Fixation of Subtrochanteric Fractures in Two Patients with Osteopetrosis Using a Distal Femoral Locking Compression Plate of the Contralateral Side

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Abstract
Osteopetrosis or Albers-Schönberg disease is a rare inherited skeletal disorder characterized by increased osteodensity and modeling defects. The fragility of such dense bone may lead to an increased incidence of fractures, especially on the hip and proximal femur. These fractures can pose problems during surgery, as the bones are hard and no medullary cavity is available for intramedullary nailing. Here we report our experience of two sisters who both presented with a subtrochanteric fracture that was fixed using a distal femoral locking compression plate (DF-LCP; Synthes, Obedors, Switzerland) of the contralateral side.

Key Words
Osteopetrosis · Fragility · DF-LCP

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Introduction

Osteopetrosis or Albers-Schönberg disease is a rare inherited skeletal disorder characterized by increased osteodensity and modeling defects [1]. Three forms are recognized: an infantile autosomal recessive malignant form, an intermediate autosomal recessive form, and an adult autosomal dominant form [2]. Infantile forms are usually diagnosed in the first year of life, and only 30% survive up to the age of six years. Patients with the intermediate form have a moderately decreased life expectancy. The benign adult form is however compatible with a normal life span [3].

The fragility of such dense bone may lead to an increased incidence of fractures, especially on the hip and proximal femur [4, 5]. These fractures can pose problems during surgery, as the bones are hard and no medullary cavity is available for intramedullary nailing. We are reporting our experience of two sisters who both presented with a subtrochanteric fracture in osteopetrotic bone that was fixed using a distal femoral locking compression plate (DFLCP Synthes, Obedors, Switzerland) on the contralateral side.

Case Report 1

A 35 year-old lady, a software engineer by occupation, sustained a trivial fall while descending the stairs in a metro station and developed severe pain in the right thigh. She was unable to get up on her own. X-ray examination revealed a transverse subtrochanteric fracture along with homogeneous hyperdense bones of the pelvis and opposite femur (Figure 1). There was hardly any medullary canal visible in the fractured femur on X-ray. We diagnosed osteopetrosis and planned for plate (DF-LCP; Synthes, Obedors, Switzerland) fixation of the contralateral side. We kept additional drill bits available in anticipation of difficulties when drilling these bones. We used a ten-hole titanium DF-LCP for the left femur. Bone drilling was indeed very difficult, but fortunately none of the drill bits were broken. The drill bits used with this device were self-tapping and 4.2 mm in diameter, which was advantageous in this case. We applied a locking screw head to the proximal and distal fragments after reducing the fracture. The design of the implant provides multiple

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Figure 1. Case 1, showing the subtrochanteric fracture in the osteopetrotic bone.

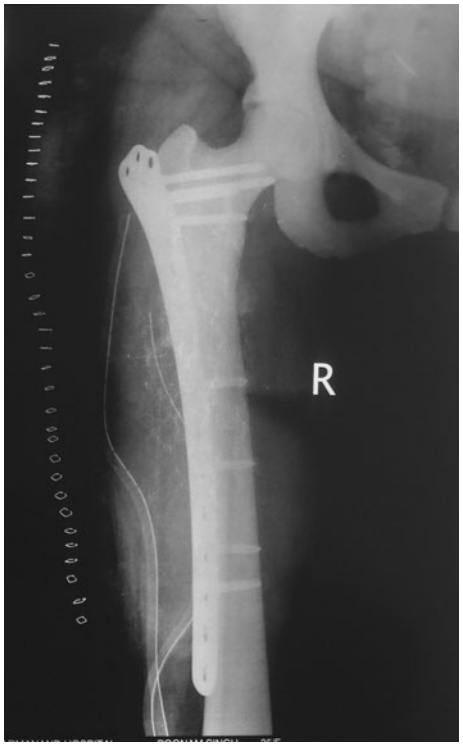

Figure 2. Immediate postoperative AP X-ray showing fixation using DF-LCP.

screw placements in the proximal femur and gives a stable fixation. Closure was done in layers and the staples were removed on the tenth postoperative day. Non-weight-bearing toe touch ambulation was started from the first postoperative day. Union was achieved at the 23rd postoperative week (Figures 2, 3, 4) and the patient was then allowed full weight bearing. At follow up she had developed an incomplete stress fracture of the left femur, which was managed conservatively.

Case Report 2

The 38 year-old sister of case 1 was already known to have osteopetrosis, and had previously presented with a subtrochanteric fracture of the left femur, which was fixed with a broad 4.5 mm LC-DCP three years ago and united. She again presented to us following a trivial fall with pain in the right thigh. X-ray revealed a subtrochanteric fracture of the right femur (Figure 5), and this time we planned fixation with a DF-LCP of the contralateral side. Fixation was achieved without any difficulty and the staples were removed on the tenth postoperative day. Immediate non-weight-bearing ambulation with a walking frame was permitted. Clinical and radiological union was achieved at the 21st postoperative week (Figures 6, 7). The patient returned to her preinjury level of activity without any residual disability.

Discussion

Osteopetrosis is characterized by increased bone deposition on unresorbed calcified cartilage or primary spongiosa [6], and the diagnosis is made by characteristic roentgenographic findings of generalized osteosclerosis primarily involving the axial skeleton and by symmetric involvement of the long bones without medullary defect [3]. The brittleness of the sclerotic bones results in severe damage, whereas the same trauma in a ''normal'' individual does not [4].

Lack of suspicion on the part of treating surgeons and inadequate planning in such cases can lead to many difficulties in the fixation of such fractures.

Various nonoperative treatment options have been reported in the past (Table 1). One of the earliest reports of nonoperative treatment was provided by

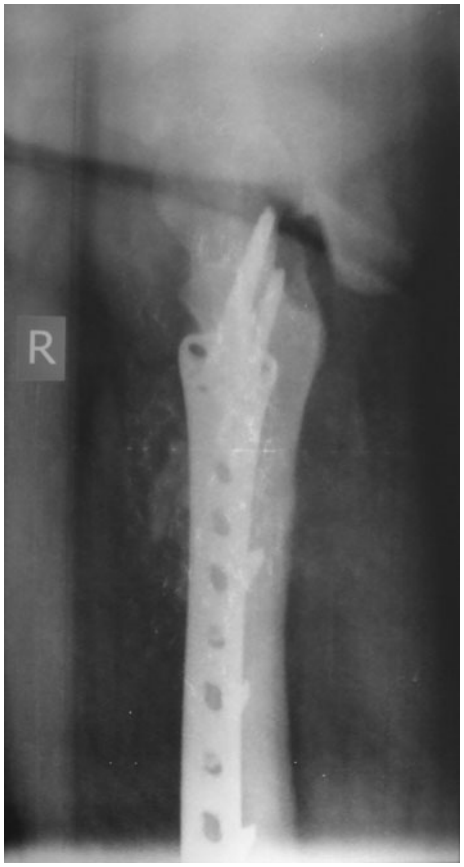

Figure 3. 23-week postoperative lateral X-ray showing union.

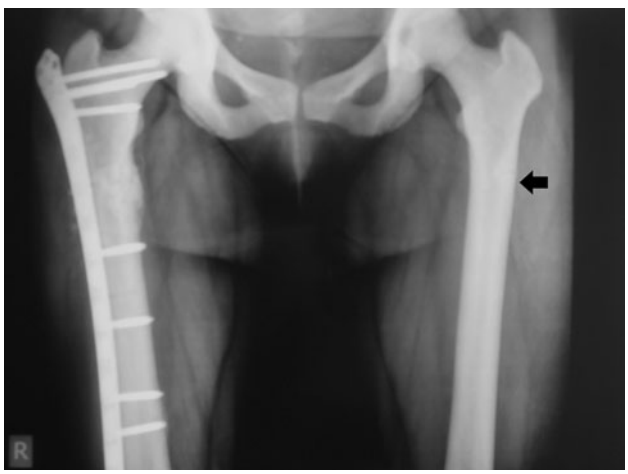

Figure 4. 23-week postoperative X-ray showing union and the stress fracture in the opposite subtrochanteric area.

Alexander [7] in 1923, when he reported the treatment of a peritrochanteric fracture in an adult female with a plaster cast which resulted in pseudoarthrosis. Hasenhultl [8] reported the treatment of a peritrochanteric fracture in a 27 year-old male with Russell

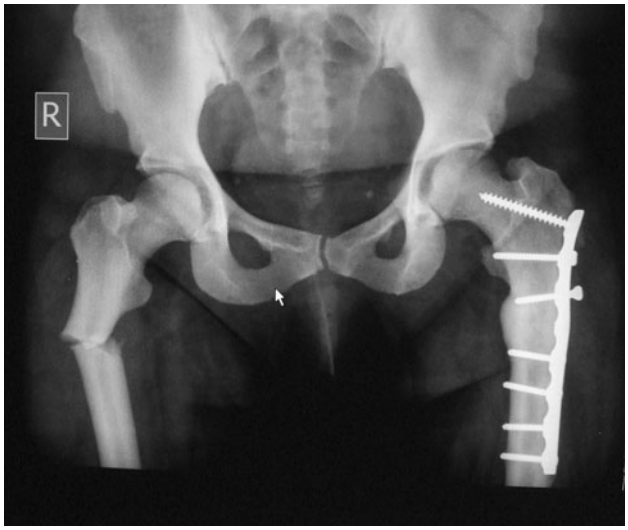

Figure 5. Case 2 showing the fracture in the right subtrochanteric area along with the previous healed fracture of the left femur.

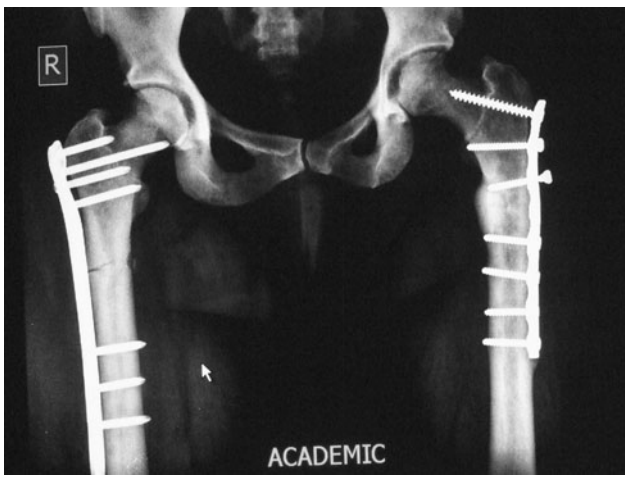

Figure 6. Six-week postoperative X-ray.

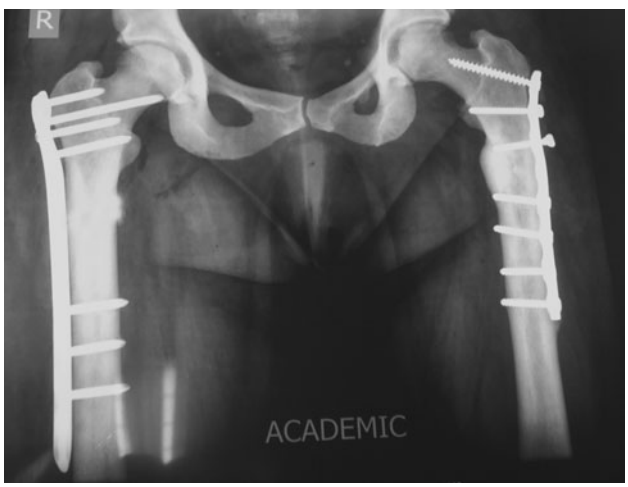

Figure 7. 22-week postoperative X-ray showing union.

Table 1. Reported studies of nonoperative osteopetrotic fractures in adults. (F: Female; M: male; R: right; L: left, P: peritrochanteric, FN: femur neck).

SN	Study	Age/gender	Fracture location	Treatment	Complication	Follow up
1	Alexander [7]	43/F	R, P	Nonoperative plaster cast	Pseudarthrosis	Nonambulatory
2	Hasenhuttl [8]	27/M	P	Russel traction		Union at 10 weeks, ambulatory
3	Dahl et al. [5]	18–22 (n = 2)/F	P	Plaster splint/cast		Union, ambulatory
4	Armstrong et al. [9]	6–16 (n = 3)	FN	Non weight bearing	Coxa vara deformity treated with a valgus osteotomy	Union, ambulatory
		8–30 (n = 2)	P	Nonoperative		Union
		Teenager/M	P	Hip spica		Union, ambulatory
		14/F	P	Traction for 10 weeks		Union at 16 weeks, ambulatory
5	Gupta & Gupta [10]	31/M	P	Traction		Union at 12 to 16 weeks, ambulatory
6	Birmingham [28]	56/M	L, P and FN	Hip spica cast	Coxa vara deformity	Union

traction where the fracture united at ten weeks. Dahl [5] reported two young patients who were treated with plaster casts for peritrochanteric fractures, which united in both patients. Armstrong [9] described the treatment of patients between the ages of 8 and 30 years. All of them were treated nonoperatively with traction and casts, and all fractures united eventually. Gupta & Gupta [10] treated a peritrochanteric fracture in an adult male with traction, and the fracture united at 16 weeks.

Many operative treatment modalities (Table 2) have been described in the past as having considerable difficulties and complications [4, 5, 7–20]. Kleinberg [15] treated a peritrochanteric fracture with plate, screws and a cortical strut graft. The plate broke and the fracture united in angular malposition. Yang [20] fixed a peritrochanteric fracture in a 21 year-old male with a Jewett nail, but placement failed. The fracture was then fixed with three screws and a hip spica cast was applied. The fracture united at 12 months but she needed hip contracture release during her course of treatment because of immobilization in the hip spica.

Milgram & Jasty [16] treated a peritrochanteric fracture in a 52 year-old lady with a Holt nail plate but encountered difficulty in placing the nail and screw. The fracture united after two years. Ashby [11] treated three patients with peritrochanteric fractures. The first one was a lady of 49 years who was initially treated by Zickel nail. The procedure was difficult and long (>6 h), with intraoperative fragmentation of the distal fragment. The lady was followed up for three years, and then X-ray showed displacement and comminution of the fracture about the nail. It was removed and a total hip arthroplasty was performed. Three years later she had an incomplete periprosthetic fracture at the tip of the femoral stem, which was treated by partial weight bearing for two months, and the fracture healed at ten months. The second patient was a 61 year-old lady with a fracture around the left hip who was treated by nail plate; union was achieved. Eight years later she had a fracture of the right hip treated with a nail plate. A deep infection developed, requiring removal of the implant. She then underwent a THA eight months later, but rehabilitation failed and she did not regain walking ability.

Gupta & Gupta [10] treated a peritrochanteric fracture in 31 year-old man with a blade plate; the fracture united after 16 weeks. Chhabra [13] described six patients with peritrochanteric fractures treated by dynamic hip screw in two cases, Kuntscher nail (K-nail) in three cases, and locked intramedullary nail in one patient. Both DHSs failed, one because of infection leading to nonunion and the other because the implant was pulled out, leading to nonunion. The fractures in the two patients with K-nails united, but the nail migrated in the third patient, after which a nail exchange was performed and the fracture healed. The fracture treated with the locked nail united uneventfully.

Among 25 reported cases (4 neck femur and 21 peritrochanteric fractures) treated by open operative methods, the nonunion rate was found to be 12% and the infection rate 12%. In the peritrochanteric group, the rate of hardware failure was 29% and the rate of

Table 2. Reported studies of operative osteopetrotic fractures in adults. (F: Female; M: male; R: right; L: left; P: peritrochanteric; S: shaft femur; FN: femur neck; ORIF: open reduction and internal fixation; DHS: dynamic hip screw).

	Study	Age/gender	Site of fracture	Treatment	Complication	Follow up
1	Klienberg [15]	35/M	L, P	Plate, screw and cortical strut graft	Plate breakage and angular malunion	Union
2	Yang [20]	21/F	L, P	Jewett nail	Failed implant, revised with screw + spica	Union at 12 months
		52/F	P	Holt nail plate		Union at 2 years
3	Belz [12]	46/F	L, S	Ext. fixator		Union at 13 weeks
4	Ashby [11]	49/F	L, P	Zickel nail	Fragmentation of distal fragment with nail placement, placed cerclage wires	Union
		52/F	L, P (3 years later)	THA after nail removal		Full weight bearing
		55/F	R, P (3 years later)	THA	Periprosthetic fracture 3 years later	Union at 10 months
		61/F	L, P (8 years prior)	Nail plate device		Union
		69/F	R, P	Nail plate device	Deep infection with hardware removal	Nonunion
		70/F	R, P	THA after 8 months	Failed rehabilitation	Nonambulatory
5	De Palma et al. [14]	27/M	R, P	Jewett plate	Union, removed plate at 1 year, re-fractured, dynamic compression plate	Union
6	Armstrong et al. [9]	1–16 (n = 3)	FN	Pins/compression screw		Union
		6–16	FN	Non weight bearing/pins	Nonunion treated with pins at 6 months	Union
		Adult (n = 3)	P	Nail plate/compression screw plate		Union
		8–30	P	ORIF		Nonunion
		8–30	P	ORIF		Union at 6 months
		31	M	P	Blade plate	Union at 12 to 16 weeks
7	Rolauffs et al. [17]	39/M	R, FN	Parallel screws	Hardware failure, osteomyelitis, Girdlestone	Girdlestone
8	Su et al. [18]	29/M	L, P	ORIF		Union
9	Chhabra et al. [13]	22/F	L, P	DHS revision after prior Jewett nail	Infection	Infection, nonunion
		22/F	R, P	DHS	Hardware failure	Pullout, nonunion
		41/F	L, P	Kuntscher nail	Rod migration with exchange nailing	Union 2 months, rod removed at 6 months
		42/F	L, P	Kuntscher nail		Union
		45/F	R, P (3 years later)	Kuntscher nail		Union
		42/M	R, P	Proximally locked intramedullary nail		Union at 2 months

reoperation was 29%, with an incidence of periprosthetic fracture of 14%.

Only one report describes the use of an external fixator for an osteopetrotic fracture, although the good result obtained suggests that this may be a reasonable option [12].

The less invasive stabilization system (LISS) was designed for the treatment of distal femoral fractures [21, 22]. It behaves as an internal fixator and hence its biomechanics are inherently different from conventional plating techniques [23]. We used distal femoral locking plates (DF-LCP; Synthes, Obedors, Switzerland) of the contralateral side of the distal femur as it matched the contour of the proximal femur on the contralateral side and had combi holes. The advantages of using a combination of two concepts – locking and compression plate fixation – with only one implant are that this combination plate can be used as a locked internal fixator to align the fragments to the shaft in a bridging manner and to achieve interfragmentary compression of simple fractures by means of a dynamic compression technique involving the placement of a lag screw through the dynamic compression unit of the plate [24, 25]. The mechanical quality of these plates gives significant advantages over other methods of fracture fixation in osteopetrotic bones.

Concerns have been raised regarding the fixation of a peritrochanteric fracture with an undisplaced fracture neck of the ipsilateral femur. Open reduction and internal fixation of a peritrochanteric fracture puts the undisplaced fracture neck of the ipsilateral femur at risk of displacement and vascular compromise unless it is also stabilized internally [26, 27]. The device used in the present study can address this problem as well, as we have the option of placing screws in the neck in many planes through the holes of this device, but we have no experience of such a fracture pattern.

Conclusion

The rarity of this condition and a lack of awareness on the part of treating surgeons can lead to the misdiagnosis of the underlying pathology of osteopetrosis in a patient presenting with long bone fracture, and subsequently poor preoperative planning and intraoperative difficulties while fixing the fractures.

Based on our experience, we would recommend the use of DF-LCP of the contralateral side for the management of fractures in dense and sclerotic pathological bones as one of the preferred modalities of treatment.

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