MASSIVE OSTEOLYSIS

Report of a Case

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There are at least four separate and unrelated conditions implied by the word “osteolysis”. The first, idiopathic hereditary osteolysis, affects carpal and tarsal bones, presents during childhood, and is inherited as an autosomal dominant disorder. There is no renal disease, no histological evidence of angiomatosis, and the osteolytic process does not pose a threat to life (Torg and Steel 1968).

The second, idiopathic osteolysis with nephropathy, affects the carpal, tarsal and adjacent tubular bones of young children who develop hypertension and azotaemia and die in early adult life. There is no histological evidence of angiomatosis and the disease is apparently not inherited (Torg and Steel 1968).

The third, haemangiomatosis with massive osteolysis (Fornasier 1970), is an extensive congenital proliferation of blood vessels affecting skin, subcutaneous tissues and muscles as well as bone. An entire limb may be involved and the limb bones may disappear.

The fourth, known variously as massive osteolysis, disappearing or vanishing bone disease, phantom bone and Gorham’s disease, is characterised by complete destruction of all or part of a bone by angiomatous tissue. The process may spread insidiously, like a locally aggressive tumour, involving adjacent bones and soft tissues, and may cause death by encroaching on vital structures. On the other hand, the growth of the lesion may become arrested spontaneously. It is not inherited and is not congenital. The following case is an example of this fourth entity.

CASE REPORT

A woman machinist aged thirty-six years, who had been in good health, presented in September 1965 with pain in the left foot first noticed while dancing. Plain radiographs of the foot revealed no bone or joint abnormality. As the pain persisted the patient was referred to a rheumatologist, who noted tenderness and slight swelling over the left second, third and fourth metatarsal bones. Clinical examination was otherwise normal. There were no angiomata in the skin, and there was no family history of bone disease, rheumatoid arthritis or tuberculosis. Although a further radiograph six months after the onset of symptoms was reported as normal (Fig. 1), the foot was placed in plaster for four weeks. On removal of the plaster, the discomfort had disappeared.

Three months later, ten months after the onset of symptoms, she returned to her doctor, complaining that she could not bear to put her left foot to the ground without a firm bandage. A further radiograph revealed a surprising change (Fig. 2). There was now considerable destruction of the shafts of the second and third metatarsal bones, with a pathological fracture of the latter. The destruction of bone was more pronounced in the cortex, the shafts were narrowed, and there was complete lack of periosteal new bone formation, suggesting a diagnosis of massive osteolysis. Estimations of blood urea nitrogen, creatinine clearance, serum calcium, phosphate, alkaline phosphatase, as well as the erythrocyte sedimentation rate, blood picture and serum agglutinins were all normal. Biopsy was done, and at operation

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Figure 1—A radiograph of the left foot six months after the onset of symptoms. At first it was thought to be normal, but there is slight scalloping of the surface of the shaft of the second metatarsal bone with some slight reduction in the width of the shafts of both the second and third metatarsal bones. Figure 2—The same foot ten months after onset of symptoms. Marked changes have occurred since the previous radiograph. The shafts of the second and third metatarsal bones are narrowed, tapered and osteoporotic, and there is a pathological fracture of the shaft of the third metatarsal bone without any callus or periosteal new bone formation.

Figure 3—Arteriograph of the foot a year after the onset of symptoms, sixteen seconds after injection of contrast medium into left femoral artery. Contrast medium persists in the metatarsal arteries, but no pathological vessels are demonstrated. The shaft of the third metatarsal bone tapers to a point, somewhat resembling a sharpened lead pencil. Figure 4—A radiograph a year and a half after the onset of symptoms. There has been almost complete resorption of the second and third metatarsal bones, although the heads remain. The shaft of the fourth metatarsal bone is thinned and is beginning to taper, and the intermediate and lateral cuneiform bones are also disappearing. Figure 5—The left foot twenty-seven months after the onset of symptoms, just before amputation. Further destruction of the fourth metatarsal and intermediate and lateral cuneiform bones has occurred, and the shaft of the first metatarsal bone is narrowed. The first tarso-metatarsal joint is now dislocated and there is disuse osteoporosis of the tarsal bones. Only vague outlines of the heads of the second and third metatarsal bones remain.
the subcutaneous and periosseous tissues of the left foot were found to be oedematous; the shafts of the second and third metatarsal bones were lying free, like sequestra, in this tissue. Cultures of the bone for tuberculosis were sterile.

Histology—Two small fragments were submitted for histological examination, which showed fibrovascular tissue with two types of vessels. Most were large and cavernous, measuring 140 microns or more in diameter, with thin walls and lined by a single layer of endothelial cells; these were either empty or contained very occasional cells and proteinaceous material resembling lymph. The other type of vessel had muscular walls up to 30 microns in thickness, but the lumina were small, being rarely larger than 14 microns in diameter. The vessels were surrounded by normal fatty marrow and fibrous tissue, through which were scattered a few chronic inflammatory cells. The abnormal fibrovascular tissue extended out from the bone into the adjacent soft tissues, where it infiltrated voluntary muscle fibres, some of which had undergone degenerative changes. In the soft tissues, the small thick-walled vessels were more prominent than cavernous spaces. In the affected bone, the residual trabeculae were small and thin, and a few had irregular moth-eaten outlines. There were virtually no osteoclasts. The histological appearances were interpreted as being diagnostic of massive osteolysis (Figs. 6 and 7).

Progress and further treatment—After operation the foot was placed in plaster. Three months later, in an attempt to determine the extent and nature of the process more accurately, left
DISCUSSION

Clinical features—As massive osteolysis occurs predominantly in children or young adults, the onset of symptoms at the age of thirty-six years is atypical. However, isolated cases have been reported as late as 58 years (Gorham and Stout 1955). There has been no significant sex predilection in reported cases, nor has any evidence of genetic transmission become obvious. Localised pain, which is sometimes caused by a pathological fracture, is the usual presenting symptom, and a history of a mild injury is often obtainable.

On physical examination the involved area is tender; systemic disturbances are not observed. In our case the patient was able to continue working and to walk without severe pain for many months despite extensive bone destruction. Some soft-tissue swelling may be present early, but there is no evidence of inflammation and the overlying skin remains normal throughout. There have been no reports of a flow bruit being heard on auscultation.

Despite extensive bone resorption, the serum calcium and alkaline phosphatase are always normal, indicating a lack of osteoclastic activity in the underlying process. The localised nature of the disease is supported by a normal erythrocyte sedimentation rate and normal blood picture. In none of the reported cases has any biochemical or endocrine abnormality been detected.
Massive osteolysis has been described arising in almost every bone except the cranial vault. There is some propensity for involvement of the shoulder girdle and pelvis, but no distribution can be regarded as typical. While the process frequently involves more than one contiguous bone, “skip” lesions, multiple foci or metastases do not occur (Johnson and McClure 1958). Kery and Wouters (1970) described one case of massive osteolysis of the left fibula and left tibia. This focus spontaneously ceased growing over a twelve-year period. The patient then developed a small painless bone defect in the right tibia. This lesion was excised and had not recurred ten years later. In this case the published radiographs and photomicrographs of the lesion from the left fibula leave no doubt that the patient had massive osteolysis in the left leg, but in our opinion the evidence for massive osteolysis of the right tibia is unconvincing.

**Radiological features**—Johnson and McClure (1958) reviewed the radiological features of thirty-two cases reported up to that time. They depicted the progression of the disease as an intraosseous process, followed by extraosseous spread. In the present case the observed radiological changes suggest that the disease began near the periphery of the bones, either in the periosteum or in the adjacent soft tissues, and then spread deep into the cortex and medulla. Thus, in Figure 1, there is a suggestion of slight scalloping of the external surface of the shaft of the second metatarsal bone, with a slight reduction in the width of the shaft of the second and third metatarsals bones. At that early stage there was no evidence of intraosseous lysis. The early involvement of two contiguous bones would point to an extraosseous process involving adjacent bones, rather than a multifocal intraosseous process in two adjacent bones.

The gradual tapering and progressive resorption of cortical bone is one of the most characteristic radiological features of the disease (Johnson and McClure 1958). The two persisting bone surfaces come together like the point of a sharpened lead pencil (Fig. 3). Intramedullary or subcortical lytic foci may be overshadowed by the predominantly extraosseous resorption of the involved bones. These intraosseous changes could be caused by nonspecific patchy local osteoporosis associated with disuse. Certainly, severe osteoporosis has been a feature of the late stage in most cases described. Whether initially an extraosseous or intraosseous process, the purity of the osteolysis, with complete lack of new bone formation even under the stimulus of a pathological fracture (Fig. 2), is a characteristic feature. Massive osteolysis sometimes remains confined to one bone but usually it progresses to involve contiguous bones. However, the bones initially involved may not completely disappear, as illustrated by the persisting remnants of the heads of the second and third metatarsals in our case (Fig. 5).

Both arteriography and venography have been used in an attempt to demonstrate the lesion, but unlike the findings in angiomatous malformations, arteriovenous fistulae and vascular tumours, in which the abnormal vessels are readily seen, it is not usually possible to show any connection between the abnormal tissue and the vascular circulation. The absence of phleboliths, coarse trabeculation and vascular or soft-tissue calcification, characteristic of angiomata, are also important negative findings. Hambach, Pujman and Maly (1958) produced a homogeneous and almost complete outline of a totally resorbed scapula by direct injection of contrast medium into the tissue, and this seems to be the best method of demonstration. Our findings confirm the opinion that the abnormal tissue of massive osteolysis had no direct communication with the vascular circulation.

**Pathology**—Gorham and Stout (1955) have provided the most detailed description of the histopathology of massive osteolysis. They suggested that the process was similar to haemangiomatosis (diffuse haemangiomatosis), a congenital condition in which the hyperplastic vascular components form arteriovenous fistulae. Lymphatics are not involved, and the abnormal vessels infiltrate skin, subcutaneous tissues, underlying deep soft tissues and bones. Whereas bones are often destroyed by diffuse haemangiomatosis, the bones and soft tissues may become
hypertrophied, resulting in a giant limb. In addition, the arteriovenous fistulae may cause cardiac hypertrophy and even cardiac failure. The present case seems to be quite distinct from diffuse haemangiomatosis. It was not congenital, there were no arteriovenous communications demonstrated radiologically and the skin and subcutaneous tissues were normal. Histologically, the sparsity of erythrocytes within the vessels’ lumina, and the lack of big muscular arteries and large thick-walled venous spaces, also indicated that the disease differed from diffuse haemangiomatosis.

Judging from this case and from others previously reported, massive osteolysis is probably best regarded as a locally aggressive and destructive tumour of vessels, with a deceptively bland histological appearance. As stabilisation occurred after sixteen years of slow progression in one case (Gorham, Wright, Shultz and Maxon 1954), it seems that unless the process involves essential parts of the bony skeleton such as the thoracic cage, eventual arrest may be expected. In this respect, massive osteolysis is analagous to aggressive fibromatosis (desmoid tumour) in that it does not metastasise and may cease growing spontaneously, but it may cause death by encroaching upon adjacent vital structures.

Treatment—Determination of the best method of treatment is difficult. Surgical treatment has consisted in amputation and local resection, with or without replacement prostheses or bone grafts. The results have been variable. Sympathectomy has been done without affecting the disease process. Results obtained with radiation therapy have been equivocal. Johnson and McClure (1958) noted arrest of the disease after two courses of radiotherapy, and from a review of the literature they concluded that it is reasonable to use ionising radiation. Operation, they thought, should be reserved for cases not responding to radiotherapy, and for the correction of deformity following spontaneous or radiation-induced arrest. In the present case operation was performed because of increasing local symptoms, and for fear that the disease would spread proximally up the leg. The patient now appears to be cured.

SUMMARY

1. A case of massive osteolysis of the bones and soft tissues of the left foot is reported.
2. Arteriography revealed slowed circulation in the foot, but the tumour vessels did not fill with contrast medium.
3. Two years after below-knee amputation of the left leg and four years after the onset of symptoms, the patient appeared to be free from disease.

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REFERENCES


