INTRAOSSEOUS LIPOMATOSIS

A CASE REPORT

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We report a case of systemic intraosseous lipomatosis involving the proximal femur, both ends of the tibia, and the tarsal and metatarsal bones. The lesions progressed during a five-year follow-up with a pathological fracture of the tibial plateau. CT scans were characteristic and helped in diagnosis but MR imaging added little information.

Intraosseous lipomatosis is a hamartomatous malformation due to hyperplasia of adipose tissue, and is fundamentally different from solitary benign intraosseous lipoma. Management involves reconstruction of any pathological fracture. Large progressive lesions should be treated by curettage and grafting in an attempt to prevent such fractures.

Intraosseous lipoma is a rare benign bone tumour, about 130 cases having been recorded (Döhler and Harms 1981; Lemerle, Gaulier and Zucman 1985; Hall, Cohen and Grumbach 1986; Buckley and Burkus 1988; Milgram 1988). Intraosseous lipomatosis is even more rare (Döhler et al 1982), and one case has been described in which it was associated with hyperlipidaemia (Freiberg et al 1974).

We report a patient who presented with slowly progressive systemic intraosseous lipomatosis. The disease involved the leg and foot bones producing pain and pathological fractures, but there was no associated disorder of lipid metabolism.

CASE REPORT

A 28-year-old man, an active sportsman and footballer, had a fracture of the lateral condyle of the right femur caused by minor trauma. This was managed surgically at another hospital by reduction and fixation with Kirschner wires. Radiographs at this time also showed ill-defined

radiolucent lesions in the distal femoral and the proximal tibial condyles of both legs.

Four years later he sustained a fracture of the lateral plateau of the left tibia in a fall from a bicycle. This was treated at Tatabánya Hospital by reduction of the plateau fracture, curettage of the underlying cystic area and bone grafting, with fixation by two cancellous screws. Because of the tumour-like appearance of the cystic lesions, the patient was referred to Semmelweis University Medical School in Budapest.

The patient, now 33 years of age, was a wine-grower, reported by one of his relatives to drink half to one litre of wine per day. He showed no evidence of alcoholic dependence or cirrhosis. He complained of some pain in both knees and both ankles on starting weight-bearing and at extremes of movement. These joints appeared to be enlarged and tender. In addition, he had moderate limitation of subtalar movement. Both knees showed 10° of valgus instability during walking. No neurological abnormalities were detected.

Laboratory examination gave normal values for haemoglobin, haematocrit, white and red cell counts, urine, hepatic and renal function, total serum protein, serum calcium and phosphorus, blood sugar, urinary creatinine, calcium and phosphorus. The serum parathormone and the 25-OHD₃ levels were also normal. There was slight elevation of the erythrocyte sedimentation rate and the serum alkaline phosphatase level. A bone marrow smear from a sternal puncture was normal.

Normal lipid metabolism was confirmed by measurement of the total serum lipid, serum cholesterol, HDL-cholesterol, and serum triglyceride, and by serum lipid electrophoresis.

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Fig. 1

Fig. 2a

Fig. 2b

Figure 1 – Ill-defined radiolucent lytic lesions in the right femoral and tibial bone ends. Figure 2a – Compression fracture of the left tibial plateau into a lytic lesion. Figure 2b – Tibial plateau reconstruction using two screws. Six months after curettage and grafting there appears to be good incorporation.

Fig. 3a

Fig. 3b

Fig. 3c

Fig. 3d

Figures 3a and 3b – Radiographs showing cystic lesions in the malleoli, talus and other tarsal bones. Figures 3c and 3d – Six months after curettage and grafting of the malleoli.

The first radiographs of the patient's long bones, taken in 1984, showed ill-defined irregular radiolucent lesions, mainly in the proximal tibiae, but also in the distal femora. Four years later, the lytic areas in the right femoral condyles were larger and more pronounced (Fig. 1). Some thickening of the weight-bearing trabeculae was visible within the lytic areas, and it was obvious that the compression fracture of the left lateral tibial plateau had occurred into a weakened radiolucent area (Fig. 2a). After homologous grafting, healing at six months was satisfactory (Fig. 2b).

Radiographs of the ankles and feet showed marked changes in the bony framework. The talus and tarsal bones were atrophic, with many cystic areas (Fig. 3). There was cortical thinning with endosteal scalloping of both malleoli; the bones appeared to be expanded around

the lytic and cystic areas. Similar radiolucent lesions were visible in the cuboid, the metatarsal heads and the bases of the phalanges. Scintigraphy showed normal activity in all the affected bone.

CT scans confirmed the presence of numerous fat-density lesions (Figs 4 and 5a). In some areas, as at the ankle, the cortex was thinned and the bone expanded, but elsewhere, as in the proximal tibial metaphysis, the lipomatous area appeared to be surrounded by a reactive sclerotic margin of rearranged and coarsened trabeculae. MRI added nothing to the CT diagnosis, since signal intensities from the lipomatous areas on weighted T_1 and T_2 images differed little from those of normal bone marrow (Fig. 5b).

At operation, the cortex of affected areas was unusually thin, but hard and sclerotic. In each lesion,

Fig. 4

Fig. 5a

Fig. 5b

Figure 4 – CT scan through the proximal tibia. There is a large lesion with an ill-defined but sclerotic margin. Figure 5a – CT scan of the femoral condyles and patella, showing irregular lytic lesions of fat density. Figure 5b – MR image, which provides less differentiation and is less useful.

Fig. 6

Figure 6 – Histology of the curetted material shows mature normal adipose tissue (haematoxylin and eosin $\times$ 250).

cancellous trabeculae had been replaced by a mass of regular yellow adipose tissue, with no connective tissue capsule. The cavities opened into the diaphyseal medullary cavity, with no osseous wall or connective tissue septum.

Microscopically, curetted material showed unencapsulated, abundantly vascularised, normal adipose tissue (Fig. 6), with the sporadic presence of bony spicules. In smaller foci, mainly around vessels and in connective tissue septa, there were areas of mild, chronic inflammatory cell infiltration.

Our management of the patient started after the grafting of the fracture of the left tibial plateau. Conservative treatment with calcium, vitamin-D and a series of three courses of calcitonin had produced no clinical or radiographic changes after one year, and it seemed that there was a danger of further pathological fractures. The satisfactory healing after grafting of the left proximal tibia led us to advise surgery on the ankle.

Both malleoli were explored, curetting the cavities and filling them with cancellous bone grafts. At one year the grafts appear to be well-incorporated (Figs 3c and 3d).

DISCUSSION

Intraosseous lipomatosis is a disease of unknown origin. Freiberg et al (1974) describe a case of 'multiple intraosseous lipomas' associated with type IV hyperlipoproteinaemia. Others have recorded the coincidence of hyperlipoproteinaemia with intraosseous xanthomatosis (Inserra et al 1984; Yokoyama, Shinohara and Wada 1988). However, our case suggests that intraosseous lipomatosis may develop in an otherwise fit patient, unassociated with abnormal lipid metabolism. We could discover no aetiological factors and no evidence that the fact that the patient was an habitual though not excessive alcohol consumer was related to the lesions.

Intraosseous lipomatosis is not synonymous with or equivalent to multiple lipomas within bone. An intraosseous lipoma is usually found in the metaphysis of a long bone, or sometimes in the calcaneus. It appears as a sharply demarcated lytic lesion, occasionally with sclerotic margins and rarely shows any central calcification. The bone may appear to be expanded with a thin cortex but no periosteal reaction. Clinical symptoms are usually absent, and most cases are recorded as secondary findings (Goldman et al 1984; Manitz and Lossnitzer 1985; Ramos et al 1985; Reig-Boix et al 1987).

Systemic intraosseous lipomatosis usually affects long bone metaphyses, and sometimes the tarsal and metatarsal bones. It is seen mainly in middle-aged patients and shows slow progression. Multiple ill-defined radiolucent lesions appear to be confluent and are continuous with the diaphyseal medullary cavity. Other margins, formed by weight-bearing trabeculae, may be sclerotic, but the adipose tissue, unlike that of a solitary intraosseous lipoma, is not encapsulated by connective tissue.

The appearance is that of hyperplasia of medullary

adipose tissue, with secondary resorption of bony trabeculae. The compression produced by excessive adipose tissue causes remodelling of the bone; it is expanded and the cortex is thinned with endosteal scalloping.

Intraosseous lipomatosis, unlike lipoma of bone, presents with ill-defined episodes of bone pain and pathological fractures caused by minor trauma. It leads sooner or later to severe secondary destruction of the involved joints.

CT scans provide a better diagnosis than plain radiographs, giving greater accuracy in defining the site and extent of the disease, and in showing the presence of adipose tissue through its characteristic density. It also shows the cross-sectional appearance of the coarsened

and reorganised bone trabeculae surrounding the lesions. By contrast, magnetic resonance imaging contributes little to the diagnosis, because it provides very similar signals from normal medullary fat and from the adipose tissue of the lesion.

The management of intraosseous lipomatosis is an unsolved problem. There is no prospect of chemotherapeutic control, so all that can be done is to treat pathological fractures by precise reconstruction, and to curette and graft those lesions which appear to provide such a hazard.

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