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Dynamic condylar screw in subtrochanteric fracture with osteopetrosis: A case report

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Abstract

Osteopetrosis is a rare hereditary disease which is characterized by increased bone density and it usually presents with increased bone mass and generalized osteosclerosis. We report the case of an adult patient with marble stone- osteopetrotic subtrochanteric fracture, which was successfully treated with Dynamic cortical screw in the light of a comprehensive literature review.

Keywords: Dynamic condylar screw, subtrochanteric fracture, osteopetrosis

Introduction

Osteopetrosis is a rare hereditary disease which is characterized by increased bone density and it usually presents with increased bone mass and generalized osteosclerosis [1]. The term osteopetrosis is derived from the Greek 'osteo' meaning bone and 'petros' stone. It was first described in 1904 by Albers-Schonberg, a German radiologist [2].

There are three clinical forms based on the age of onset, inheritance pattern, and clinical features: (i) infantile or malignant osteopetrosis, (ii) intermediate, and (iii) adult onset or benign osteopetrosis. Osteopetrosis tarda, which is also known as marble bone disease, is a subtype of autosomal dominant osteopetrosis type II (ADO type II) [3]. Osteopetrotic bone is weak and prone to fracture by minor trauma, despite the sclerotic radiographic appearance of the thickened cortices and its material hardness, and the areas of concentrated stress such as the femoral neck and subtrochanteric areas are especially susceptible [4,5].

The classic radiological features of osteopetrosis comprise diffuse sclerosis, affecting the skull, spine, pelvis and appendicular bones, medullary compartment often obliterated by immature unresorbed bone, bone modeling defects at the metaphyses of long bones, such as funnel – like appearance (Erlenmeyer flask deformity) and characteristic lucent bands, bone- in-bone appearance particularly in the vertebrae and phalanges, focal sclerosis of the skull base, pelvis and vertebral end plates-sandwich vertebrae and “rigger-jersey” spine and transverse fractures [6]. There is ample amount of literature regarding treatment of osteopetrotic fractures concentrates on that of children or on the difficulty and the weaknesses of operative intervention in adults.

We report the case of an adult patient with marble stone- osteopetrotic subtrochanteric fracture, which was successfully treated with Dynamic cortical screw in the light of a comprehensive literature review.

Case Report

The aim of our study is to report a case of a 35 year female with subtrochanteric fracture referred to our hospital with the complaints of hip pain and the inability to walk, which occurred after a minor fall which the patient sustained 15 days back after a trivial fall from the bed, based on the patient history. There is history of multiple previous fracture, once in her tibia, which was managed conservatively and then it went into malunion (Figure 1).



Fig 1: Old fracture of tibia in a case of osteopetrosis showing sclerotic bone treated conservatively and later malunited

Also there is a history of left subtrochanteric fracture around two years back, which was treated conservatively and it also resulted in malunion.

Physical examination revealed pain with palpation as well as loss of hip range of motion. The extremity was externally rotated and shortened. Movement elicited pain in the area of the right hip. Radiographs of the right femur revealed a fracture of the subtrochanteric region and of the spine revealed multiple osteopetrotic changes (Figure 2, 3).



Fig 2: Subtrochanteric fracture of right side 15 days old. X Ray plate shows sclerotic and thick femur with absence of medullary canal.



Fig 3: X Ray spine shows case of osteopetrosis sclerotic subend plate changes at multiple level giving sclerotic-lucent-sclerotic appearance

Ocular and neurological and other systemic examinations showed normal vital findings. Laboratory test results were within normal limits including the serum calcium and alkaline phosphate levels while x ray showed subtrochanteric fracture. Based on the patient history, physical examination findings, and laboratory and imaging test results, along with consultation remarks, the patient was diagnosed with osteopetrosis tarda and osteopetrotic femoral proximal fracture.

The patient was put on traction and advised to undergo surgery in view of the nature of the fracture and the requirement of early ambulation. Patient was then taken up for surgery and was then originally planned for proximal femoral nail (intramedullary device) but while awling, the bone was too hard and the awling failed and so this procedure was abandoned. Patient was then planned for extra medullary fixed angle plate. The DCS along with cortical screws was chosen to circumvent these problems. The fracture was reduced on a traction table after opening the area. Fixation was held with a dynamic condylar screw with three drill bits breaking while applying cortical screws. Fluoroscopic assessment revealed an adequate reduction and fixation. No perioperative complications were observed.

In the postoperative period, there was no scar related complication, either. Postoperatively, quadriceps exercises were encouraged on the first postoperative day. Range of motion exercises were started within the limits of pain. X Ray done showed adequate reduction and fixation (Figure 4).



Fig 4: Bilateral subtrochanteric fracture. Left side fracture had occurred two years ago and was treated conservatively resulted in malunion .Right side was managed by surgical intervention

The histopathological examination confirmed the diagnosis of osteopetrosis. No complications including infection, nonunion, or avascular necrosis (AVN) were observed in the follow-up period.

Discussion

Osteopetrosis, which is a group of conditions, is a heterogeneous hereditary disease characterized by significantly increased bone density due to osteoclast dysfunction. Multisystemic involvement like anemia, pancytopenia hepatosplenomegaly, renal tubular acidosis, compressive nerve palsies and mental retardation needs modified anaesthetic technique. Facial deformities like proptosis, high arched palate, broad facies, hypertelorism, mandibular hyper or hypoplasia, limited mandibular movements make oral intubation difficult while bony encroachment of nasal turbinates may preclude nasal intubation ^[7]. Autosomal dominant osteopetrosis is the most common form with an estimated prevalence of 1 in 20,000 births. Age of onset is late childhood or adolescence ^[6]. The life expectancy of patients with intermediate osteopetrosis is moderately reduced, whereas adult patients with benign ADO has a normal life expectancy ^[8]. In adult benign ADO, bones are prone to fractures due to increased bone density and sclerosis with an increased rate of hip and proximal femoral fractures in type II (a phenotypic variant of Adult ADO). Several case reports and small-scale case series are available in the literature and review of the literature revealed case reports in which conservative treatment modalities were used; however, procedure-related complications including nonunion and coxa vara were also reported ^[9]. Technical difficulties include bending of drill bits or screws during surgery using drilling or carving due to hard fragile sclerotic bones and a narrow medullary canal. It should be kept in mind that drilling hard bones and internal fixation may complicate surgery in patients with osteopetrotic fractures ^[10]. Subtrochanteric fractures of the femur demand a special consideration in orthopedic traumatology, given the high rate of complications associated with their management ^[11]. When the intramedullary devices cannot be used for technical reasons, as in our case, the dynamic condylar screw provides a reasonable alternative, especially in the third world where costly image intensifier facilities are not available as a norm in hospitals due to financial constraints. Because of the familiarity of most surgeons with the sliding hip screw techniques in the treatment of trochanteric fractures, the adaptation is enhanced. The position of the condylar screw makes it possible to stabilize the proximal fragment and hence the fracture with extra screws ^[12].

Conclusion

Our suggestion to the readers is that surgery is an effective treatment modality in patients with osteopetrotic fractures, although intraoperative technical difficulties may be experienced and fracture healing is slower than normal and possible postoperative complications during the follow-up period should be taken care of. Special focus should be given to these patients with osteopetrosis due to their fragile bone status resulting from defects in osteoclast function and consequent impaired wound healing. The DCS provides a good alternative to intramedullary nails in the fixation of subtrochanteric fracture in the severely osteopetrotic bone.

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