Upsala J Med Sci 109: 123–130, 2004

A Two- stage surgery for severe femoral neck deformity due to fibrous dysplasia: A case report

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ABSTRACT

Various kinds of surgical treatments have been reported for varus deformity of the proximal femur due to fibrous dysplasia. We report a case of two-stage corrective osteotomy for severe varus-retroversion deformity of the femoral neck due to monostotic fibrous dysplasia. The patient was an 18 year-old man. On initial examination, the spina malleollar distance was 88 cm on the right side and 83 cm on the left. Plain radiography showed prominent varus deformity of his left proximal femur. The morphology was 130 degrees on the right side and 85 degrees on the left. Computed tomography revealed 60 degree retroversion of the femoral neck.

A two-stage surgery was performed, consisting of curettage and bone grafting followed by corrective osteotomy 16 months later. A 55 degree valgus osteotomy was performed in the subtrochanteric region. After osteotomy and 40 degree internal rotation of the shaft, a 130 degree angle plate was used for osteosynthesis. Postoperative radiological examination showed a morphology of 140 degrees and computed tomography revealed a 20 degree retroversion of the femoral neck. No recurrence or varus deformity was seen at four years after surgery. Although the leg length discrepancy was 2.5 cm, the patient had no difficulty in one foot standing and no restriction of ADL (activity of daily living).

The well-known progressive varus Shepherd's crook deformity in the polyostotic form of fibrous dysphasia is associated with limb shortening, limping, and occasionally chronic fatigue fractures with disabling pain (1). Various kinds of surgical treatments have been reported for this type of varus deformity (1–5). Curettage and bone grafting is one of the most common and simple treatments. However, this method often gives bad results as the grafted bones are absorbed and that the progress

Received 13 January 2004
Accepted 29 January 2004

Key words: monostotic fibrous dysplasia, osteotomy, two-stage surgery.

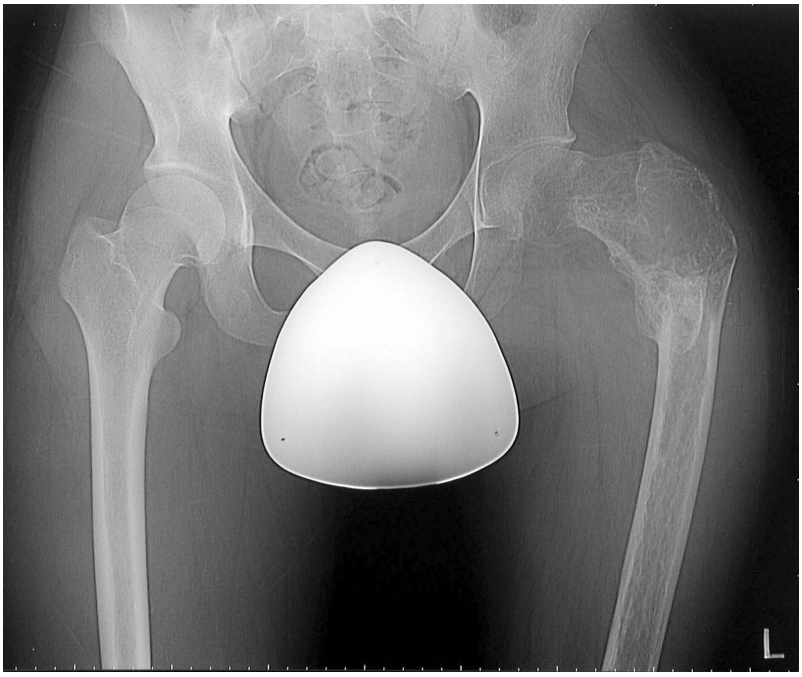

Fig. 1. Plain radiography showing a 5cm x 5.5cm sized radiolucent lesion with ground glass appearance and marginal sclerosis in the left femoral trochanteric region.

of varus deformity van not be prevented (6). We report a case of two-stage corrective osteotomy for severe varus-retroversion deformity of the femoral neck due to monostotic fibrous dysplasia.

CASE REPORT

The patient was an 18 year-old man. His chief complaint was left coxalgia. In August 1996, he noticed pain in his left buttock and thigh. In February 1997, he fell to the ground and hit his buttock. He was hospitalized with the diagnosis of left femur trochanteric pathological fracture. He underwent conservative treatment.

He was referred to our clinic because of pain when walking. On initial examination, the spina-malleollar distance was 88 cm on the right side and 83 cm on the left with a leg length discrepancy of 5 cm. His left trochanter protruded laterally and the left lower extremity was externally rotated. Plain radiography demonstrated a 5cm × 5.5 cm sized radiolucent lesion with ground glass appearance and marginal sclerosis in the left femoral trochanteric region. The femoral calcar was partially interrupted with a very thin cortex. Plain radiography showed prominent varus deformity of his left proximal femur. The morphology was 130 degrees on the right side and 85

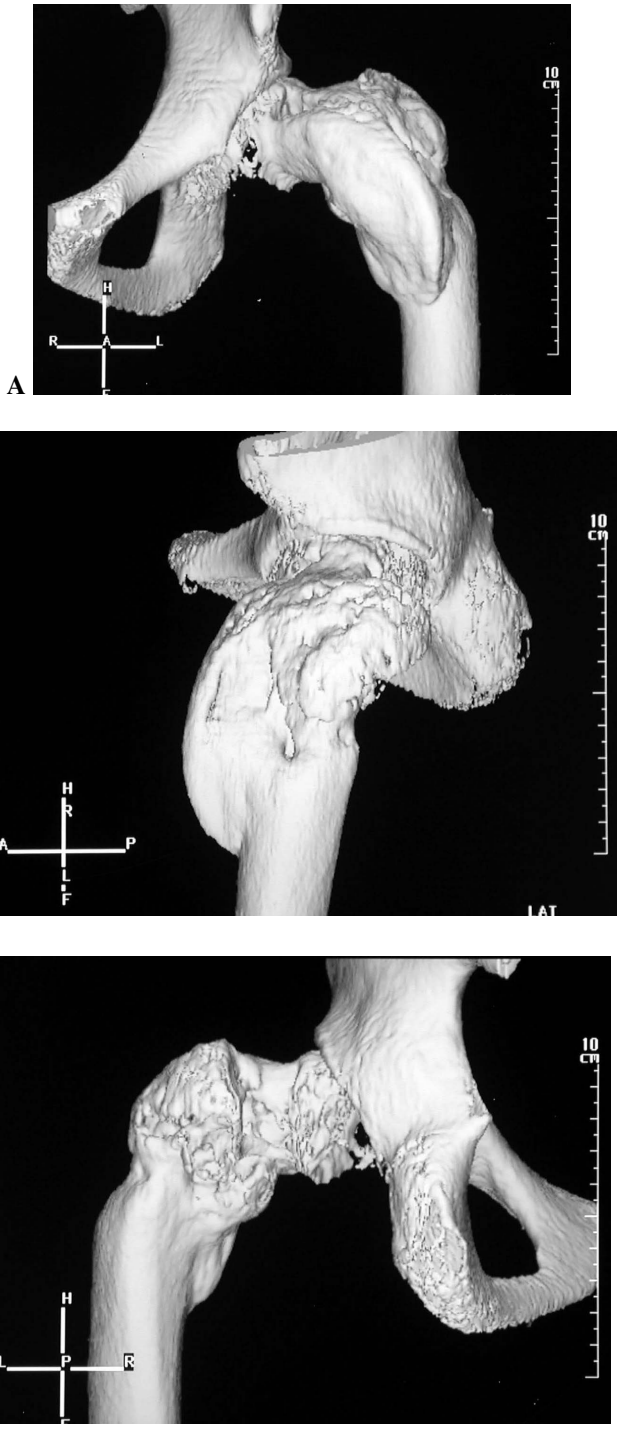

Fig. 2. Three dimensional CT visualizing varus and retroversion of the femoral neck. (A: anterior view, B: lateral view, C: posterior view).

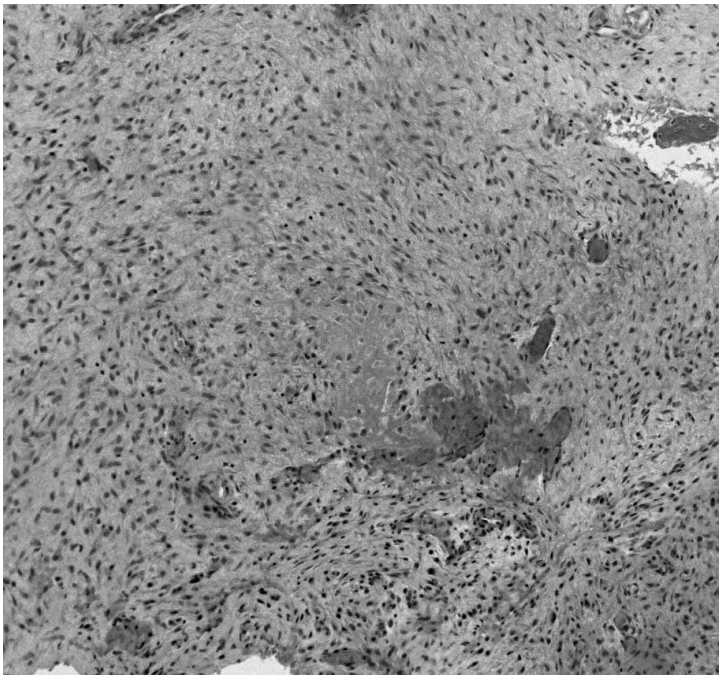

Fig. 3. Microphograph of the lesion showing cellular spindle cell proliferation with woven bones.

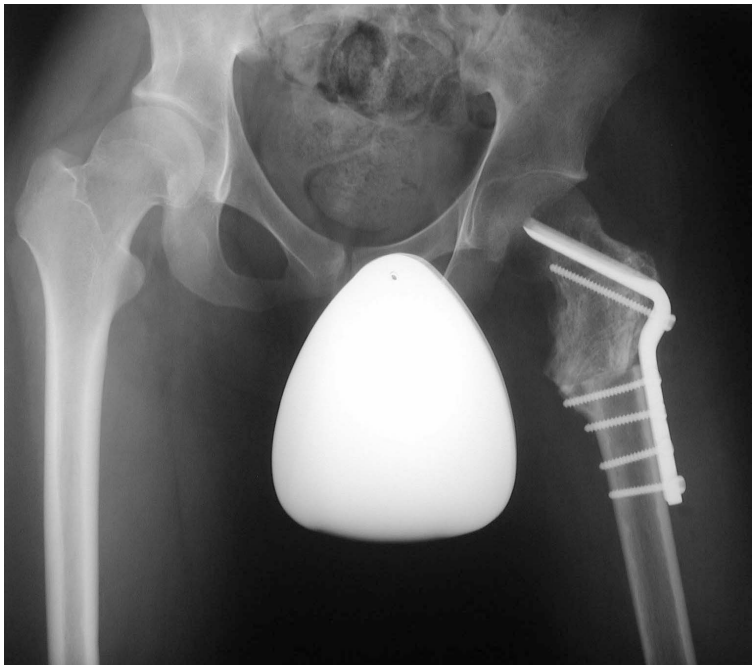

Fig. 4. Plain radiograph after surgery showing morphology of 140 degrees.

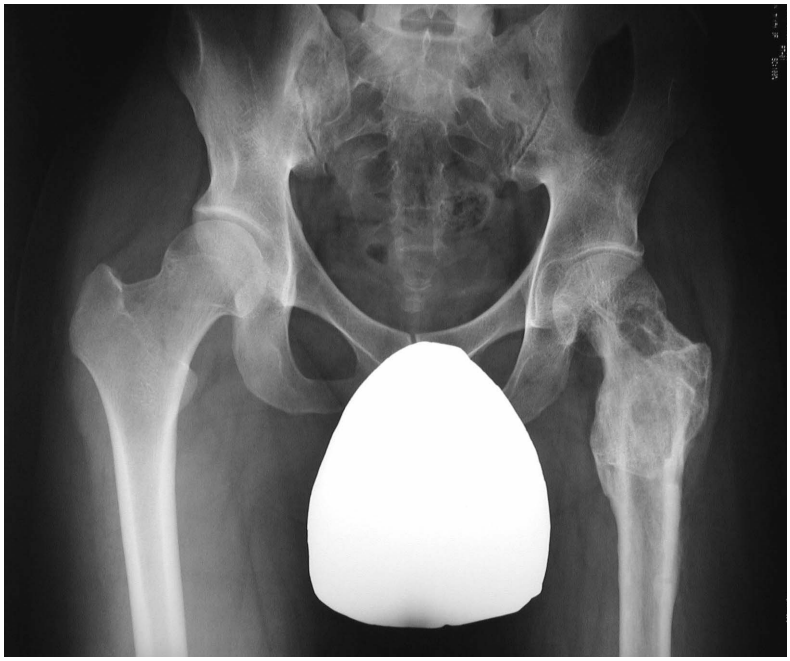

Fig. 5. Plain radiography showing no recurrence or varus deformity at four years after surgery.

degrees on the left (Fig. 1). Computed tomography (CT) revealed a 60 degree retroversion of the femoral neck.

Three-dimensional CT visualized varus and retroversion of the femoral neck (Fig. 2). MRI showed low signal intensities from the femoral head to the lesser trochanter on T1 weighted images and lobulous high signal intensities on T2 weighted fat suppression images. Bone scintigraphy showed increased abnormal uptake only in the left trochanteric region. The histological diagnosis of the open biopsy specimen was fibrous dysplasia (Fig. 3). The patient was diagnosed with monostotic fibrous dysplasia occurring in the femoral trochanteric region.

Operative treatment was chosen because of walking difficulty due to the severe hip deformity, leg length discrepancy and high risk of pathological fracture.

A two-stage surgery was performed, consisting of curettage and bone grafting followed by corrective osteotomy after 16 months. A 55 degree valgus osteotomy was performed in the subtrochanteric region. After osteotomy and 40 degree internal rotation of the shaft, a 130 degree angle plate was used for osteosynthesis. Postoperative radiological examination showed a morphology of 140 degrees and computed tomography revealed a 20 degree retroversion of the femoral neck (Fig. 4). No recurrence or varus deformity was seen at four years after surgery (Fig. 5). Although the leg length discrepancy was 2.5cm, he had no difficulty in one foot standing and no restriction of ADL (activity of daily living).

DISCUSSION

Pathological fractures and deformities are common in patients with fibrous dysplasia. The dysplastic bone is biomechanically unsound, and fatigue fractures often occur even with normal stresses. Fractures that result from fibrous dysplasia are often managed non-operatively, as it is thought that these fractures usually heal without difficulty(1, 6, 7).

Treatments for lesions in the proximal part of the femur, however, are troublesome. Symptomatic involvement of the proximal part of the femur frequently requires surgical intervention to achieve relief of pain and resumption of unrestricted activity. The forces of weight and muscle pull on the mechanically weakened bone in this area, producing an unrelenting propensity for fracture and varus deformity. Recurrent fractures and relentless progression of the varus deformity in spite of aggressive orthopedic treatment are characteristic involvements in this disease(1, 8, 9).

Curettage and bone grafting have been the mainstay of treatment for symptomatic and even asymptomatic lesions of fibrous dysplasia. Nakashima et al. reported that six of their eight patients who had lesions of the femoral neck were managed successfully with curettage and bone grafting (3). Stephenson et al. noted better results after curettage and bone-grafting in patients who were eighteen years or older (7). However, Enneking et al. described that the results of curettage and autogenous cancellous bone-grafting were unsatisfactory, since the grafts were incorporated quickly and eventually were replaced by dysplastic tissue (1). However, they reported good results after fibula graft without curettage. Guille et al. reported that curettage and cancellous or cortical bone grafting did not appear to have any advantage compared with osteotomy alone in the treatment of symptomatic lesions of fibrous dysplasia (6). In the present case, the varus deformity was so advanced that fibular grafting could not be chosen. A two stage surgery consisting of curettage and bone grafting followed by corrective osteotomy was chosen because of the severity of the varus deformity, and we obtained good functional results. One of the possible reasons for the poor results after correction of varus deformity is thought to be the high recurrence rate in cases of polyostotic fibrous dysplasia. The present case was a monostotic fibrous dysplasia, which has a lower recurrence rate as compared with the polyostotic type. We allowed 16 months to confirm that there was no recurrence of the lesion after curettage and bone grafting, and then, corrective osteotomy was carried out. We believe the patient's young age and good stability of the osteotomy design also contributed to good results.

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