Sometimes the cortex is forced out and protrudes as a small or large lump from the surrounding normal bone.

The tumour usually appears in the epiphyseal region and has been found in both young and old persons.

The correct diagnosis is frequently not made clinically, but if, as in our case, one feels a soft, elastic tumour in the centre of a bony-hard lump on the bone, one ought to think of the

possibility of its being a haemangioma. Palpable pulsation in the tumour is not necessary.

In some of the cases mentioned the diagnosis has been made from the roentgenograms, but in most of the cases a definite diagnosis was only made after biopsy. A solitary tumour is generally mistaken for a chondroma, while in cases of multiple haemangiomata, xanthomatosis, osteitis fibrosa cystica and disseminata have sometimes been considered as differential diagnoses. Usually it has not been difficult to exclude malignancy.

Cases who have been troubled by the tumour have most frequently been treated surgically. It has usually been sufficient to curette the tumour and chisel off the protruding bone edges. Bone grafting has been used in cases where fracture has occurred through the tumour region and the tumour has been big. While surgery gives the quickest relief to the patient's troubles, roentgen irradiation for a suitable period is recommended for cases where the tumour is not accessible to surgery. The prognosis is considered very good, even in cases with multiple tumours.

SUMMARY

The author has searched the literature up to May, 1947, and has found reports of 12 cases of cavernous haemangioma of the long bones. In 10 cases the tumour was situated in the epiphyseal region, and in 2 in the diaphysis.

The author adds a case of haemangioma cavernosa of the diaphysis of the left tibia to the literature; this is the third case reported of a haemangioma in this position.

In advanced cases the tumour can be diagnosed by clinical examination.

RESUME

L'auteur a passé en revue la littérature jusqu'en mai 1947 et a trouvé le compte rendu de 12 cas d'hémangiome caverneux

des os longs. Dans 10 cas, la tumeur était située dans la région épiphysaire et dans 2 dans le diaphyse.

L'auteur leur ajoute un cas d'hémangiome caverneux du diaphyse du tibia gauche; c'est le troisième cas d'hémangiome ayant cette localisation qui ait été rapporté.

Dans les cas avancés, la tumeur peut être diagnostiquée par l'examen clinique.

ZUSAMMENFASSUNG

Verfasser hat das Schrifttum bis zum Mai 1947 untersucht und Mitteilungen über 12 Fälle von kavernösem Hämangiom der langen Knochen gefunden. In 10 Fällen hatte der Tumor in der Epiphysenregion gelegen und in 2 Fällen in der Diaphyse.

Verfasser vermehrt das Schrifttum mit einem Falle von Haemangioma cavernosum der Diaphyse der linken Tibia; dies ist die dritte kasuistische Mitteilung eines Hämangioms in dieser Lage.

In vorgeschrittenen Fällen lässt sich der Tumor mittels klinischer Untersuchung erkennen.

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