MASSIVE OSTEOLYSIS

Report of Two Cases

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In 1838 and 1872 the history of an eighteen-year-old man with an unusual type of bone lesion was reported in the Boston Medical and Surgical Journal. The humerus disappeared completely in the course of eleven years and twice he sustained a spontaneous fracture of the bone. In spite of the abnormality he was able to do manual labour until his death at the age of seventy.

Since Jackson's account, many cases of this type have been reported in the literature under such titles as massive osteolysis, essential osteolysis, phantom bone, vanishing bone, and disappearing bone. The pathology was described by Gorham, Wright, Shultz and Maxon (1954), and since then the term Gorham's syndrome has also been used.

Massive osteolysis is uncommon. Twenty-four cases were traced by Gorham and Stout up to 1955. In 1961 Abell and Badgley mentioned that the total number of cases was thirty-three. It was stated to be forty-nine by Gorham and West (1964) and "about fifty" by Caulet, Fandre, Adnet, Coffin, Pennaforte and Mathey (1968).

This paper reviews the two patients with massive osteolysis observed for a long time. In 1958 these cases were demonstrated by T. A. A. van der Weijer.‡

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Case 1—A girl aged twelve fell on the ice and thereafter noticed some pain in the right thigh, which at first did not restrict her activity. Two months later the patient bumped against a table and fractured the upper third of the right femur (Fig. 1). Treatment by sustained traction was applied, but the fracture failed to unite. A biopsy specimen was obtained, and histological

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Case 1. Figure 2—Radiograph taken six months later showing osteolytic changes involving the proximal third of the right femur. Figure 3—Two months later the process has involved the proximal half of the femur and has extended into the pelvis.

Case 1. Figure 4—Photomicrograph of the biopsy specimen taken when the fracture had failed to unite despite adequate immobilisation by traction. Many thin-walled blood vessels, some filled with red blood cells, are seen. (Haematoxylin and eosin, ×90.) Figure 5—Photomicrograph of the second biopsy specimen taken eight months after the fracture. Bone trabeculae are very thin and the intertrabecular spaces are occupied by connective tissue containing numerous thin-walled vessels. (Haematoxylin and eosin, ×90.)

Case 1. Figure 6—The fatty marrow of the second biopsy specimen contains numerous small and large capillaries. (Haematoxylin and eosin, ×180.) Figure 7—Photomicrograph of the second specimen showing hyaline cartilage in the proximal part of the femur invaded by communicating sinusoids filled with red blood cells and surrounded by connective tissue. (Haematoxylin and eosin, ×90.)
examination showed many thin-walled blood vessels in the endosteal connective tissue. Some of the vessels were filled with red blood cells (Fig. 4). No sign of malignancy was noted.

The disease process progressed. After two months the proximal end of the right femur had disappeared and after four months the proximal third of the bone was affected (Fig. 2). During the next two months the process began to involve the right half of the pelvis (Fig. 3).

At exploratory operation only a cavity was found in place of the femur. The cavity contained serum and its wall consisted of thick fibrous tissue.

Histologically, bone tissue was scanty, the trabeculae were very thin and the intertrabecular spaces were occupied by connective tissue. Numerous thin-walled vessels containing a few red blood cells were present (Fig. 5). The fatty marrow also showed many small and larger capillaries (Fig. 6). The proximal part of the femur showed communicating sinusoids in the hyaline cartilage: these were filled with red blood cells and connective tissue (Fig. 7).

The results of laboratory investigations, including erythrocyte sedimentation rate, blood counts, Wassermann reaction, protein spectrum, serum calcium, phosphorus and alkaline phosphatase, and calcium balance, were normal. Neurological and endocrinological examinations as well as arteriography were negative.

Gradually the right thigh became swollen and fluctuant, though there was no pain and no pyrexia. At this time the right lower limb was greatly shortened: the limb was supported in a Thomas's splint.

When fourteen years old she suddenly developed symptoms of severe infection in the region of the bone lesion. The fluid aspirated from the osteolytic region contained staphylococci. The cavity was drained immediately and antibiotics were administered, but the patient died within forty-eight hours in severe septic shock.

Case 2—The patient in this case is now aged thirty-six years. At the age of ten years he suffered from vague discomfort in the left leg, and he sought medical advice four months later. He was then in good general condition. There was local swelling at the lateral side of the left leg. The head of the fibula felt soft but was not painful. Fluctuation was noted. There was no pyrexia and no increase of local warmth. Radiographs showed a large shadow in the soft tissues about the fibula, the proximal third of which had disappeared (Fig. 8). The laboratory findings were normal, as was neurological examination.

Six months later progression of the lesion had occurred. Two-thirds of the fibula had disappeared. The area was explored and a cavity was found in place of the fibula. The cavity contained clear fluid and its wall consisted of thick fibrous tissue.

Histologically the bone had been replaced by fibrous tissue and the number of bone trabeculae was greatly reduced (Fig. 13). In some regions of the fibrous tissue thin-walled vessels were present (Fig. 14).

Progress—In the next year the proximal end of the tibia showed some radiological changes, and three months later it fractured spontaneously. After six months this fracture had united, but the greater part of the fibula had disappeared (Fig. 9). During the next two years laboratory findings were normal and there was no pain.

Twelve years later shortening of the left leg measured nine centimetres. There was pronounced atrophy of the muscles. The patient was able to get about in a caliper and had
Case 2. Figure 9—Antero-posterior and lateral radiographs twenty-one months later. The tibia is now involved and the pathological fracture of the tibia that had occurred six months previously has united. The greater part of the fibula has now disappeared. Figure 10—Ten years later the process has not extended in the left tibia, but the fibula has been further eroded.

Case 2. Figure 11—Antero-posterior and lateral radiographs of the right tibia twelve years after the left leg had been affected. The right tibia has a well delineated bone defect in the middle third with cortical thickening around it. Figure 12—Ten years later there had been no progress in the lesion in the right tibia.
little trouble with the left leg, but at this time a small painless swelling was noted on the right tibia. Radiographs showed a well delineated bone defect with thickening of the cortex in the middle third of the tibia (Figs. 10 and 11).

![Image of radiograph showing bone defect](image)

**Fig. 13**
Case 2—Photomicrograph of the lesion of the left fibula from the biopsy specimen taken six months after the onset of the condition when the patient was 10 years old. The bone had been replaced by connective tissue. (Haematoxylin and eosin, × 80.)

![Image of photomicrograph showing fibrous tissue](image)

**Fig. 14**
Case 2. Figure 14—In some areas the fibrous tissue contained thin-walled blood vessels. (Haematoxylin and eosin, × 210.) Figure 15—Photomicrograph of the lesion in the right tibia which occurred ten years later. Biopsy revealed a fibrous marrow with thin-walled capillaries and some osteoclasts. (Haematoxylin and eosin, × 210.)

This part of the right tibia was explored and the bone defect completely excised. Microscopically the tissue showed a fibrous marrow with some thin-walled capillaries and some osteoclasts (Fig. 15).
The disease process in the right tibia did not progress in the subsequent ten years, and at present the patient is free of complaints (Figs. 12, 16 and 17).

DISCUSSION

Leger, Ducroquet and Leger (1949) mentioned three types of osteolytic diseases: 1) osteolysis associated with an infection such as osteomyelitis or with rheumatoid arthritis; 2) osteolysis associated with disease of the central nervous system, such as tabes dorsalis, syringomyelia, leprosy or myelodysplasia; and 3) osteolysis "dite essentielle", characterised by spontaneous disappearance of a part of the skeleton without concomitant symptoms. This third form of osteolysis with complete, spontaneous and progressive disappearance of bone was called massive osteolysis by Gorham and Stout (1955). They differentiated it from atrophy of bone due to inactivity, from the acute inflammatory atrophy associated with trauma described by Sudeck, and from lytic processes in bone due to tumours, hyperparathyroidism or other metabolic diseases.

According to Gorham and Stout the most characteristic histological abnormality in massive osteolysis is the change of bone into connective tissue. This connective tissue contains many thin-walled vessels, sometimes with red blood cells. In other areas freely anastomosing vascular spaces lined by endothelial cells can be seen. The fatty marrow also contains some dilated blood vessels.

This process does not resemble ordinary haemangioma of bone, which remains localised and which, though it may destroy bone, never "dissolves" it completely. The process resembles the proliferation of capillaries observed in the soft tissue of congenital arteriovenous fistulae. The term used by Gorham and Stout was "haemangiomatosis", implying a proliferative process. According to these authors massive osteolysis is not the result of osteoclasis, but of active hyperaemia, mechanical forces, slight changes in pH and other unknown causes. Gorham (1960) proved experimentally the importance of active hyperaemia in the development of massive osteolysis.
Butler, McCance and Barrett (1958), and Milner and Baker (1958) found osteoclasts in cases of massive osteolysis. Milner and Baker cited the first publication of Gorham in which it was stated: "Histologically much of the cortical and cancellous bone had disappeared and that which remained was surrounded by osteoclasts". Gorham responded in an article in 1964: "Milner and Baker believed that osteoclasts are of primary etiological importance in massive osteolysis. Most observers, however, share our experience that osteoclastosis is rarely associated with massive osteolysis. There are two possible reasons why osteoclasts may or may not be found in any given case of osteolysis: Follis (1952) believed that osteoblasts destroy bone as readily as osteoclasts; Hancox (1949) stated that the life span of osteoclasts is only about twenty-four hours and that the greater part of their functional activity occurs in the first eight hours. Unless biopsies of human bone are taken during the relatively short period of their functional activity, osteoclasts might be rarely observed."

Johnson and McClure (1958) distinguished two stages in the process of massive osteolysis. The proliferation of capillaries in the endosteal connective tissue is characteristic of the first stage. In the later stage the bone tissue is replaced by fibrous tissue although complete avascularity is rare. These authors reported on the relative frequency of massive osteolysis in different bones. The sequence is as follows: clavicle, scapula, proximal end of humerus, ribs, iliac bone, ischium, sacrum. The disease has not been observed in the calvarium or in the distal bones of the extremities. The monostotic form occurs more frequently than the polyostotic.

In general the patient is young and both sexes are equally affected. The process is painless: it starts suddenly, progress is rapid and finally the bone is replaced by a thin layer of fibrous tissue surrounding a cavity. Despite this, the function of the extremity is good. Laboratory findings are normal.

According to the usual clinical and histological criteria the cases of our two patients are typical of massive osteolysis. In the second case the fibrotic form of the disease was dominant. After twelve years a lytic process began in the opposite tibia. Microscopically this second lesion showed a very fibrous marrow, which contained some thin-walled capillaries, but these were less numerous than in the original lesion. Some osteoclasts were observed. In the literature no case was mentioned in which a second lesion developed in another bone after a period of standstill.

Reviewing the literature of massive osteolysis for the period 1965–68 we found eight reported cases (Hadjidekow, Airanow and Welitschkow 1965; Knolle and Meyer 1965; Stern 1965; Louyot, Gaucher, Benoit and Combebias 1966; Kyllonen 1967; Schneider and Schimke 1967; Tucker 1967; Caulet et al. 1968). There were two reports of essential osteolysis with nephropathia (Lagier and Rutishauer 1965; Torg and Steel 1968).

SUMMARY

Two cases of massive osteolysis of the lower extremities are reported. In one case a second lesion occurred in the opposite tibia and the process stopped after early total resection.

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