Case report

An unusual presentation of patella and fifth metatarsal base fractures in a patient with osteopetrosis

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1. Introduction

Osteopetrosis is a rare heterogeneous group of inherited bone dysplasias characterized by diminished osteoclast mediated bone resorption and leading to increased bone density. Also known as marble bone disease, it was first described by Albers Schönberg, a German Radiologist in 1904.¹

The most common clinical presentation of Autosomal Dominant Osteopetrosis is frequent fractures in approximately 75% of the patients. Other manifestations include hip osteoarthritis, facial nerve palsy and mandibular osteomyelitis.² However, 20–40% of the patients remain asymptomatic.

Management of these fractures is associated with significant complications, both intra and post-operative. To add up, the variable healing response of the bone poses a significant challenge to the treating orthopaedic surgeon. Most of the fractures in these patients involve long bones³ (femur and tibia). There are rarely any cases reported in literature of fractures involving flat or sesamoid bones in these patients.

In this report, we describe the unusual presentation of fractures of the patella and fifth metatarsal base in a patient with osteopetrosis.

2. Case report

A 40 year old gentleman with a known history of Autosomal Dominant Osteopetrosis and previous multiple fractures, sustained an injury to his knee due to fall while skiing. He had tenderness and swelling in the anterior aspect of his knee but an intact extensor mechanism. The X-ray showed a minimally displaced fracture of his patella. The patient was managed non-operatively with a cylindrical cast in view of the minimal displacement and intact extensor mechanism. Three weeks later, he sustained a twisting injury to his foot and was diagnosed to have an undisplaced fracture of the 5th metatarsal base. The metatarsal fracture was also managed non-operatively. Subsequent X-rays done after 2 months of the index injury showed no evidence of callus formation or bone healing (Figs. 1 A, 2 A). After discussion with the patient, it was decided to surgically fix both the fractures.

The patella fracture was fixed with 4.0 mm partially threaded cancellous screws and the fifth metatarsal with a 2.7 mm locking plate. Despite the transverse pattern of the patella fracture, tension band wiring was not chosen because we were wary that the wires might not penetrate the sclerotic bone. Intra-operatively, there was fibrous tissue at the fracture site, no evidence of healing and a complete absence of cancellous bone in the patella. A new drill was used for drilling each hole and a high power drill system was used in this case. The drilling and tapping of the bone was uneventful. However, one of the screws broke when it was inserted halfway into the inferior pole of patella. The retained portion of the screw was buried in the bone and a decision not to remove the screw was taken. Another screw could not withstand the high torque needed to insert the screw. For fear of another screw breakage, it was

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Fig. 1. Pre- (A) and post-operative (B) radiographs of the right knee. Post-operative radiographs show one broken and one partially inserted screw.

decided not to fully insert it. One screw was successfully inserted fully (Fig. 1B). The fifth metatarsal fracture was fixed with locking plate (Synthes 2.4/2.7 mm LCP). This procedure was uneventful (Fig. 2B). Intravenous cefazolin for 24 h was administered to the patient as per hospital protocol.

The patient was kept non weight bearing on his left foot in the post-operative period for 6 weeks followed by progressive weight bearing. His right knee was kept in an articulated knee brace allowing 0–90° of motion initially. The immediate post-operative period was uneventful with good recovery. However, at 5 months, he developed an abscess on his left foot near the surgical scar for which he underwent an operative debridement and 6 weeks of intra-venous cefazolin as recommended by Infectious Disease Physicians. The implants were not loose and hence were retained. The wound healed eventually. He was kept on oral suppressive cloxacillin until 3 weeks post implant removal which was performed 1 year post fixation. At 1 year follow-up, both his fractures had healed clinically. Minimal callus was seen in the patella fracture though the 5th metatarsal fracture showed complete bony bridging (Fig. 3). He was ambulating well for long distances without aid and his knee range of motion was 0–130° of flexion.

3. Discussion

Fractures in osteopetrosis typically affect long bones, especially the proximal femur, tibia, humerus and foot bones. Some uncommon sites like clavicle,⁴ olecranon and distal humerus^{5,6} have been described as well. Though diaphyseal and metaphyseal fractures in long bones are common, fractures involving flat bones or small metaphyseal bones are uncommon. To the best of our knowledge, there is no published report in literature on patella fractures managed surgically in patients with osteopetrosis.

Management of fractures in patients with osteopetrosis has always been a challenge for the orthopaedic surgeon. Intra-operative complications include difficulty in drilling the bone due to sclerosis, frequent breakage of drill bits and implants and problems with inserting intramedullary nails.⁷ The amount of heat generated during drilling of bone is associated with thermal necrosis of the bone. Many intra-operative measures have been described previously to minimize these intra-operative complications.⁸ Frequent change of drill bits,⁸ use of slow speed high torque drill bits, metal cutting bits, high speed burs, periodic cooling of bones, avoiding intramedullary implants and predrilling before k wire insertion⁸ are some of the strategies that are recommended while fixing these fractures.

Standard surgical management of transverse fractures of the patella is tension band wiring. However, in view of possible difficulty encountered in drilling a K-wire through hard sclerotic bone, use of an alternative technique of screws⁹ for fixation was chosen. We used non-cannulated screws to avoid drilling a guide wire for cannulated screws. Our fixation was complicated by one screw breakage and one incomplete screw insertion. As we used high power drills and a fresh drill bit for each drilling, none of the drill bits broke during the procedure.

The fracture of the fifth metatarsal was an undisplaced Zone 2 fracture, as described by Torg.¹⁰ The standard management of this fracture is non-operative and that was our first option in this case. However as the patient was to undergo surgery for his patella, the metatarsal fracture was also fixed after discussion with the patient. We used locking screws to avoid the step of using a tap and thus minimize instrument breakage. However, it was noticed that the torque limiting screwdriver could not be used to tighten the screw as the force needed to insert the screw was much higher than the torque required to lock the locking screws. As a consequence, a standard screwdriver had to be used to tighten the locking screws.

Fig. 2. Pre- (A) and post- operative radiographs (B) radiographs of knee and left foot.

Fig. 3. Final post-operative X-ray. Note absence of complete bone bridging in the patella fracture.

Fractures in osteopetrotic bones are associated with a high incidence of delayed union, non-union, abnormal bone healing and osteomyelitis in the post-operative period. The bone healing is abnormal due to the absence of bone remodeling. Molecular defects in osteoclasts cause an imbalance between bone formation by osteoblast and resorption by osteoclasts, leading to sclerotic brittle abnormal bone.¹¹ Although there is decreased bone resorption, the number of osteoclasts is elevated because of upregulation in the absence of bone resorption. Absence of ruffled border in osteoclasts on electron microscopy explains the dysfunctional nature of osteoclasts in osteopetrosis.³ DePalma et al.¹² studied the fracture callus in a patient with Autosomal Dominant Osteopetrosis. According to them, the fracture callus was more or less normal in initial stages and composed of a rich bone-forming cell component. However, the tissue showed no Haversian organization a year later.

In our patient, the fractured patella did not show any appreciable callus formation or bridging of bone despite the patient being asymptomatic with full function at 1 year. Though most of the fracture callus description focuses on diaphyseal fractures, nothing has been described about fracture healing in sesamoidal bones in fractures with osteopetrosis. We were not able to explain absence of callus and bony bridging in the fractured patella post-operatively.

Osteomyelitis is also common in these patients because of absence of white cell function and diminished vascular supply.¹³ Our patient sustained an abscess formation with a possible osteomyelitis of the fifth metatarsal. The wound healed uneventfully after one debridement and 6 weeks of intravenous antibiotics. We kept him on oral suppressive antibiotics till the implant was removed one year after surgery.

4. Conclusion

Our case reports the first case of fracture of patella fixed surgically in a patient with osteopetrosis and explains the dilemma and difficulty encountered by the authors in fixing this common fracture in an uncommon condition.

Conflict of interest

None.

Source(s) of support

Nil.

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References

1. Albers-Schönberg HE. Röntgenbilder einer seltenen Knochenerkrankung. *Munch Med Wochenschr.* 1904;51:365–368.
2. Shapiro Frederic. Osteopetrosis: current clinical considerations. *Clin Orthopaedic Relat Res.* 1993;294:34–44.
3. Milgram JW, Jasti M. Osteopetrosis: a morphological study of 21 cases. *J Bone Joint Surg Am.* 1982;64:912–929.
4. Nakayama J, Fujioka H, Kurosaka M, Kitazawa H, Maesawa N, Tomioka M. Surgery for clavicular and humeral fractures in an osteopetrosis patient: a case report. *J Orthop Surg.* 2007;15(2):251–254.
5. Calvin C, Rajesh B, Richard DL. A case report: treatment of a medial condylar humerus fracture in an adult with osteopetrosis. *Internet Med J.* 2012;12:.
6. Miguel AF, Carlos MO, Rodrigo FP, Sebastian S, Linda V. Recommendations for fracture management in patients with osteopetrosis: case report. *Arch Orthopaedic Trauma Surg.* 2015;135:351–356.
7. Rakesh G, Nikhil G. Femoral fractures in osteopetrosis: case reports. *J Trauma Inj Infect Crit Care.* 2001;51:997–999.
8. Amit B, Manesh V, Lennox CME. Challenges in the management of fractures in osteopetrosis! Review of literature and technical tips learned from long-term management of seven patients. *Inj Int J Care Injured.* 2009;40:1167–1171.
9. Berg Eugene E. Open reduction internal fixation of displaced transverse patella fracture with figure-eight wiring through parallel cannulated compression screws. *J Orthop Trauma.* 1997;11(November (8)):573–578.
10. Torg JS. Fractures of the base of the fifth metatarsal distal to the tuberosity. *Orthopedics.* 1990;13:731–737.
11. Jakub T, Steven LT, Paul JO. Osteopetrosis review article. *New Engl J Med.* 2004;351:2839–2849.
12. De Palma L, Tulli A, Maccauro G, Del Torto M. Fracture callus in osteopetrosis. *Clin Orthopaedic Relat Res.* 1994;308:85–89.
13. Armstrong DG, Newfield JT, Gillespie R. Orthopaedic management of osteopetrosis: result of a survey and review of literature. *J Pediatric Orthopaedics.* 1999;19:122–132.