Case of Monostotic Fibrous Dysplasia in the Hand

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Summary

A case of monostotic fibrous dysplasia in the proximal phalanx of an otherwise healthy, twenty-five year old is discussed.

Fibrous dysplasia in the hand is rarely seen. Our patient presented with a swelling in his proximal phalanx. X-rays showed a lytic lesion. The lesion was treated with excision biopsy and cancellous bone grafting. Histological examination excluded malignancy but was diagnostically inconclusive. At recurrence, two and a half years later, a diagnosis of fibrous dysplasia was made. Intractable pain necessitated a ray amputation of the affected phalanx. The indications for surgical intervention in cases of fibrous dysplasia are discussed.

Introduction

Fibrous dysplasia of bone is characterised by localised replacement of the bone interior by fibro-osseous connective tissue. It is a relatively common bone disorder and is most frequently seen in children and young adults in the 2nd to 3rd decades.

Fibrous dysplasia may be accompanied by extra-skeletal manifestations, such as abnormal cutaneous pigmentation and endocrinopathies. Fibrous dysplasia is often monostotic but may be polyostotic. The more common sites where monostotic fibrous dysplasia occurs are the ribs, the maxilla, mandible, femur and tibia. Involvement of the hand is rare. In 1986, the Bone Tumors Studying Group reviewed the literature on bone tumors and dystrophies localized in the hand and found that only five cases of fibrous dysplasia in the hand had been described. The group reported that the diagnosis was often missed before operation.

Keywords

fibrous dysplasia, monostotic fibrous dysplasia, hand, phalanx

Figure 1: X-ray (right hand) after recurrence (November 1997)
Histological examination of the biopsy specimen showed connective tissue consisting of spindle cells and areas of bone formation. This was suggestive of either a reparative bone process superimposed on a fracture through a bone lesion or less likely, local callus formation surrounding an aneurysmal bone cyst. No evidence of malignancy was found. There was good post-operative recovery and the patient was discharged from the Hand Clinic after a short period of follow-up appointments.

Two and a half years later (November 1997), the patient returned with a recurrence at the same site. X-rays confirmed a cystic lesion of approximately double the dimensions to the one previously removed (Figure 1). An excision biopsy was performed through the old scar and several specimens were sent for histological examination. Bone grafting was not performed in this intervention. Microscopic views of the excised fragments revealed poorly oriented bony trabeculae in C-shapes or circle forms compatible with a diagnosis of fibrous dysplasia. There were no osteoblasts rimming the trabeculae (Figure 2).

The patient complained of intractable pain at the site despite apparently complete excision of the lesion. A ray amputation of the ring finger was finally opted for. The procedure was performed through a racquet incision along the right 4th metacarpal. After careful dissection, the digital nerves and flexor tendons were excised as proximally as possible. The slip of extensor digitorum longus to extensor digiti minimi was preserved. An osteotomy was performed at the base of the 4th metacarpal and after repair of the deep transverse ligament, the 3rd and 5th metacarpal bones were approximated and stabilised with transverse K-wires. Histology of the excised phalanx confirmed fibrous dysplasia. No evidence of malignant change was present. The K-wires were removed after 4 weeks. Wound healing was satisfactory.

Six months post-op, the patient was satisfied with the appearance and function of his hand. Strength studies showed the grip strength of his right hand to be practically equal to that of his left.

Discussion

Patients with fibrous dysplasia are most often evaluated and treated by orthopaedic surgeons. The indications for surgery in a patient with suspected fibrous dysplasia include:

1. Establishing a diagnosis. Although the X-ray appearance is usually typical, biopsy may be necessary to confirm the diagnosis.
2. Relief of intractable bone pain. This may involve the removal of affected bone followed by vascularised bone grafting. The latter has been successfully performed with fibrous dysplasia of a metatarsal, so as to maintain a functional, pain-free foot and at other sites such as the humerus and radius. In the case discussed above, intractable pain was treated by ray amputation.
3. Correction of skeletal deformity. Surgery is advised when significant deformity is present or when pathological fracturing occurs.

4. Malignant change. Sarcomatous transformation of fibrous dysplasia, most commonly to osteosarcoma, is a serious and rare complication occurring in a fraction of a per-cent (0.5%) of all cases. Sarcomas may arise spontaneously or following irradiation. Early diagnosis and adequate treatment may improve the prognosis. (The use of radiotherapy for the treatment of fibrous dysplasia is controversial. It is probably not advisable as it may increase the risk of sarcomatous change.)

Apart from surgery, preliminary studies with Pamidronate (Aredia®) suggest that some fibrous dysplasia patients may derive considerable benefit from this therapy.

References