Ilizarov technique for correction of the Shepherd’s crook deformity: a report of two cases

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ABSTRACT

We report 2 patients with fibrous dysplasia who underwent correction, using the Ilizarov technique, of Shepherd’s crook deformities and pathological fractures of the left femurs. A 12-year-old boy underwent an opening wedge osteotomy and a 48º gradual correction, whereas a 43-year-old woman underwent a 34º acute correction without osteotomy at the fracture site. Both patients could initiate early weight bearing. Final leg function was excellent and alignment was maintained. No complications were encountered. Both patients had no difficulty sleeping and no major complaints about the Ilizarov technique. It is more important to achieve accurate alignment than resection of the lesion. The Ilizarov technique is effective for treating the Shepherd’s crook deformity in patients with fibrous dysplasia.

Key words: fibrous dysplasia of bone; fixators, external; Ilizarov technique; limb deformities, congenital

INTRODUCTION

The shepherd’s crook deformity of the proximal femur in patients with fibrous dysplasia may result in limb shortening, limping, fatigue, pathologic fracture, and hip pain. Various surgical treatments have been reported including curettage and cancellous bone grafting, valgus osteotomy, plate fixation, intramedullary nailing, and cortical bone grafting. We report on 2 patients with fibrous dysplasia in whom the Ilizarov technique was used to correct Shepherd’s crook deformity with pathological fracture of the left femur.

CASE REPORTS

Case 1
In 1992, a 12-year-old boy presented to our hospital with left thigh pain and a limp. He had been diagnosed with polyostotic fibrous dysplasia at age 3 and underwent curettage and cancellous bone grafting of the left femoral neck at age 4. Radiographs showed a pathological fracture of the left femur (at
a cystic intertrochanteric lesion), varus deformity of the femoral neck, and 1.5-cm shortening (Fig. 1a). He underwent curettage and cancellous bone grafting after insertion of a frozen allogeneic fibula into the femoral head through the greater trochanter. The fracture was then fixed with the Ilizarov apparatus and the centre of angulation was osteotomised (Fig. 1b). After 8 days, an opening wedge osteotomy was performed and a 48º gradual deformity correction continued for 42 days. Full weight bearing was started after the deformity correction was completed. The neck-shaft angle was corrected from 77º to 148º (Fig. 1c). This overcorrection was made because the varus deformity was expected to increase during growth. The duration of external fixation was 184 days. At the 5-year follow-up, growth was completed and the neck-shaft angle was 130º. At the 7-year follow-up, a fibrodysplastic lesion had recurred at the cancellous and cortical bone graft, but femoral alignment was maintained (Fig. 1d) and the patient was free from pain and had no limp. No infection, malunion, fracture, or pin breakage was encountered. The patient had no difficulty sleeping and no major complaints about the Ilizarov technique.

**Case 2**

In 2002, a 43-year-old woman presented to our hospital with thigh pain and gait disturbance after a fall. She had been diagnosed with monostotic fibrous dysplasia at age 9 and underwent curettage and cancellous bone grafting of the left femoral neck. She underwent conservative treatment for a femoral neck fracture at age 12. Radiographs showed a subtrochanteric pathological fracture at a cystic lesion and varus deformity of the femoral neck (Fig. 2a). It was fixed with an Ilizarov apparatus and then a 34º acute deformity correction was performed at the fracture site without open osteotomy. The neck-shaft angle was corrected from 85º to 130º (Fig. 2b). Full weight bearing was started at day 5. The duration of external fixation was 145 days. At the 2-year follow-up, no progression of the varus deformity was evident (Fig. 2c). The patient had no hip pain or limp. No complications were encountered. The patient had no difficulty sleeping and no major complaints about the Ilizarov technique.

**DISCUSSION**

Varus deformity of the proximal femur is common in patients with fibrous dysplasia. Curettage and cancellous bone grafting has been the treatment of choice for the Shepherd’s crook deformity, but unsatisfactory results such as bone graft resorption and recurrence of the lesion and varus deformity have been reported.9 Valgus osteotomy and internal fixation with a plate or intramedullary nailing can stabilise the fracture and prevent additional fractures and deformity.2 A single procedure limits the correction angle attainable, so large
deformity corrections require 2-stage operations. Overcorrection is necessary to achieve the correct mechanical axis because varus deformity usually increases during growth. Internal fixation is less useful in a large deformity correction as it disregards the mechanical axis. Full weight bearing can start only after bone union (weeks 6 to 16).

A strut cortical bone graft provides structural support for the proximal femur with fibrous dysplasia, with or without internal fixation, but the bone graft may be resorbed by the fibrodysplastic tissue, as in our first case.

The goals of treatment are to regain normal gait and relieve pain. Complete eradication of the lesion is infeasible. It is more important to achieve a good alignment of the bone fragments than resection of the lesions. Fibrous tissue is more active in young patients; when the fibrous tissue matures after puberty, mechanical stress is relieved. Shearing forces must be reduced by achieving mechanical alignment and adequate correction. If vascularity is preserved at the fracture site, the fracture will fuse within the normal healing period.

The advantage of the external fixator is its ability to modify the correction, lengthen limbs, minimise invasive surgery, and enable early weight bearing. No re-operation to remove implants is needed as in internal fixations. A ring fixator (Ilizarov apparatus) can provide 3-dimensional adjustment and residual deformity correction, although a unilateral fixator is easier and more comfortable for patients. Small wire and half pin insertions at different levels and in different directions enable rigid stabilisation and early weight bearing. Deformity correction using the Ilizarov technique can achieve accurate alignment without curettage of fibrodysplastic lesions.

REFERENCES