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Fibular Grafting for Fibrous Dysplasia with Pathological Fracture of Proximal Femur: A Case Report

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Abstract

Fibrous dysplasia is a congenital, no hereditary skeletal disorder where a portion of a bone develops abnormally. It accounts for approximately 7% of all benign bone tumors. It is a developmental anomaly of bone formation in which the marrow is replaced by fibrous tissue. The bones most frequently involved are the long bones: femur (most common), skull, and the ribs.

Keywords: Fibrous dysplasia; Pathological fracture; Fibular grafting

Case Report

History

A 7 yrs old female child from muzzafarnagar presented to the OPD with the history of fall while playing. Subsequently she developed pain and deformity in the left lower limb and was unable to bear weight on the affected limb.

Examination

On local examination overlying skin was normal, deformity with shortening of limb was present. Skin over other body area was normal and no deformity was seen on the other limbs.

Investigations

X-rays of the affected limb showed lytic expansile lesion in metaphyseal region of the proximal femur with diapyseal extension along with ground glass appearance with thinning of cortex associated with fracture in subtrochanteric region of femur (Figure 1). X-rays of other limb were within normal limits. Her blood investigations showed raised alkaline phosphatase (402 IU/L reference range 5 IU/L to 112 IU/L), raised ESR (23 reference range 1 mm/1st hour to 20 mm/1st hour). Other investigations were within normal limits. Based on clinical and radiological features diagnosis of mono-ostotic fibrous dysplasia with pathological subtrochanteric fracture was made.

Management

After presentation limb was immobilized with skin traction and adequate analgesia was given. An operative intervention with curettage, internal stabilization along with bone grafting was done.

Patient was placed in a supine position; the buttock of the affected side was cushioned and the proximal femur was approached. The cavity which was filled with blood and grayish white tissue was adequately exposed. Adequate curettage of cavity was done and bone gap between the proximal and distal fragment was bridged with about 10 cm of fibular strut graft taken from the contralateral side. Internal fixation was done using two enders nail of appropriate size with one passing through the fibular graft (which was predrilled using a 2.5 mm drill bit) and the other ender nail also passing through both the fragments but outside the fibular graft (Figure 2). Following this cancellous bone graft from iliac crest was placed in cavity. Wound was closed in layers over the drain. The patient had an uneventful recovery. Post operatively two units of blood were transfused and boot and bar cast was given to prevent rotational deformity. Fifteen days following surgery sutures were removed and hip spica cast was applied.

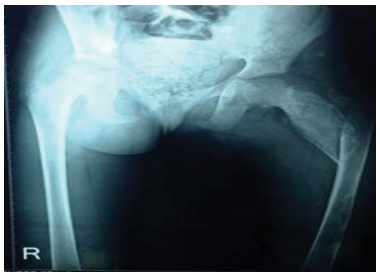

Figure 1: Pre- Operative AP view of pelvis with both hip.

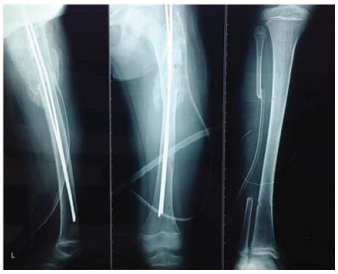

Figure 2: Post operative Xray showing fibular graft with enders nail and fibula taken from the contralateral side.

Discussion

The term fibrous dysplasia was originally proposed by Lichtenstein [1,2]. Fibrous dysplasia can be classified into one of three categories. Monostotic fibrous dysplasia involve only one bone, and many of these patients remain asymptomatic unless a fracture or swelling occurs. The polyostotic form is more severe, involving multiple bones. Nearly any bone in the body may be affected, including the long bones of the extremities, skull, vertebrae, pelvis, scapula, ribs and bones of hand and feet. Often one side of the body (in particular, one of the lower extremity) is more severely affected, resulting in deformity and limb

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length discrepancy [3,4]. Craniofacial involvement occurs in nearly 50% of patients with polyostotic disease. The third category, polyostotic form with endocrine abnormality, is the least common form. Precocious puberty, premature skeletal maturation, hyperthyroidism, hyperparathyroidism, acromegaly and cushing's syndrome can occur in these patients [5]. The triad of precocious puberty (endocrinopathy) , café au lait spots, and polyostotic bone involvement is commonly referred to as mcCune-Albright syndrome [6].

Repetitive microfractures can lead to a "shepherd's crook" deformity with pain, significant various at the femoral neck, shortening of the femur, an obvious Trendelenburg gait, and limited mobility. The condition begins before birth. It is caused by a gene mutation that affects the cells that produce bone. The gene mutation causes the cells to form an abnormal type of fibrous bone. This fibrous bone gradually grows and expands over a period of years, causing a weakened area of the bone. The area of weak bone can cause pain. It can crack (fracture) the bone, and may lead to deformity.

The radiographic appearance is characteristic with the lucent area having a granular ground glass appearance with expansion of the involved area of bone along with deformity that is usually seen as bowing [7].

Magnetic resonance imaging (MRI) will show the involved area of bone and may be helpful to determine whether areas have become cancerous. When the lesions of fibrous dysplasia are actively growing, blood tests may show elevated levels of the enzyme alkaline phosphatase. However, these abnormalities are not specific for fibrous dysplasia. They can be seen in other conditions with high bone turnover, including normal growth.

With monostotic fibrous dysplasia, it may be difficult to differentiate small lesions from simple bone cysts on radiographs. Less often, small lesions may be confused with histiocytosis or enchondromas. In these cases a biopsy may be necessary. Larger lesions with cortical thinning and a ground-glass appearance usually do not require a biopsy to confirm the diagnosis. Polyostotic fibrous dysplasia is readily identified on radiographs [8].

Presence of fibrous dysplasia is not an indication for surgery. Areas of fibrous dysplasia that are not symptomatic may be observed with periodic x-rays and not treated if they are not progressing. Braces may occasionally be used to prevent fracture, but they have not been effective in preventing deformity. Bisphosphonates have been effectively used in the relief of pain because it decreases the activity of cells that dissolve bone.

Operative intervention is needed when repeated pathological fracture have occurred, when lesion causes significant or progressive deformity or when associated pain becomes persistent. Pathological fracture can occur after mild trauma, they are often minimally displaced and they heal at a normal rate. Delayed union or non-union is not a problem but progressive deformity [9].

The primary goal of treatment is to realign the deformed bone, particularly in the weight bearing lower extremities. If the fracture occurs in the long bone or if involves the proximal femur, surgical intervention is the preferred approach. Internal fixation maintains proper alignment and can be achieved with intramedullary rods in the long bones or with compression screws with side plates in the proximal femur [9].

Additionally, scooping out (curettage) of the fibrous dysplasia is generally performed along with bone grafting. Osteotomies may be required to achieve satisfactory alignment if shepherd's crook deformity is present.

Enneking et al. reported that cortical strut grafting was effective in strengthening the bone in the proximal femur [10].

Conclusion

For carefully selected patients, cortical bone grafting is an excellent procedure for fibrous dysplasia. Curettage with bone grafting is effective because it has chance of local recurrence. Cortical bone grafts provide strong structural support to bone biomechanically weakened by fibrous dysplasia. Despite the successful use of internal fixation and near anatomic bone realignment, progressive deformity can still occur, leading to the need for additional surgery.

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