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PRIMARY LYMPHANGIOMA OF BONE

By

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It is well known that lymphangiomas and haemangiomas have many features in common. One of these is the localization, which is similar in both (Wrede, 1911; Winkler, 1924), with one exception. Although primary haemangioma of bone is a fairly common tumour, especially in the vertebral column (Herzog, 1944), primary lymphangioma restricted to bone is exceedingly rare. Cases of lymphangioma of bone reported in the literature have mostly originated in adjacent tissue, thus involving the bone only secondarily (Wrede, 1911; Mandel, 1927; Herzog, 1944). In other cases the lesions in bone have not been pure lymphangiomata but mixed haem- and lymph-angiomata (Wrede, 1911; and apparently also Bickel and Broders, 1947, as reported by Gorham et al., 1954). In the review by Gorham et al. on *Disappearing Bones* references are made to a few additional, less thoroughly investigated cases in which bone-destruction may have been caused by lymphangiomatous tissue. On scrutiny of the literature we have found only two completely investigated cases (Harris and Prandoni, 1950; Cohen and Craig, 1955) of apparently pure lymphangioma (or 'multiple lymphangiectasis') evidently arising in the bone marrow or periosteal tissue. In both these cases lymphangiomatous tissue (or lymphangiectasis) was present also in organs other than the skeleton.

In view of the rare occurrence of lymphangiomatous tissue in bone we are reporting a case of bone-destruction in which the roentgen findings were similar to those in many of the cases reported above. On microscopic examination thin-walled, irregular, vascular structures were found in the marrow spaces, which might be identified as constituting a cavernous lymphangioma. The vascular structures seemed

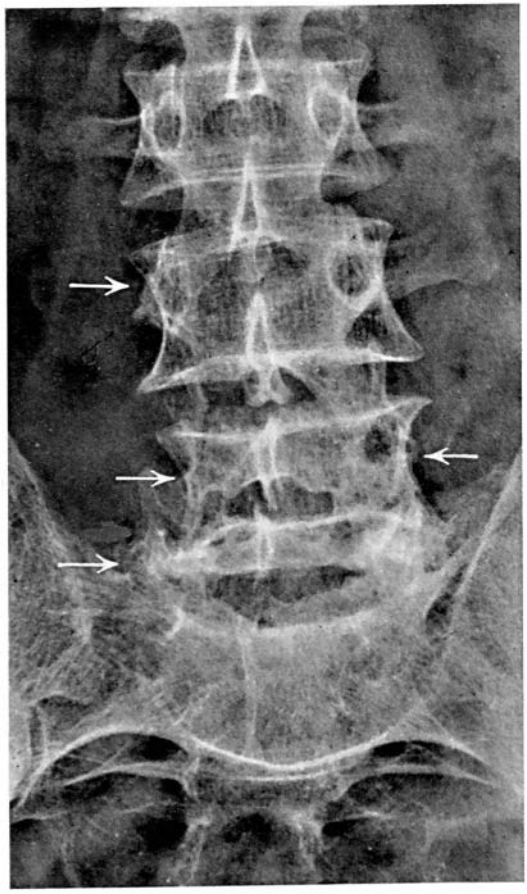

Fig. 1.
Frontal radiogram, lumbar spine. Right transverse processes of L.3 and L.5 almost completely missing. Both transverse processes and pedicles of L.4 totally absent.

to be restricted to the bone and to be free from haemangiomatous tissue. There were no signs of systemic disease.

CASE HISTORY

A builder's labourer aged 55 years was admitted to the Royal Academic Hospital in September, 1955, with a two-month history of backache. The pain was of moderate degree, and was made worse by exercise. There was numbness and a radiating pain in the leg. He had had a similar attack lasting one month 3 years before. There was nothing

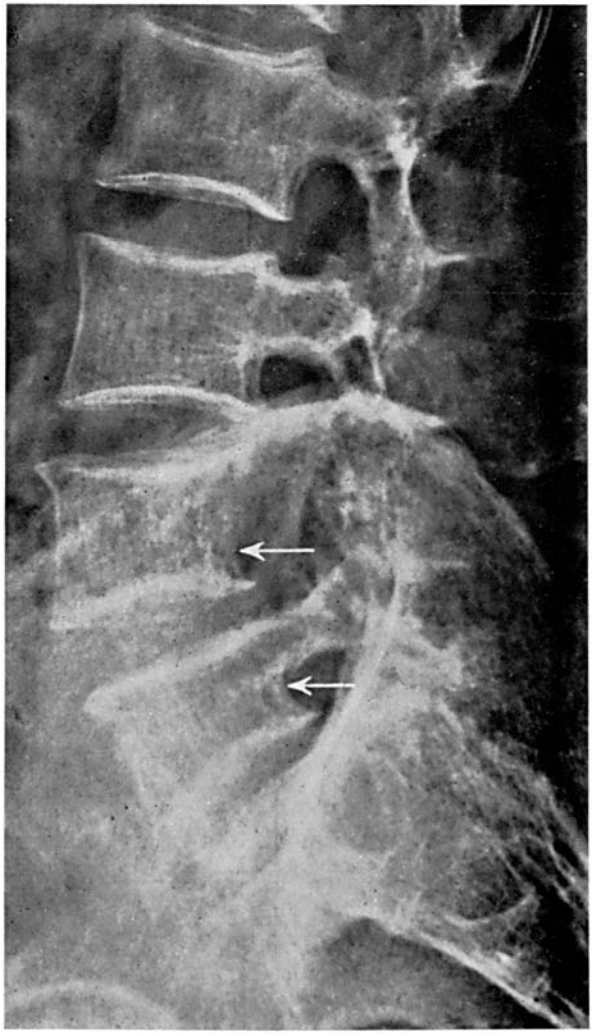

Fig. 2.
Lateral view of lumbar spine. Anterior part of arch of L.4 absent. Posterior boundaries of bodies of L.4 and L.5 show excavation.

further of interest in his past history or family history. On examination no abnormal physical signs were found. There were no developmental anomalies. The following tests gave normal results: red-cell count, haemoglobin concentration, total and differential leukocyte counts, and estimation of chemical and cellular constituents of the urine and cerebro-spinal fluid. The Wasserman reaction of the cerebro-spinal fluid

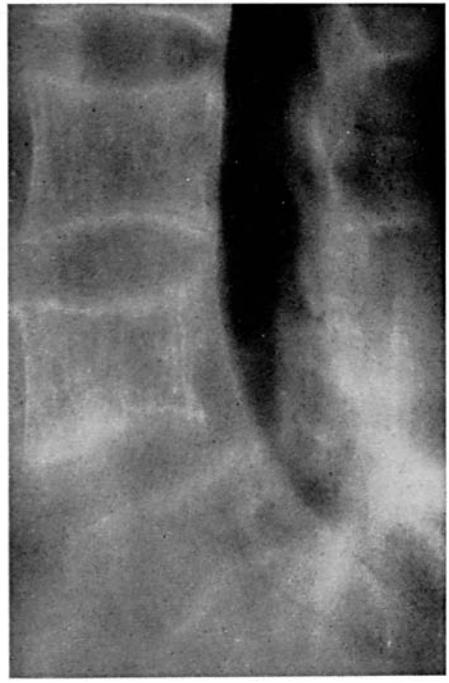

Fig. 3.
Oxygen myelography of lumbar spine. Tomogram. No expansive process within the dura to cause the skeletal changes.

was normal. The erythrocyte sedimentation rate (Westergren method) was 20 mm per hour. There was no melaena and no Bence-Jones protein in the urine.

Roentgen examination. In the frontal view of the lumbar region (Fig. 1) the right transverse processes of L.3 and L.5 were almost completely absent. Both transverse processes and pedicles of L.4 were missing. In the lateral view of the lumbar region (Fig. 2) the anterior part of the arch of L.4 was absent. The posterior edges of L.4 and L.5 showed excavation.

At oxygen myelography of the lumbar spine (Fig. 3) no intradural expansive process was found to explain the changes in the arch of L.4 and the posterior contours of L.4 and L.5. The bone-structure of the bodies of the affected vertebrae was coarse (Fig. 4). No pathological changes were evident in other parts of the skeleton (skull, sternum, thorax), but coarseness of bone-structures was present in the right iliac bone and on the right side of the sacrum (Fig. 5). It was note-

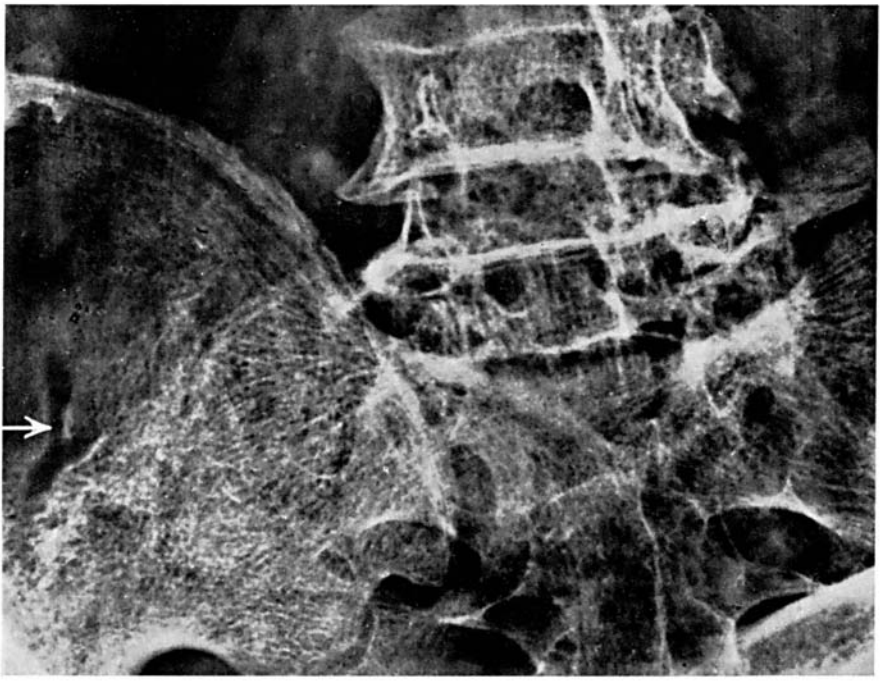

Fig. 4.
Note the coarse structure of the bodies of L.4 and L.5. The arrow indicates the wide nutrient canal of the right ilium.

worthy that the nutrient canal of the right ilium was much wider than that of the left ,(Fig. 5). The roentgen findings suggested atypical haemangioma.

Operative Findings. Laparotomy revealed almost total absence of the right transverse processes of L.4 and L.5. The right transverse process of L.3 was also eroded, only a thin shell of bony tissue being left. Nowhere could any pathological changes be found in the soft tissue surrounding the eroded vertebrae. The surgeon's opinion was that the pathological process seemed to be confined to the bone. The absence of haemorrhage was conspicuous. Biopsy specimens were taken from the transverse processes of all three vertebrae involved, from the lateral part of the right part of the fourth lumbar vertebra, and from adjacent soft tissue.

Histological examination. The biopsy specimens consisted of small pieces of cancellous bone, hyaline cartilage, ligamentous connective tissue, and minute bundles of striated muscle and islands of fat. The

Fig. 5.
Note coarse structure of the right side of the sacrum and right ilium. Nutrient canal of the latter is strikingly wide. The arrows indicate the nutrient canals of each side.

loose, fatty tissue of the marrow spaces of the cancellous bone contained many thin-walled cavernous structures forming an angiomatous structure (Fig. 7). The cavernous spaces were mostly empty, but a few contained inspissated serous material (Fig. 8). Isolated erythrocytes only were seen in the lumina. The angiomatous tissue was confined to the bone marrow and the immediately adjacent periosteal connective and fatty tissues (Fig. 6). In these last-mentioned parts the stroma between the angiomatous spaces was fairly densely fibrous, as in ordinary cavernous angiomata. The muscle bundles and their interstitial fatty tissue were not infiltrated by the angiomatous tissue. In a few places there was a suggestion of intimate connexion between the cavernous spaces and the larger branches of the nutrient vessels of the bone (Fig. 8). The vascular spaces showed no endothelial proliferation. There were extremely few smooth-muscle cells in the walls, and only isolated

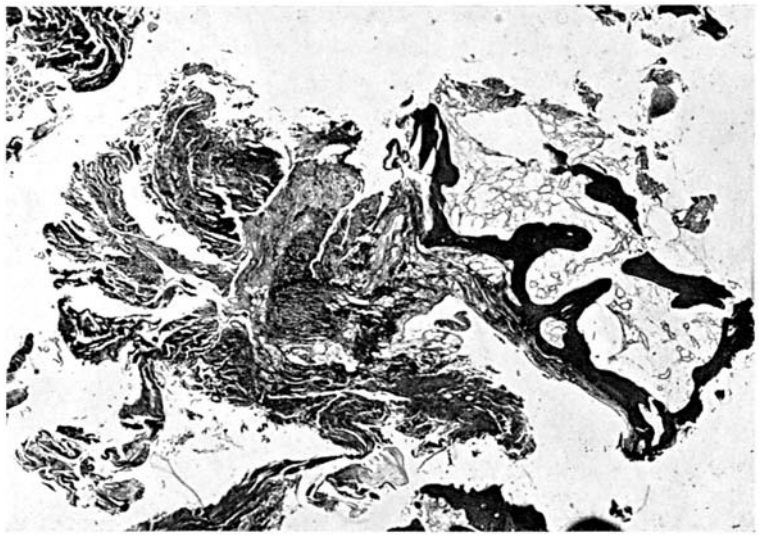

Fig. 6.

Low-power view of part of biopsy specimen from pedicles and transverse processes of L.3, L.4, and L.5 and adjacent tissue. The lymphangiomatous tissue is situated in the marrow spaces of the spongiosa (to the right) and the periosteal tissue, but does not invade the ligamentous connective tissue, fat, or skeletal muscle (centre and left). van Gieson stain (× 17).

elastic fibrils. The bone marrow was slightly infiltrated by granulocytes, plasma cells, and lymphocytes (Fig. 7). Very few haemopoietic cells were present.

No bacteriological specimen was taken at operation.

The postoperative course was uneventful. Roentgen therapy was tried, two courses of 6 × 300 r and 4 × 300 r being given. There was no symptomatic improvement. A corset was supplied, and the patient instructed to rest.

No improvement was evident at follow-up examinations 3, 9, and 12 months later, and roentgenograms showed the same picture as at operation.

DISCUSSION

The clinical, roentgenological, histological, and operative findings would make it seem probable that the bone-destruction in this case is pressure atrophy (Winkler, 1924) due to pressure on the bony tissue by the irregular, thin-walled vessels demonstrated in the marrow spaces and periosteum. These vessels are either completely empty or filled

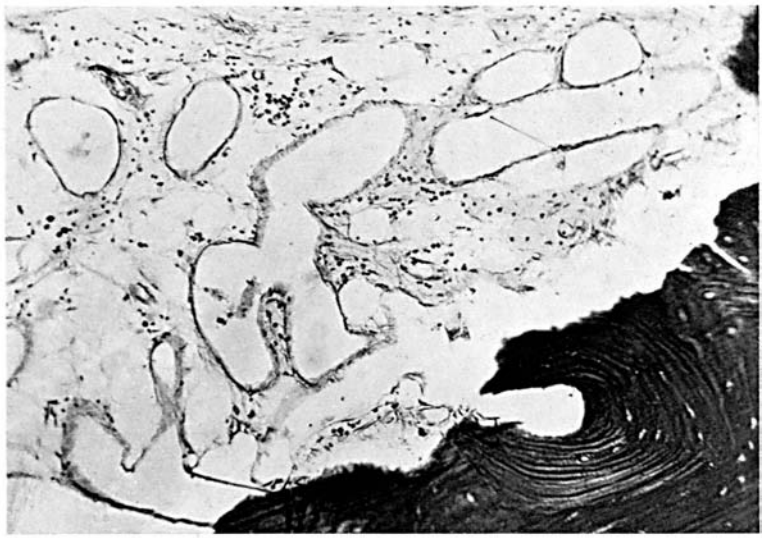

Fig. 7.

High-power view of the lymphangiomatous tissue in the bone marrow. The vascular spaces are empty, and are lined with flattened endothelium. There are practically no muscle cells in the thin walls. The loose, interstitial fatty tissue contains a few granulocytes and haemopoietic cells. The bony trabeculae (lower right) are normal. van Gieson stain (× 135).

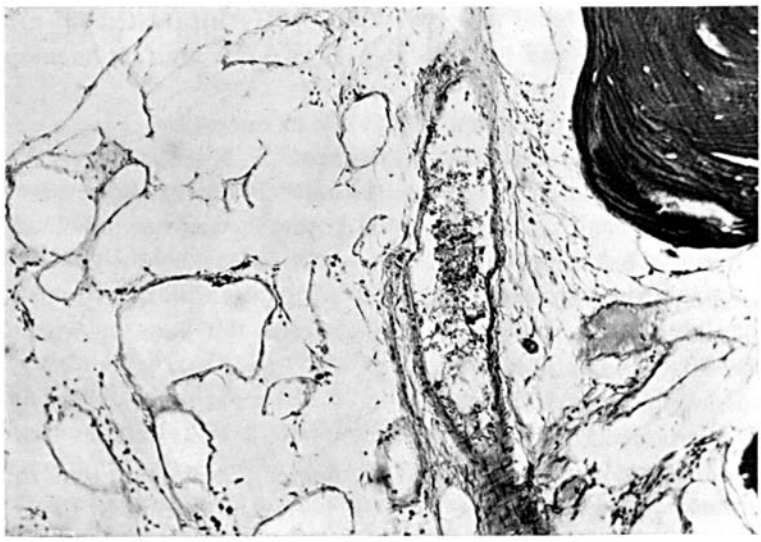

Fig. 8.

Another section of the marrow spaces showing the intimate relationship of the lymphangiomatous tissue to a blood vessel (centre right). A few of the angiomatous spaces contain inspissated serous material (lower right). van Gieson stain (× 135).

with an inspissated, serous fluid containing only isolated red cells. The walls of the vessels consist of thin endothelium, collagenous connective tissue with isolated elastic fibrils, and possibly also thin streaks of smooth muscle here and there. Thus the vessels are with great probability of lymphatic character.

In the normal bony skeleton true lymph vessels are found only in the periosteum. 'Lymph-sheaths' have been described both about the blood vessels of the haversian canals and at the boundaries between bone and periosteum and bone and bone marrow (Broman and Häggqvist, 1945). The actual marrow spaces do not seem to contain any lymphatic vessels (Custer, 1949). The presence of wide, thin-walled, irregularly formed lymph vessels in the marrow spaces would thus probably indicate a heterotopic malformation or true tumour formation. The vascular spaces can hardly be explained by stasis or inflammation. Except that firm, fibrous connective tissue is not present between the vascular spaces, the lymphatic structures fulfil remarkably well the criteria set out by Winkler (1924), Thomas (1942), and others, for cavernous lymphangioma. In addition, interstitial connective tissue is obvious in connexion with the angiomatous vascular spaces in the periosteal tissue.

It is a matter of debate whether cavernous lymphangioma consists of true tumour tissue or a malformation of the angio-hamartoma type (Parsons and Ebbs, 1940; Thomas, 1942; Willis, 1948; Jacobs and Kimmelstiel, 1953), and we do not wish to express an opinion in this paper. If in this case the lesion is to be interpreted as a congenital malformation, it is difficult to explain why the symptoms first appeared in late adult life. Most of the cases of lymphangiomatous tissue in bone quoted in the introductory paragraph were children or young adults. Wrede (1911) and Mandel (1927) maintain, however, that changes of this nature may remain latent for long periods, and therefore not give rise to symptoms until the patient has reached adulthood. They illustrate their theory with case histories. The localization of the lesion in the case we describe is the same as in 'fissurale Lymphangiome' (Winkler, 1924; Mandel, 1927).

To judge from the operation and histological findings, the cavernous lymphangiomatous tissue in this case was limited to the bone marrow and periosteal tissue. Thus in principle it showed the same distribution in the bony tissue as did the angiomatous formations in the cases described by Harris and Prandoni (1950) and Cohen and Craig (1955), which were considered to have arisen in the bone.

The presence of wide nutrient canals in the iliac bone and the suggestion of an intimate relationship between the lymphangiomatous tissue and the wide blood vessels (Fig. 8) would indicate that the lymphangiomatous tissue originated from the lymph vessels and lymph-sheaths that accompany the blood vessels (Cohen and Craig, 1955) of the bony tissue. Similar widening of the nutrient foramina was noted in one of Wrede's (1911) cases (the one diagnosed as secondary lymphangioma), as was mentioned by Herzog (1944).

To judge from the biopsy material the lymphangiomatous tissue in our case did not contain a haemangiomatous component, since the blood vessels encountered showed no such changes. Only small parts of the area affected, as shown on the roentgen films, were examined histologically, however, and it is possible that haemangiomatous structures were present elsewhere. This would seem to have occurred in Bickel's and Broder's (1947) case, to judge from a paper published by Gorham et al. (1954). Unfortunately no opportunities have been available for obtaining further biopsy specimens from other parts of the affected bone.

No satisfactory explanation is forthcoming for the moderate increase in erythrocyte sedimentation rate, either from the clinical findings or the biopsy material. It can hardly be connected with the slight inflammatory changes which were found in isolated patches of the bony tissue.

SUMMARY

A 55-year-old man with a history of backache was found on roentgen examination to have osteolytic changes in the lumbar spine, sacrum, and right ilium, with enlargement of the nutrient canal of the ilium. The changes partly appeared in the form of complete erosion of certain parts and partly as coarseness of structure in general. At operation no changes were seen in the tissues adjacent to the destroyed bone. Biopsy specimens from some of the diseased parts showed histologically many thin-walled vascular spaces apparently restricted to the bone marrow and periosteal tissue. The changes were compatible with a cavernous lymphangioma which was assumed to bear an intimate relationship to lymphatics accompanying the blood vessels of the bone.

RESUME

Chez un homme âgé de 55 ans ayant des douleurs dans le dos, on a trouvé à l'examen radiologique des modifications ostéolytiques de la

colonne lombaire, du sacrum et de l'ilion droit, avec élargissement du canal nourricier. Les modifications apparaissaient partiellement sous forme d'érosion complète, partiellement sous forme d'une structure en générale plus grossière. A l'opération on n'a constaté aucune altération des tissue adjacents à l'os endommagé.

La biopsie de spécimens de quelques-unes des parties malades a montré histologiquement beaucoup d'espaces vasculaires à parois très minces, apparemment limités de la moëlle osseuse et du tissue périostal. Les modifications ressemblaient à un angiome lymphatique caverneux destiné à assurer un rapport étroit avec les vaisseaux lymphatiques accompagnant les vaisseaux sanguins de l'os.

ZUSAMMENFASSUNG

Bei einem 55 Jahre alten Mann mit Rückenschmerzen wurde röntgenologisch osteolytische Veränderungen in der Lendenwirbelsäule, dem Kreuzbein und rechten Darmbein mit einer Erweiterung des foramen nutritium des Darmbeines gefunden. Die Veränderungen zeigten sich teilweise in Form von vollständigen Erosionen gewisser Teile, während andere Stellen eine Grobheit der Struktur im allgemeinen aufweisen. Bei der Operation konnten keinerlei Veränderungen der den zerstörten Knochen anliegenden Gewebe entdeckt werden. Gewebsproben von einigen der erkrankten Teile zeigten histologisch viele dünnwändige Gefässräume, die anscheinend nur auf das Knochenmark und das periostale Gewebe beschränkt waren. Diese Veränderungen waren vereinbar mit dem Bilde eines kavernösen Lymphangioms, das vermutlich eine enge Beziehung zu den Lymphgefässen, die die Blutgefässe des Knochens begleiten, hat.

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