One-stage osteotomy and fixation using a long proximal femoral nail and fibular graft to correct a severe shepherd’s crook deformity in a patient with fibrous dysplasia: a case report

Himanshu Kataria, Neeraj Sharma, Rajesh Kumar Kanojia
Department of Orthopaedics, Lady Hardinge Medical College, New Delhi, India

ABSTRACT

We report a case where a one-stage osteotomy and fixation, using a long proximal femoral nail and fibular graft, was performed to correct a severe shepherd’s crook deformity (70° varus and 50° retroversion) of the femoral neck with a pathological stress fracture in a patient with fibrous dysplasia. The neck shaft angle was corrected to 125°. At the 57-month follow-up, the patient was free of pain and had no limp or evidence of recurrence.

Key words: bone nails; fibrous dysplasia, monostotic; fracture fixation, intramedullary; osteotomy

INTRODUCTION

In patients with fibrous dysplasia, the shepherd’s crook deformity is usually associated with varus deformity, limb shortening, limping, and occasionally, chronic fatigue fractures with disabling pain. Although it is usually managed with fixation using plates and screws, concentration of stress on the lower portion of the plates may induce secondary deformities. It is also difficult to fit plates conforming to the contour of the deformed femur.1,2 Fixation using an intramedullary nail and flexible nails has a high recurrence rate.3,4 Multiple corrective osteotomies using intramedullary devices in two stages is therefore the recommended procedure for a severe varus retroversion deformity.1,5 We report a case where a single-stage duoplanar subtrochanteric osteotomy and intramedullary nailing was used to correct a severe shepherd’s crook deformity of the proximal femur with a pathological stress fracture in a patient with monostotic fibrous dysplasia.

CASE REPORT

In July 2003, a 15-year-old male presented with a painful right hip and a limp. His right trochanter protruded laterally and his leg was rotated externally and shortened by 7-cm. He walked with a marked Trendelenberg lurch and the Trendelenberg test was positive. Radiographs showed a severe varus deformity of the right proximal femur (Fig. 1a). Three-dimensional computed tomography revealed a large expansile lytic lesion with a patchy area of ground...
glass haze in the upper metadiaphyseal and femoral neck, with evidence of a cortical breach suggestive of pathological fracturing. A 70° varus deformity and 50° retroversion were also present in the femoral neck (Fig. 1b). Bone scintigraphy confirmed the abnormal uptake and monostotic nature of the lesion. A frozen section biopsy subjected to histopathological examination confirmed the diagnosis of fibrous dysplasia.

Radiographs were used to determine the appropriate size and location for a closed wedge osteotomy of the proximal femur. The patient was placed in a supine position under an image intensifier and underwent a duoplanar osteotomy (valgus osteotomy with a 55° lateral closing wedge) of the subtrochanteric region. Antegrade femoral nailing with 2 screws achieved good purchase in the femoral head. A 6-cm-long, non-vascularised, fibular cortical graft was split in 2 to provide a spacer and mechanical strut effect on the lateral femur.

Postoperatively, partial weight bearing was allowed at 4 weeks. Full weight bearing was allowed at 5 months after radiological union. The neck shaft angle was corrected to 125° (Fig. 2a). At the 57-month

Figure 1  (a) A radiograph and (b) 3-dimensional computed tomographic scans showing an expansile lytic lesion in the right femorotrochanteric region with a patchy area of ground glass haze in the upper metadiaphyseal and femoral neck. The severe varus deformity and retroversion of the femoral neck and head is seen in the axial view.

Figure 2  Radiographs showing (a) bone union at 5 months and (b) a well-consolidated femorotrochanteric region with the neck shaft angle preserved after 57 months.
follow-up, the patient was free of pain, had no limp, a normally oriented patella and residual leg shortening of 1 cm. The Trendelenberg test was negative and he had no lurch in his gait. The proximal femur had consolidated, with no recurrence of varus deformity and retroversion and no change in the neck shaft angle (Fig. 2b). The patient was able to perform his activities of daily living normally.

DISCUSSION

Pathological fractures and varus deformities are common in patients with fibrous dysplasia, particularly in the proximal femur, because of the forces exerted by weight and muscle pull on the weakened bone. The fractures and varus deformities recur often, despite aggressive treatment.\(^3,5\) Cortical grafts are preferable to cancellous grafts or bone graft substitutes because of their superior physical quality. The cancellous graft is incorporated quickly and replaced by dysplastic tissue.\(^6\) The fibular graft yields better results than curettage or cancellous grafting.\(^6\)

A 2-stage procedure (curettage and bone grafting followed by corrective osteotomy) is advocated for severe varus deformities.\(^7\) We preferred to use a single-stage procedure, as the lesion was monostotic and carried a low risk of recurrence.

Using plates and screws for fixation in the vicinity of weakened bones may not provide sufficient stability for successful correction of deformities and pathological fractures. Stress fractures can occur in the distal regions of plates. The risk of refracture is high after implant removal.\(^1,2,5\) Multiple osteotomies and intramedullary nailing can prevent fractures and improve the gait, but these have a high varus deformity recurrence rate.\(^3-5\) A long intramedullary nail that traverses the neck and has a firm purchase in the normal head can prevent subsequent loss of the neck shaft angle.\(^5\)

Fixation with a proximal femoral nail is particularly applicable for shepherd's crook deformity in patients with fibrous dysplasia, because of its ability to stabilise the neck and correct the deformity. Multidirectional nails or reconstruction nails have the same qualities, but approaching the pyriform fossa is difficult because of the severe varus deformity and muscle contracture while inserting a multidirectional nail.\(^5\)

The long proximal femoral nail is preferred because (1) it is easier to insert as the trochanter, rather than the pyriform fossa, is the entry point; (2) the size of its screws is larger, providing better purchase in the femoral head; and (3) it has a long central lever arm to hold in the distal diaphysis to prevent recurrence of deformity. It can accommodate osteotomies at multiple levels when required. Nonetheless, the dense fibrous bone is difficult to ream.

REFERENCES

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