Acta Orthop. Belg., 2006, **72**, 786-789

CASE REPORT

Ossifying fibroma of long bones in adults : A case report

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Ossifying fibroma (osteofibrous dysplasia) is a rare fibro-osseous lesion made up of fibrous tissue with woven bone formation. It is most commonly found in the tibia and fibula of children ten years of age or younger. The most important differential diagnosis is monostotic fibrous dysplasia, which is radiologically similar but without woven bone rimmed by active osteoblasts like ossifying fibroma on histological examination. No epitheloid cells are found as in adamantinoma. We report the case of a 45-year-old woman who had a 12-month history of pain and slight swelling. Radiographs showed a multilocular radiolucent lesion with sclerotic rim in the proximal tibia. The lesion was curetted and the defect was packed with bone graft and acrylic cement. Microscopic examination showed active osteoblasts rimming the irregulary woven bone. One-year follow-up showed good functional recovery without recurrence of the lesion.

Keywords : osteofibrous dysplasia ; ossifying fibroma.

INTRODUCTION

Ossifying fibroma (osteofibrous dysplasia) is a rare fibro-osseous lesion most commonly found in tibias and fibulas of children under the age of ten.

The first report of osteofibrous dysplasia was in 1921 by Frangenheim (*4*) who considered it congenital osteitis fibrosa. In 1966 Kempson (*7*) used the term "ossifiyng fibroma" to describe two lesions in the tibia of young children that generally resembled fibrous dysplasia but were histologically distinct from that condition in that the bone trabeculae in the abnormal tissue were covered by active osteoblasts. The lesion has also been referred to as "congenital pseudoarthrosis" and "congenital fibrous defect of the tibia". In 1976 Campanacci (*2*) used the term "osteofibrous dysplasia of the tibia and fibula" for this lesion to emphasize the anatomic location, developmental origin, and histologic similarity to fibrous dysplasia.

CASE REPORT

A 45-year-old woman, born in Morocco presented with a twelve-month history of pain and slight swelling of her left knee. There was no history of trauma. She had no relevant previous medical history, and was only on antihypertensive treatment.

Physical examination showed swelling over her proximal left tibia and local pain. There was no

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No benefits or funds were received in support of this study

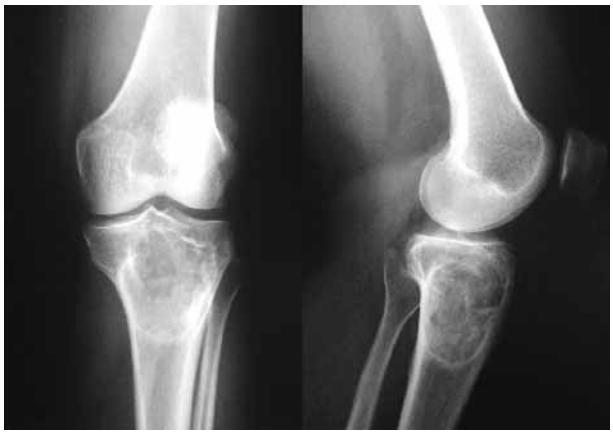

Fig. 1. — Anteroposterior and lateral radiographs showed a multilocular radiolucent lesion with marginal sclerosis in the proximal tibia.

joint effusion. There were no proximal lymphadenopathies present.

Results of laboratory tests were all within normal limits. Radiographs of the tibia and CT-scans showed a multilocular radiolucent lesion with marginal sclerosis in the proximal tibia. The lateral cortex of the tibia appeared expanded and attenuated (fig 1).

The lesion was curetted and the defect was packed with allogeneic bone graft filling the metaphyseal defect and polymethylmethacrylate bone cement filling the intramedullary space with two cancellous cross screws (fig 2).

Microscopic examination showed a fibrous stroma and scattered bony trabeculae rimmed by active osteoblasts (fig 3).

One-year follow-up showed good functional recovery without recurrence of the lesion.

DISCUSSION

Ossifying fibroma is a rare condition usually affecting the tibia and fibula in the first two decades of life. Although this lesion was radiographically similar to monostotic fibrous dysplasia, Kempson (*7*) considered it as another pathological entity based on its clinical aggressive behaviour and active osteoblastic rimming of the woven bone on histologic examination. Later on, Campanacci and

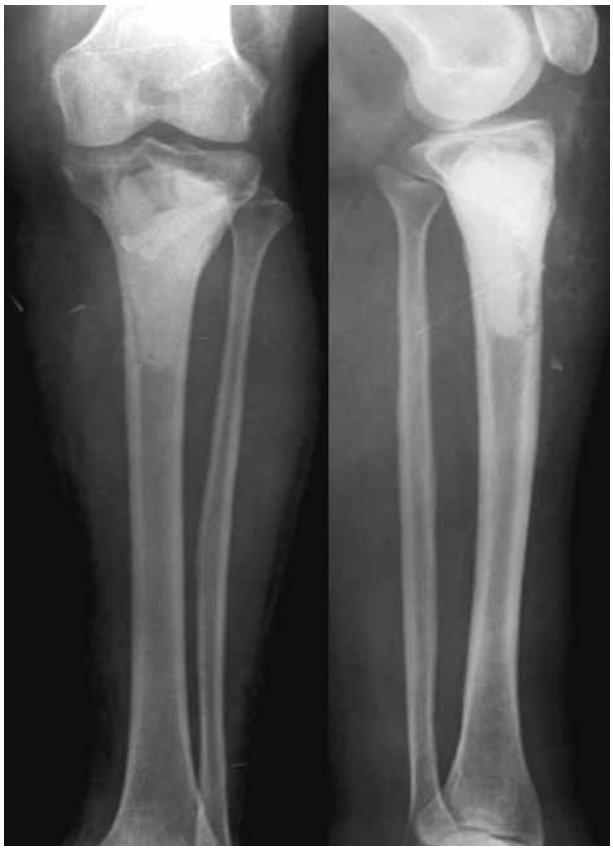

Fig. 2. — Postsurgery radiograph after curettage and allogeneic bone graft, acrylic cement and two cancellous-bone screws.

Laus (*1*) proposed the term "osteofibrous dysplasia of tibia and fibula", from their study of 35 patients, suggesting its histological similarity to fibrous dysplasia and predilection for the tibia and fibula.

The most common site is the middle third of the diaphysis of the tibia, followed by the upper third and the distal third (*2, 7*). In the fibula the mid-diaphysis is generally involved. The symptoms almost always appear in the first decade of life, most often before the age of five (60% of cases), and very rarely in adults. There is no gender preponderance.

The most common clinical manifestation is enlargement of a bone associated with a bowing deformity. It usually runs a protracted course with minimal symptoms. Pathological fracture may be a presenting feature. Another significant complication, described in 1989, is the coexistence of an adamantinoma within an osteofibrous dysplasia-like

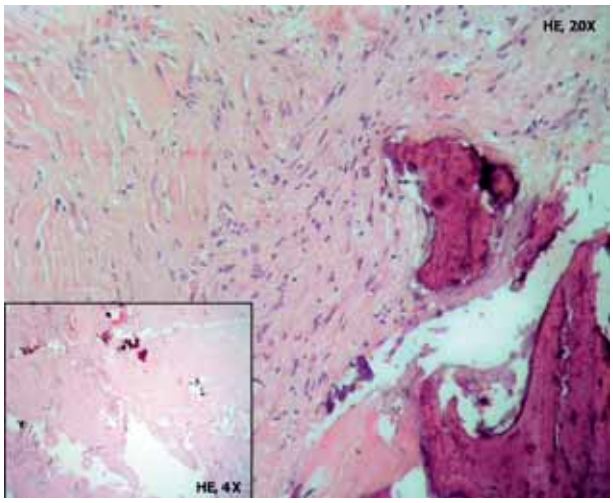

Fig. 3. — Photomicrograph of histological section. Spindle-shaped stromal cells have a storiform pattern with woven bone rimmed by active osteoblasts (haematoxylin-eosin, ×4 ; ×20).

lesion, but the relationship between these two conditions is unclear (*6*).

The typical roentgenographic feature of the lesion is a multilocular, confluent, eccentric, or intracortical osteolysis with surrounding sclerotic margins in the diaphysis or metaphysis. The cortex is expanded but not breached. Bowing of the bone becomes evident if a pathologic fracture occurred previously. When the lesions are small the radiographic presentation often is similar to that of metaphyseal fibrous defect or chondromyxoid fibroma. Large lesions sometimes have an appearance similar to that of fibrous dysplasia, with the cortical location being the most helpful and distinguishing feature. Some larger lesions, especially those consisting of multiple lucencies with interconnecting sclerosis, are difficult to distinguish from adamantinoma, although osteofibrous dysplasia tends to have a less aggressive appearance.

The main differential diagnoses are fibrous dysplasia, adamantinoma and non-ossifying fibroma (*2*). Fibrous dysplasia is predominantly intramedullary while eccentric expansion, and intracortical osteolysis is present in osteofibrous dysplasia. It usually involves the femur and the ribs, whereas osteofibrous dysplasia has a predilection for the tibia and fibula.

It has previously been considered that the diagnosis could be made on the basis of the characteristic clinical and radiological features, but since the observation of coexisting epithelioid tumours it is mandatory to have a histological confirmation of the diagnosis.

Ossifying fibroma or osteofibrous dysplasia presents two fundamental patterns ; fibrous tissue rimmed by bone trabeculae surrounded by active osteoblasts and a "zonal" architecture (*1, 2*). The fibrous tissue often has a stellate pattern and is rather loose, with delicate collagen fibers ; sometimes it is more dense, with bands of packed collagen fibers. The fibroblasts generally are well differentiated. The central fibrous area with new bone formation radiating to the outer area of more mature anastomosing lamellar bone spicules contributes to the distinctive zonal architecture of this lesion (*2*). Histologically, fibrous dysplasia has more cellular and less fibrous tissue, the woven bone is not rimmed by active osteoblasts and these is no zonal architecture (*2, 7*). However, after trauma the bone spicules in fibrous dysplasia may show osteoblastic rimming and the stroma may become dense. In such cases there are also haemosiderin deposits, haemorrhage, and inflammatory cells, which usually are not seen in osteofibrous dysplasia (*7*).

Adamantinoma of the long bones usually affects the middle portion of the tibia and can produce a radiographic appearance similar to that seen in cases of osteofibrous dysplasia. An association between adamantinoma and fibrous dysplasia was first suggested by Dockerty and Meyerding in 1942 (*3*) and studies from the Mayo Clinic also have emphasized this relationship (*6*). In 1992, it was demonstrated that keratin-positive cells were present in 93% of the cases of osteofibrous dysplasia studied but none showed progression to adamantinoma (*11*). One must, however, be cautions in extrapolating keratin positivity in osteofibrous dysplasia to a relationship with adamantinoma, because some authors suggest hat osteofibrous dysplasia may be one of a variety of mesenchymal lesions that show keratin positivity. Histologically, small islands of epithelial cells are found in adamantinoma.

The other characteristic feature of osteofibrous dysplasia is its aggressive clinical behaviour. The

recurrence rate varied from 64% to 100% after curettage, even a wide resection including the periosteum did not prevent its recurrence (*10*). The most important factor that affects the outcome is the patient's age.

Nakashima *et al* reported a 100% recurrence in ten cases treated in patients younger that ten years of age (*8*). Goergen *et al* reported two cases of ossifying fibroma : a 3.5-year-old boy had multiple recurrences after excision and resection, and a 16-year-old girl had a successful result after a single excision (*5*). Campanacci and Laus found that recurrence never occurred if patients were seen after the age of ten years, and progression of the lesion tended to become stabilized before puberty (*1*). In their report, a seven-year-old boy had two recurrences after excision of an ulnar lesion, whereas the other three patients, who were treated after 18 years of age, had excellent results. The authors suggested that surgery should not be attempted in patients younger than 15 years of age.

In patients younger than 15 years of age, the presence of epithelioid tumours as reported in 1989 by Keeney *et al* (6) must be excluded, as they can mimic osteofibrous dysplasia, but may be distinguished by the presence of epithelial cells on histological examination. If osteofibrous dysplasia is confirmed, conservative treatment with bracing to prevent fracture and regular follow-up examination is advised. Definitive surgery with marginal excision and bone grafting is considered if the lesions persist after 15 years of age. Wide extraperiosteal resection and bone grafting is reserved for patients who have repeated fractures or a rapidly progressing lesion, but it is seldom necessary.

The prognosis of osteofibrous dysplasia is usually good if it is treated correctly. During the first years of life, it should not be treated surgically because the lesion may undergo spontaneous repair and there is a high probability of recurrence if surgically treated. Between 5 and 10 years of age the indications for surgical treatment are influenced by the presenting symptoms, and the surgical treatment should be delayed as long as possible. Marginal excision or curettage and bone grafting will cure this disease when performed after 15 years of age. However, in a recent publication, Lee *et al* have recommended segmental extraperiosteal excision in all cases and excision and reconstruction in extensive lesions, in view of the risk of persistent morbidity if left untreated, and of the possible association of osteofibrous dysplasia with adamantinoma (*9*). The surgical management of this lesion thus remains controversial.

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