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Case Report

Osteopetrosis Case Report of BL Femur FX with Low Energy Trauma, Case Report and Literature Review

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Abstract

Osteopetrosis *is* a type of bone metabolism disorder that causes osteoclasts to fail properly, leading to impaired bone resorption and poor bone remodeling.

The primary orthopedic presentations and the most common manifestation in patients with osteopetrosis fractures, especially long bones

The literature has described several operative methods to treat osteopetrotic subtrochanteric fractures.

Our study reports on the results of an internal fixation procedure performed on a patient who is known to have osteopetrosis who had a bilateral proximal femur fracture.

Keywords: Osteopetrosis, Bilateral proximal femur fracture, long bone fracture, Infection

Introduction

Osteopetrosis (OP) is a rare inherited skeletal condition resulting from osteoclast dysfunction, which leads to a lack of bone resorption and an increase in bone density. This disorder is characterized by hard bones, however prone to breakage, and often has a narrow or completely absent medullary canal [1-2]. Osteopetrosis is categorized into three types: autosomal dominant OP (ADO), intermediate autosomal OP (IAO), and autosomal recessive OP (ARO) based on the level of severity and inheritance mode [3].

The primary orthopedic presentation in patients with osteopetrosis are fractures, scoliosis, hip osteoarthritis, and osteomyelitis. The most common manifestations, observed in 75% of cases, are frequent fractures primarily affecting the long bones [4]. The most commonly fractured long bones are the upper third of the femur bone and the upper third of the tibia bone [5]. The problem in patients with OP does not arise from the bone-healing ability of osteocytes, as it is comparable to that of individuals without this condition. Studies showed that benign osteopetrosis typically takes around 12 months for fracture union [6]. However, the problem arises from the nonfunctional osteoclasts, leading to a lack of a remodeling phase [6].

The orthopedic surgeon always faces significant challenges due to the unpredictable nature of bone remodeling. Special care must be taken when treating fractures in patients diagnosed with OP [4].

Currently, there is a lack of consensus regarding the preferred approach for treating fractures in patients diagnosed with

osteopetrosis, whether it be through conservative or surgical methods [7]. The majority of fractures occurring in patients with osteopetrosis were commonly managed through conservative treatments, such as skeletal traction and plaster fixation. This approach is favored due to the increased susceptibility to infection with operative intervention in these individuals [8]. However, conservative methods to treat intertrochanteric, subtrochanteric, and femoral neck fractures can lead to complications such as coxa vara deformities, delayed bone union, and nonunion [9].

There are few cases reported in the literature of peritrochanteric/subtrochanteric fractures, most of which are asynchronous bilateral subtrochanteric fractures; however, only six cases of which are of bilateral Synchronic subtrochanteric fractures [1,10-12]. Management of peritrochanteric fractures in those patients is challenging, with an emphasis on the ongoing debate and the controversy surrounding the best modality to treat these fractures. Further, Simultaneous subtrochanteric fractures complicate the matter more [1,10-12].

We report a case of an osteoporotic patient with a bilateral Proximal femur fracture resulting from low-energy trauma and treated with plate and screws bilaterally.

Herein, we present the clinical and radiological findings within a comprehensive literature review and management recommendations

Case Presentation

Patient Information

A 28-year-old patient presented to the emergency department after a history of falling on the top of a car around 2 meters in height.

The patient presented complaining of bilateral thigh pain with the inability to bear weight.

Clinical findings

The patient was conscious, alert, and oriented on physical examination, with a Glasgow Coma Score of 15/15 and vitally stable. The patient presented with deformity in both thighs, tenderness bilaterally, and a reduced range of movement. The patient had normal neurological exams, distal pulses, and perfusion.

Diagnostic evaluation

An X-ray of the bilateral thigh AP and lateral and a pelvis X-ray showed a bilateral femur fracture (Right proximal third femur fracture and Left Subtrochanteric femur fracture) (Figure 1).

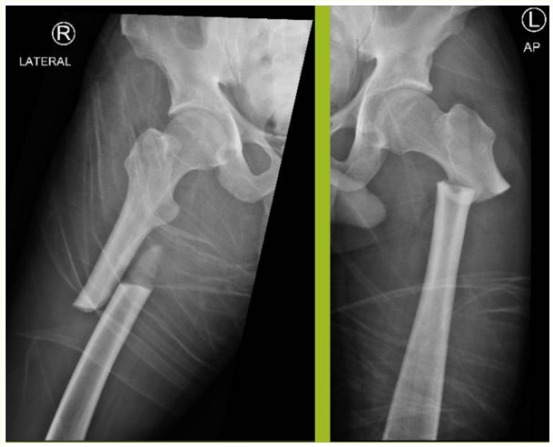

Figure 1: Bilateral femur fracture (Right proximal third femur fracture and Left Subtrochantric femur fracture).

Both sides of the distal femur were affected by an "Erlenmeyer flask deformity," as seen on the radiographs (Figure 2).

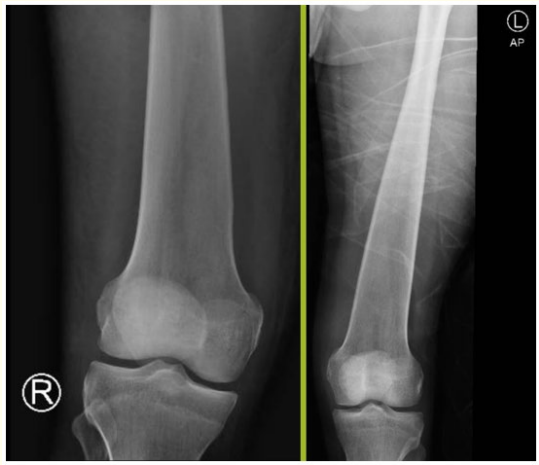

Figure 2: "Erlenmeyer flask deformity" of bilateral distal femur.

An investigation of laboratory parameters revealed normal levels of thyroid hormones and alkaline phosphatase and a negative sickling test. On the Skeletal survey, there was no evidence of other skeletal deformities. Before this injury, the patient had not suffered from fractures or recurrent infections.

Initial therapeutic intervention

Initially, bilateral skin traction was applied, and surgical treatment was indicated. The patient consented to open reduction and internal fixation with a plate and screws.

We started with the right side with a trial of close reduction with an intramedullary nail because the right femur was less sclerotic than the left side. As expected, the nail did not pass through the bone, so we proceeded with the surgery utilizing a lateral approach for both sides with plates and screws (Figure 3). Types of plates (Right proximal femur fixation done by (LC-DCP: Limited Contact Dynamic Compression Plate and Left subtrochanteric fracture was fixated with proximal femoral locking plates).

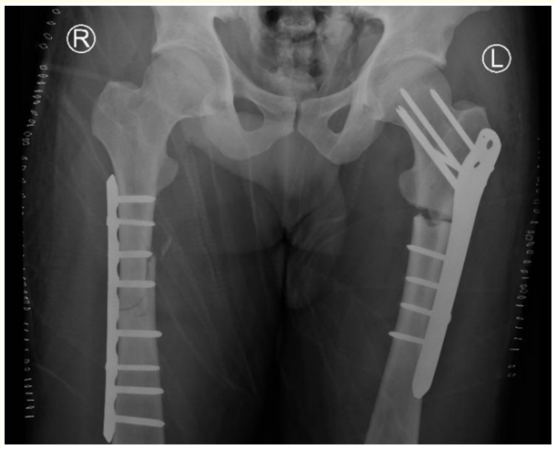

Figure 3: Post-operative x-ray with bilateral proximal femur fixation with plates and screws.

Postoperative course

The patient had an uneventful postoperative course. He was advised and instructed for non-weight bearing mobilization on the lower limbs and allowed for mobilization using a wheelchair for eight weeks.

Follow-up Visits and Assessment of Outcomes and Interventions

The patient presented after two weeks of surgery for clip removal. The wounds were clean at that time, with no signs of infection or discharge after 15 weeks of follow-up, signs of healing and callus formation were seen in the x-ray. Week 20th post-surgery, the Patient started to have surgical site infection with minimal discharge from the Left side, on the proximal site of the wound; daily dressing with broad-spectrum oral antibiotics was given to the patient for two weeks but with no improvement. On week 22, the patient was admitted and Operated on with the removal of the left femur implant (had soft tissue and necrotic bone positive culture, treated with appropriate course of antibiotics). At week 33, the infection resolved, the patient completed his full course of antibiotics, and his X-ray of the Left femur showed healing and callus formation (Figure 4). The patient mobilized with non-weight bearing for 6 weeks followed by mobilization as tolerated with crutches.

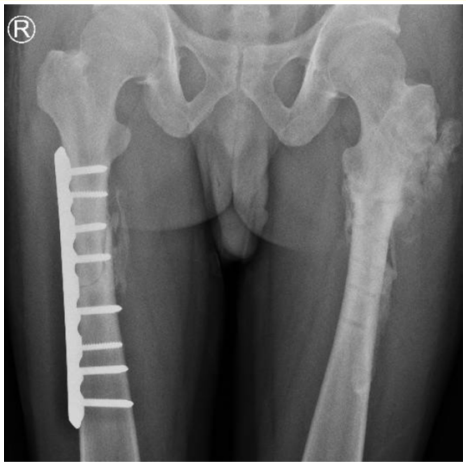

Figure 4: X-ray of the Left femur showed healing and callus formation after 33 weeks post-initial surgery.

Discussion

Multiple studies over the years have published case reports, extensive reviews, and a summary of the literature on femur fractures in OP patients [1,10-12]. In their cumulative review, approximately 57 cases of peritrochanteric fracture were presented [1,10-12].

Dual-energy X-ray Absorption DXA is a more accurate method of measuring Bone Mineral Density (BMD) than conventional

radiography. Paccou J., *et al*. 2018 would involve an ideal definition of the Z-score in BMD as +2 at both the spine and hip, where Z-scores exceeding + 6 can be considered extremely high [2]. Our patient had a Z-score of + 5.9 on the spine, which corresponds to a higher result compared to his age group, On the other hand, his hip was excluded due to fixation done before shifting him to do the DEXA (Figure 5). Patients with osteopetrosis cannot accurately predict fracture risk with this test. Nevertheless, it will provide a clue for some patients who were never diagnosed due to normal clinical and laboratory results.

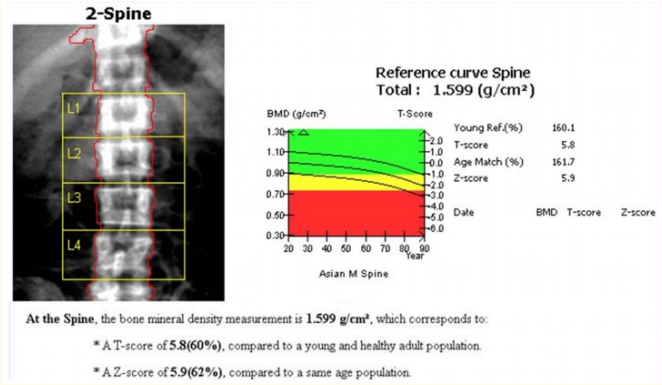

Figure 5: Bone Mineral Density (BMD) showing a Z-score of + 5.9 on the spine region.

Management options for operative peri trochanteric fracture used in the reported cases included Dynamic hip screws (DHS), dynamic condylar screws (DCSs), intramedullary nails (IMNs), proximal femoral anti-rotation intramedullary nails (PFNAs), locking compression plates (LCPs) and total hip arthroplasty (THA) [1,11]. Conservative treatment was by immobilization with restricted weight-bearing, plaster cast, or traction [12].

Birmingham P and McHale KA 2008 [12] reviewed 38 cases of femur fractures from 1954 until 2008 literature, 25 of which were treated operatively [12]. Thirteen cases were treated conservatively, three of which were of femur neck fractures and ten cases of peri trochanteric fractures; by direct comparison, they observed a lower complication rate when compared to operative treatment. The most common complication of conservative treatment was coxa vara deformity in 31.0% of cases, the majority of which were with femoral neck fractures and a nonunion rate of 7.6%.

A more recent study discussing the operative management of peritrochentric fractures among those patients conducted by Ding., *et al*. 2021 reported five cases from their institute and 32 more cases from the 1954-2018 literature [11]. Their review also included six cases of femoral neck fractures, three cases of femoral shaft fractures, and a total of 50 operations [11].

The complication rate was as follows: (1) a 6.0% nonunion rate, (2) a 6.00% infection rate that failed to achieve union, (3) a hardware failure rate of 16.00%, and (4) a periprosthetic fracture rate of 6.0% [11].

They observed the tendency of recent literature towards favoring operative options as more advances and understanding of challenges encountered when treating OP patients and how to overcome them led to a decrease in the complications rate reported. To support their claim, they compared cases published before 2005 to cases published after with a complication rate of 54.55% vs. 21.05% respectively, a reoperation rate of 27.27% vs. 21.05% respectively, and a nonunion rate of 18.18% vs. 10.53% respectively [11].

Hua X., *et al*. 2020 study summarized literature regarding the operative management of osteopetrosis subtrochanteric fracture from 2010 to 2020; from their review and experience, they recommended the option of open reduction and internal fixation, taking into consideration the long healing process OP patients encounter [1]. Furthermore, they highlighted the importance of keeping in mind the possible difficulties encountered that arise from the poor bone blood supply and relative osteopenia, the increasing susceptibility of OP patients to infection, and osteomyelitis, as our patient got his infection after 22 weeks of the surgery. The increased density and stiffness of osteoporotic bone increased susceptibility to drill bit breakages and iatrogenic fractures [1]; as we tried to apply the nail to the right femur but could not, it was a challenging and exhausting process.

However, with extra care and knowledge on methods to overcome such complications and how to deal with them when encountered, the operation can become a feasible option [1,10-12].

In patients with osteopetrosis, fracture calluses have been demonstrated to be abnormal, with unorganized woven bone and a lack of lamellar organization even after healing [13], as seen in figure 4.

After removing the left side implant and receiving the appropriate antibiotic, our patient resumed full activity on both sides. He remained free of further complications.

Conclusion

Our study reports on the results of an internal fixation procedure performed on a patient who is known to have osteopetrosis and had a bilateral femur fracture. Orthopedic surgeons have to keep in mind the postoperative complications for patients with OP. We also emphasize the need to consider the challenging and exhausting process intraoperatively from the osteoporotic bone, which has increased density and stiffness. A narrow medullary canal and hard fragile sclerotic bones can cause drill bits or screws to bend during surgery. A single fixation method can never be preferred to fix such bone fractures. Choosing the most appropriate method is impossible, so the treatment should be based on the surgeon's expertise and surgical resources.

Disclosure of Interest

The authors declare that they have no conflicts of interest concerning this article.

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