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Case report

Metachronous bilateral subtrochanteric fracture of femur in an osteopetrotic bone: A case report with technical note[☆]

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ABSTRACT

Osteopetrosis is a rare inherited skeletal disorder characterized by increased density. The increased fragility of such dense bone results in a greater incidence of fractures, especially around hip and proximal femur. The surgical treatment of such fractures is difficult due to hard but brittle structure of bone. Herein we report a case of bilateral subtrochanteric fracture in an osteopetrotic patient. It was fixed using a dynamic hip screw with plate.

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1. Introduction

Osteopetrosis (OP) is a disease of osteoclasts that results in failure of bone remodeling. Despite the sclerotic radiographic appearance of the thickened cortices and its material hardness, osteopetrotic bone is weak and prone to fracture by minor trauma. Areas of concentrated stress such as the

femoral neck and subtrochanteric region are especially susceptible to these fractures.^{1,2} Surgical treatment is related to risks of infection and hardware failure in such patients.³ Here we report the case of an adult patient with autosomal dominant osteopetrosis and a bilateral subtrochanteric fracture treated successfully with dynamic hip screws and plate.

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2. Case report

A 45-year-old man with known autosomal dominant osteopetrosis sustained left subtrochanteric fracture of the hip after experiencing pain and spontaneously collapsing to the floor after trivial trauma. He was treated for right subtrochanteric fracture 4 years back (Fig. 1). On examination, patient was afebrile with stable vital signs. The left thigh was tender to palpation, swollen and local temperature was not raised. The patient was unable to lift the affected lower limb and passive rotation was painful and not transmitted proximally. The pelvis was stable without tenderness on palpation. There was no distal neurovascular deficit in both lower limbs. Radiographs revealed sclerotic bones consistent with osteopetrosis. X-rays of hip and thigh showed fracture of left subtrochanteric femur with united fracture of right subtrochanteric region fixed with dynamic hip screw (DHS) which was operated 4 years back. Patient was taken to the operating room for open reduction internal fixation with a dynamic hip screw (DHS) implant with side plate and bone grafting (Fig. 2). The DHS implant was chosen as it had been effective for a similar type of fracture in the opposite lower limb in the same patient and ease of application of a surface implant than intramedullary implant in osteopetrosis. Due to the extreme bone density of osteopetrotic bone and the lack of femoral canal, the use of an intramedullary device was deferred.

The procedure was technically challenging due to high density and brittle nature of bone.

The surgical technique was modified as we had encountered broken drill bits and guide wire in the last surgery on opposite lower limb. This time we used a 3.5 mm Steinmann pin with pointed end was used to start hole of lag screw of DHS till $\frac{3}{4}$ the of neck length as checked on AP & lateral view of hip, then the guide wire of lag screw was inserted till subchondral bone of femoral head. Subsequently the hole was reamed with a new triple reamer and tapped before inserting the final lag screw of DHS. Similarly the 4.5 mm holes of the side plate of DHS were started with 2.5 mm pointed wire till the far cortex and the rest of the hole was then drilled with 3.5 mm drill bit, 4.5 mm tapped and the screws inserted. The procedure took longer than usual time because of difficulty in inserting guide wire, drilling, tapping and screw insertion in hard bone. A few guide wires were bent and many drill bits were required for the procedure. Constant irrigation and drilling pauses were used to prevent heat necrosis. Fresh autogenous iliac crest bone grafting was inserted at the fracture site. The closure of wound was done in layers. The postoperative period was uneventful. The soft tissue healed without complications and the patient was discharged on postoperative day fourteenth. Patient was followed up at regular interval. He was initially allowed toe touch walking. Partial weight bearing walking was allowed at 6 weeks. At the end of 11 months radiological examination of fractures showed good bone alignment and completed bony union with intact hardware (Fig. 3).

Fig. 1 – A: Shows the subtrochanteric fracture in the osteopetrotic bone on the right side. **B:** Postoperative AP X-ray shows fixation using DHS with side plate on the right side, yellow arrow showing the fixed subtrochanteric fracture. **C:** Four months postoperative period showing callus at fracture site.

Fig. 2 – A: Subtrochanteric fracture in the osteopetrotic bone on the left side after 4 years. **B:** Postoperative X-ray of subtrochanteric fracture in the osteopetrotic bone on the left side fixed with DHS along with the previously healed fracture of the right femur (yellow arrow in fig. part B).

3. Discussion

Osteopetrosis is a rare inherited skeletal dysplasia characterized by osteoclast dysfunction, impaired bone resorption and poor bone remodeling resulting in generalized sclerosis of bone. Osteopetrosis varies in severity depending on the time of recognition and is classified into three primary types either an

infantile malignant type osteopetrosis, inherited in an autosomal recessive inheritance pattern; “intermediate” autosomal recessive osteopetrosis; and an adult benign type autosomal dominant osteopetrosis. The adult type has its onset in later childhood and is usually discovered accidentally during a radiographic examination for another condition.⁴ Hematological abnormalities such as anemia and thrombocytopenia are common in osteopetrosis and require perioperative correction. Orthopedic presentations in osteopetrosis include periodic back pain, bone pain, recurrent fractures especially in lower limbs, degenerative arthritis and infections.⁵ The long bones are most frequently affected, fractures of the femoral neck and proximal (upper third) shaft being most commonly reported.^{1,6} Furthermore, a tendency to poor healing of bone and soft tissue related to the abnormal bone metabolism has been observed. Lack of knowledge on the part of treating surgeons and inadequate preoperative planning in osteopetrosis cases can lead to many difficulties in the fixation of such fractures. Operative intervention has a high rate of intraoperative and postoperative complications so therefore requires appropriate planning before management. Some cases in the literature have been reported in which femoral fractures are treated conservatively with good outcome. Various non-operative treatment options including hip spica plaster cast, traction, splint and non-weight bearing have been used in the past.^{5–9} Conservative management of intertrochanteric and subtrochanteric fractures can lead to delayed healing or pseudoarthrosis.^{5,9}

The surgical treatment in these case are challenging due to considerable difficulties during procedure complications. The brittleness and hardness of the osteopetrotic bone and the bony obliteration of the marrow cavity are the risks of iatrogenic fracture. The fragility and hardness of the bone reduces its ability to hold screws and insertion of nail.³ The surgeons should be cautious during drilling and reaming. Frequent change of drill bits, regular cleaning of the tap and wash out of the screw tract and periodic cooling of the bone to prevent bone necrosis and subsequent implant loosening and infection need to be done.^{3,10}

Fig. 3 – Postoperative X-ray after 11 months showing union.

We feel that our use of 3.5 mm and 2.5 mm pins to start the holes prior to drilling greatly reduced the incidence of bent guide wires, broken drill bits as compared and encountered in previous surgery done on opposite side. It can thus be extrapolated that self-drilling, self-tapping screws with pointed sharp tips may be easy and fast to insert with a screw bit.

Any use of mallet risks fracture of the brittle bone, and therefore should be avoided. Although all such precautions can lead to prolonged operating time and increases the anesthetic risks to the patients.¹⁰ The various operative treatment methods for treatment of subtrochanteric fractures in osteopetrotic patients have been described in literature with different outcomes. The authors have used nail plate devices, DHS, distal femoral locking plates, nails (Zickel, Jewett, Kuntscher, proximally locked), total hip arthroplasty and external fixator for fixation of such fractures.^{5,11–14} Some authors preferred nails (load sharing implant) over plates and screw devices (load bearing) for treatment of long bone fractures of osteopetrotic bone as screw holes and plate ends create areas of increased stress and are prone to failure because of the increased demand placed on them during union.¹⁰ An alternative for reducing the stress in plate fixation is the use of a locking plate.

There is no single method of fixation which is superior for fixation of such fractures so therefore treatment method in these cases should be individualized depending upon expertise of surgeon and availability of surgical resources. There are various postoperative complications reported in the literature including broken plates and screws, distal fractures due to nail placements, periprosthetic fractures, infections, delayed union and migration of implants. Recently Srivastava et al reviewed all operative cases of (25 previously reported cases including 4 neck femurs and 21 pertrochanteric regions) fractures in osteopetrotic patients published in the literature and among them the nonunion rate was found to be 12% and the infection rate was 12%. In the pertrochanteric group, the rate of hardware failure was 29% and the rate of revision surgery was 29%, with an incidence of periprosthetic fracture of 14%. They 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.¹⁵

We selected and used DHS for our patient, the foremost reason being it had been successful in healing of a similar fracture type in the opposite lower limb in the same patient. The other reason being its sliding mechanism allows some axial loading of subtrochanteric fracture, though to a lesser extent than dynamic interlocking nails as the lag screw plate sliding mechanism is at an angle to the femur bone. This axial loading is important in such bones as they have reduced capacity to remodel and heal, and need physicochemical stimulus like loading to help in fracture healing. The fact that we achieved good results despite the difficulties of drilling and reaming the bones in same patient is good enough reason not recommendation to have used DHS.

4. Conclusion

The treatment method for treating osteopetrotic patients should be individualized. Although conservative treatment is

a good option for treating such cases, surgical treatment should be used in adults especially in high stress subtrochanteric area of femur as it gives good functional result. Meticulous preoperative planning with anticipation of forthcoming technical difficulties is essential in the management of fracture. DHS a universal and convenient to use implant, with some modification in technique can be used to fix such fracture. Close monitoring of patients during rehabilitation is required to detect complications.

Conflicts of interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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