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Peri-Implant Fracture In Osteopetrosis – A Case Report

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ABSTRACT

We report a case of osteopetrosis complicated by a left proximal femur peri-implant fracture with broken implants. The patient underwent implant removal followed by open reduction and internal fixation using a contralateral distal femur locking compression plate. Osteopetrosis is characterized by increased bone density and brittleness, making fracture fixation challenging. Preoperative planning is essential to minimize operative time and reduce intra-operative complications, which are more likely due to the unique bone quality. During surgery, careful consideration of the bone's brittleness and hardness guided the approach, emphasizing the importance of gentle manipulation and appropriate selection of implants. Conventional plating techniques remain the cornerstone of fracture management in osteopetrosis, but the unique challenges of this disease require the surgeon to be vigilant about potential hardware failure. Fracture union was eventually achieved at 18 months, illustrating the prolonged healing time often seen in this rare metabolic bone disorder. The extended period to union highlights the need for intensive postoperative care and regular monitoring to detect potential complications early. This case underscores the necessity of individualized surgical strategies and the importance of a multidisciplinary approach to managing osteopetrosis-related fractures. Postoperative rehabilitation and patient education play crucial roles in ensuring optimal outcomes in these patients.

INTRODUCTION

Osteopetrosis, a disease characterized by a group of skeletal abnormalities was first described in 1905 by the German radiologist, Heinrich Albers-Schonberg. It is a developmental abnormality resulting from the osteoclast dysfunction. Osteoclasts does the function of resorption and remodeling. Because of the defect in this process patient experience excess bone growth leading to functional and structural defect in skeletal anatomy. In spite of the dense body the bone is brittle and week because of the absence of remodeling due to osteoclast dysfunction and insufficient bone marrow development[1-6]. This ultimately leads to the common complication of fracture and the management of this rare entity is challenging. Fractures are most commonly seen in long bones. If the fracture is left untreated the fracture will go for varus malunion or non-union. Cases are reported using locking plate to treat an osteopetrotic femoral fracture, even then resulting in nonunion. Hence surgical management is indicated for this rare disease.

STATEMENT OF INFORMED CONSENT

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

CASE REPORT

A 40 Year old female came to OPD with alleged history of slip and fall while walking and sustained injury to her left thigh (trivial trauma). Post injury patient was not able to weight bear in her left lower limb.

Patient gives a similar history 10 years back for which she underwent Open Reduction and Plating in the same limb.

X-ray skull lateral view revealed diffuse thickening of the facial bones, calvarium most significantly in the region of the frontal and occiput. Her blood investigations had a marginal increase in alkaline phosphatase, with normal calcium, magnesium and phosphate values.

Patient was diagnosed as post traumatic left proximal femur fracture with implant breakage with mal-united right proximal femur fracture and non-union of right clavicle fracture with Osteopetrosis.

Patient's children were screened for Infantile Recessive Osteopetrosis and were found to be safe.

LOCAL EXAMINATION OF LEFT THIGH:

- 15 cms healthy surgical scar present in lateral aspect of proximal end of left thigh.
- Swelling & Tenderness present over junction of proximal and middle 1/3rd junction
- Abnormal mobility & Bony crepitus present
- Loss of transmitted movements present
- Bony discontinuity present
- Left Hip ROM- Painful and restricted
- No Distal Neurovascular deficit.

Xray pevlis with both hips showing Left proximal femur fracture with Implant Breakage with Mal-united right proximal femur fracture.

Full length bilateral lower limb xray

Chest xray pa view showing Non-union of Right Clavicle.

PROCEDURE

Patient was planned for implant removal and femur nailing. Intra-operatively we had difficulty in removing the broken implants. There were screws breakage which was left in-situ and the broken plates were removed with great difficulty. Patient was then planned for TENS nailing, which also became vain because of difficulty in drilling. After the above failed attempts patient was proceeded with open reduction and internal fixation with contralateral distal femur locking compression plate to proximal femur boycotting the broken screw. Post operative period was uneventful.

Post operative xray

POST OP PERIOD

Patient was on regular follow and she obediently followed her physiotherapy. There were no signs of union till 1year period of follow up post-surgery and was on walker support. Patient was intervened non operatively with bisphosphonate therapy. Patient finally showed signs of union by 18 months and complete union was achieved by 2 years from the day of surgery. She is now asymptomatic and has resumed her day-to-day activities without any fresh fractures and without the use of walker support. Fracture union was found to be satisfactory with these 2 years follow up with a good clinical outcome.

Post operative xrays at 2 years follow up

Clinical images at 2 years follow up

DISCUSSION

Osteopetrosis is a rare hereditary skeletal disorder in which there is an increased bone density with a defect in remodeling, where bone resorption is also impaired due to the defect in osteoclast activity. This makes the bone fragile, hence it is also called as brittle bone disease, marble bone disease. This disease is further classified as Infantile autosomal recessive osteopetrosis and Adult autosomal dominant osteopetrosis. Based on genetic mutations they are further classified in Adult dominant osteopetrosis. There is also a delay in consolidation because of failure in remodelling[7]. In a study, by Birmingham et al. there an infection rate of 12 %, non-union rate of 12% and an implant failure rate of 29% in their peri-trochanteric group[8]. Kumbaraci et al. managed bilateral proximal femur fractures with proximal femoral nail anti-rotation but used regular femur nail in pediatric population who had medullary canal[9]. Since we had a little medullary canal in our patient we attempted for TENS, which failed after prolonged drilling. Still there are case reports where Dynamic Hip screw was fixed for sub-trochanteric fractures in this population, in spite of having difficulty in drilling[10]. There are multiple studies where the fractures were managed by Locking plates, augmented with BMP, Allografts[11,12].

There are also other case reports where the plate fixation was added with meshed fresh frozen femoral head. Fracture union was achieved by 6 months after the surgery[7]. Only in Bhargav et al, study there was a locking plate used to achieve osteosynthesis for a femoral fracture with subsequent nonunion in a

patient with osteopetrosis. Certain studies recommended bone morphogenic protein grafting as bone substitute, which provokes mesenchymal cells to induce its osteo inductive properties for cell differentiation, thereby exhibiting appropriate effect on bone and callus leading to fracture healing[11]. Hence pre-operative planning minimises the duration of the operation and the risk of intra-operative complications. We must consider the brittleness and hardness of the bone during fixation. The principle in managing osteopetrosis fracture is byconventional plating. Fracture union was achieved at 18 months in our case. The longer time to union demand intensive postoperative care in this rare metabolic bone disease. Having a great difficulty in surgical stabilization in osteopetrosis, further research is necessary to elucidate the optimal form of treatment in this rare patient population.

CONCLUSION

Preoperative planning minimises the duration of the operation and the risk of intra-operative complications. We must consider the brittleness and hardness of the bone during fixation. The principle in managing osteopetrosis fracture is by conventional plating. Fracture union was achieved at 12 months in our case. The longer time to union demand intensive postoperative care in this rare metabolic bone disease.

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