SYSTEMIC LIPOMATOSIS OF BONE

A CASE REPORT


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We report the case history of a woman aged 32 years on whom we performed curettage of intra-osseous lipomata of both talar bones. Radiographs showed multiple osteolysis of the extremities with predilection for the hands and feet. The radiological and pathological characteristics of the intra-osseous lipomata and of comparable disturbances of mesenchymal bone structures are discussed. The pathogenesis of the syndrome presented here is likely to be a multicentric hamartoma.

Intra-osseous lipomata are very rare tumours in bone. A report of 47 patients (Döhler and Harms 1981) showed that such tumours often occur in the metaphyses of the long bones and in the os calcis. In the literature we found no reports of an affected talus, bilateral or polytopic localisation, coincident cysts or other bone tumours. Salzer and Salzer-Kuntschik (1965) discuss a different pathogenesis of intra-osseous lipomata involving real neoplasms and primary or secondary hyperplasia of fat cells.

The macrodystrophia lipomatosa is a rare form of localised gigantism. This congenital, usually unilateral anomaly is characterised by a marked increase in the growth rate of mesenchymal elements, especially of adipose tissue. Irregularly thickened and lengthened phalanges of the hands and feet, sometimes with severe articular deformity, are important radiographic features of the syndrome (Goldman and Kaye 1977). Three cases of cystic angiomatosis of the bone (one with extraskeletal lymphangioma) were reported by Schajowicz et al. (1978). The age of the patients ranged from 7 to 11 years; lesions of the hands and feet were not found and spontaneous regression of the osteolysis occurred in two patients. The authors assumed that a multicentric congenital malformation (for example a vascular hamartoma of bone and soft tissue) was the cause. In our patient similar skeletal lesions, suggestive of a multicentric hamartoma, were revealed after radiographic examination. However, no multicentric intra-osseous lipomata have been reported until now and the age of our patient is unusual for symptomatic hamartoma. The prognosis of the peripheral osteolysis remains doubtful.

CASE HISTORY

In 1976 the patient presented with increasing pain in the left ankle. There was slight swelling due to synovitis, but no restriction of movement. Radiographic examination showed large cysts in each talus and smaller cysts in the adjacent bones (Figs 1 to 4). At operation, the articular surface of the left talus was found to be intact but the neck of this bone was soft. After fenestration a haematinic and gelatinous substance was found in the talus. There was no fluid or hyaline matrix. Curettage and autologous bone-grafting were performed. The histological examination revealed an intra-osseous lipoma (Figs 5 and 6). A patellar-tendon-bearing brace ensured that the foot took no strain for six months.

Fig. 1
Radiographs of the left ankle taken at the first examination in 1976. Figure 1—Lateral view: multicystic destruction of the talus. Sharply defined, but no sclerotic margins. Irregularly thick trabeculae. No ballooning of the bone. Figure 2—Anteroposterior view: there is a cystic lesion in the fibula. The lateral cortex is thinner than normal.

Fig. 2

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Radiographs of the right ankle taken in 1976. Figure 3—Lateral view: findings are similar to those found in the left talus. Note the radiolucent zone in the anterior portion of the calcaneus, the tibia and in the cuboid bone. Figure 4—Anteroposterior view; large subcortical cyst of the fibula. The patchy deossification of the distal fibula is similar to that seen in the right humerus.

Histological sections of the curetted tumour of the left talus: intra-osseous lipoma. Figure 5—Abundant univacuolar adipose tissue. Fibrosis and many capillaries can be seen. Figure 6—Margin of the tumour; two sections of blood vessels with thick walls. Vessel walls of such thickness are not found in normal fatty marrow. (Haematoxylin and eosin, ×97.)

Radiographs of the right ankle. Follow-up three years after curettage and autologous bone-grafting of the talus. Figure 7—Anteroposterior view; intact talus, increased size of the fibular cyst. A radiolucent zone in the styloid process of the tibia is obvious. Figure 8—Lateral view; osteolysis of the cuboid and the os calcis. Figure 9—Tarsometatarsal bones and forefoot. Oblique view; disseminated osteolysis of the foot. No rims or ballooning. Extremely thin cortex of the fifth metatarsal and of the phalanges of the second and fifth toes.
In 1977 the same clinical signs appeared in the right talus. The treatment and the histological features were identical to those on the left. Follow-up at three years revealed that the ankles were functioning properly. The patient did not complain of any pain and there were no signs of hereditary disease. No tumours of the soft tissues and no visceral symptoms were found. The radiographs taken of the hands the previous year by the orthopaedic surgeon to whom the patient first presented showed cystic bone defects in both hands. Since there was a possibility of pathological fractures further investigations were undertaken. Nuclide bone imaging of the skeleton showed an increased uptake in the right ankle and right knee; the isotope uptake of the other bones was normal. Systematic radiographic examination (Figs 7 to 13) revealed peripheral osteolysis marked by small honeycomb-like cysts and by large confluent areas. Some of the defects were sharply defined and others showed a gradual transition to normal marrow. The lesions localised centrally or peripherally in subcortical bone affected the diaphyses, metaphyses, and epiphyses of long bones, but when the lesions were situated subcortically in the diaphyses of long bones the cortex was much thinner than normal. The distribution of these defects was asymmetrical. There were neither reactive changes of the adjacent bone structures nor sclerosing edges. Periosteal reactions or ballooning of the bone was not seen.

**DISCUSSION**

It is impossible to differentiate intra-osseous lipomata from simple bone cysts by radiography alone since trabeculae and central calcifications may be seen in both (Hart 1973; Dick 1976). Expansion of the cortex was noticed in only 21 of 46 intra-osseous lipomata (Döhler and Harms 1981). The localisation and the radiographic features of solitary subchondral bone cysts are similar to the osteolysis in our patient. However, the cysts are not filled with adipose tissue, but with a yellow coloured serum-like fluid. Histologically the fibrous membranes may resemble synovial structures (Dick 1976). There are no reports in the literature of multiple subchondral bone cysts.

Intra-osseous lipomata are characterised by a neoplastic overgrowth of univacuolar fat cells. Two thirds of these tumours contain bone trabeculae which are often atrophic. A fibrous capsule, capillaries in place of sinusoids, and the lack of haemopoietic bone marrow are further characteristics of intra-osseous lipomata. The latter may be regarded as a diagnostic feature only when haemopoiesis is normally present. Harms and Grebe (1972) demonstrated that the number of capillaries in the lipomata was twice as high as that in normal adipose tissue. This difference is an important method for detecting lipomata.
Some of these tumours can be classified as angiolipomata or haemangiolipomata. Microthrombosis may be related to the vascularisation of the lipomata. Formation of the microthrombi results in fibrous occlusion of the vascular lumen and it is possible that the interstitial fibrosis in our patient may have arisen from previous microthrombosis.

The osteolysis in our patient cannot be grouped with any of the bone dysplasias already described in the literature (Spranger, Langer and Wiedemann 1974). Since two of the cysts appeared to be radiographically identical and had the same histological features, it is conceivable that this unknown syndrome represents a multicentric hamartoma of intra-ossous adipose tissue.

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REFERENCES