

Case Report of Femoral Neck Stress Fracture in X-linked Hypophosphatemic Rachitis Patient Treated with Mechanical Axis Correction and a Literature Review

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Learning Point of the Article:

Treatment of adult hypophosphatemic patients with coxa vara and a femoral neck stress fractures can be surgical treatment with femoral cephalomedullary nail for combined deformity correction and fracture fixation.

Abstract

Introduction: X-linked hypophosphatemic rachitis (XLHR) is the most common cause of hereditary rickets that can lead to long bone deformities requiring multiple surgical correction procedures. In addition, high rates of fractures are reported in adult XLHR patients. This study aimed to report a case of femoral neck stress fracture in XLHR patient treated with mechanical axis correction. No previous studies demonstrating a combined valgus correction and cephalomedullary nail fixation were identified in the literature.

Case Report: A 47-year-old male patient with XLHR attended the outpatient clinic with severe left hip pain. X-rays revealed a left proximal femoral varus deformity and a femoral neck stress fracture. After 1 month without improvement of pain, and no radiographic sign of healing, correction of the proximal femoral varus deformity and fixation of the cervical neck fracture was achieved by a cephalomedullary nail. At 8 months follow-up, hip pain relief was achieved with radiographic healing of the femoral neck stress fracture and the proximal femoral osteotomy.

Conclusion: A review of the literature was performed to identify any case report of femoral neck fractures fixation due to coxa vara in an adult. Both coxa vara and XLHR can cause femoral neck stress fracture. This study presented the surgical technique for treating a rare case of femoral neck stress fracture in a XLHR patient with coxa vara. Pain relief and bone healing were achieved by combined deformity correction and fracture fixation with a femoral cephalomedullary nail. The technique for deformity correction and cephalomedullary nail insertion in the patient with coxa vara is shown.

Keywords: X-linked hypophosphatemic rachitis, stress fracture, femoral neck fracture, coxa vara, adult, cephalomedullary nail.

Introduction

X-linked hypophosphatemic rachitis (XLHR), with an incidence of 3.9/100,000 live births, is the most common cause of hereditary rickets [1]. The etiology of XLHR is mutations in the PHEX gene which generates high levels of fibroblast growth factor 23 (FGF23) causing a defective bone mineralization [2]. XLHR usually occurs in early years of life with bowing legs and short stature that lead to pain and difficulties in walking [3].

Hypophosphatemic ricket conventional treatment includes active vitamin D analogs and oral inorganic phosphate [4]. However, a new antibody drug against FGF23, burosumab, has shown through clinical trials that it is helpful for pediatric patients with XLHR, preventing lower limb deformity [5]. Considering the surgical treatment of adult patients with XLHR, few studies are available in the literature, highlighting the use of intramedullary fixation after realignment osteotomies which are effective for long healing times, and additionally, recurrence of

Author's Photo Gallery



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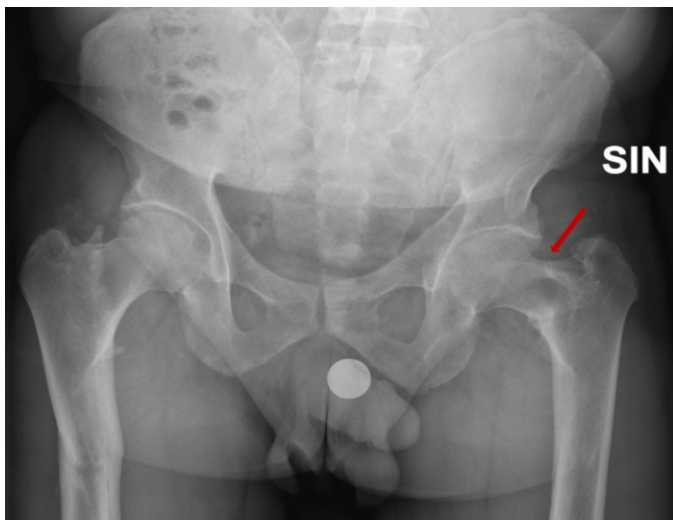


Figure 1: First X-ray of the patient. The femoral neck stress fracture is highlighted with red arrow and proximal varus deformity of the left femur.



Figure 2: Second X-ray after 1 month of conservative treatment. The continuous stress fracture of the left femoral neck is highlighted with red arrow.

deformity can be prevented in the long-term [6].

We present a rare case of XLHR with severe pain due to femoral neck stress fracture treated by mechanical axis correction and femoral cephalomedullary nail. SCARE guidelines were followed to report this case [7].

Case Report

A 47-year-old male patient with known history of XLHR attended the outpatient clinic after severe left hip pain, which escalated gradually during the past 8 years. This pain was load-related, as well as initiation pain, and there was no related

trauma. The clinical examination of the patient showed a positive Trendelenburg sign, moderately tight internal and external rotation, but good abduction and adduction. Previous surgery on the left femur included one midshaft corrective osteotomy in his adolescence, and the patient had been complaining about mild bilateral hip and spine pain for the past 15 years. The patient had no known allergies nor smoking history, height 178 cm, and BMI 27.1 kg/m². The patient has been under continuous check up from an endocrinologist for XLHR.

The X-ray revealed a left femoral neck stress fracture in

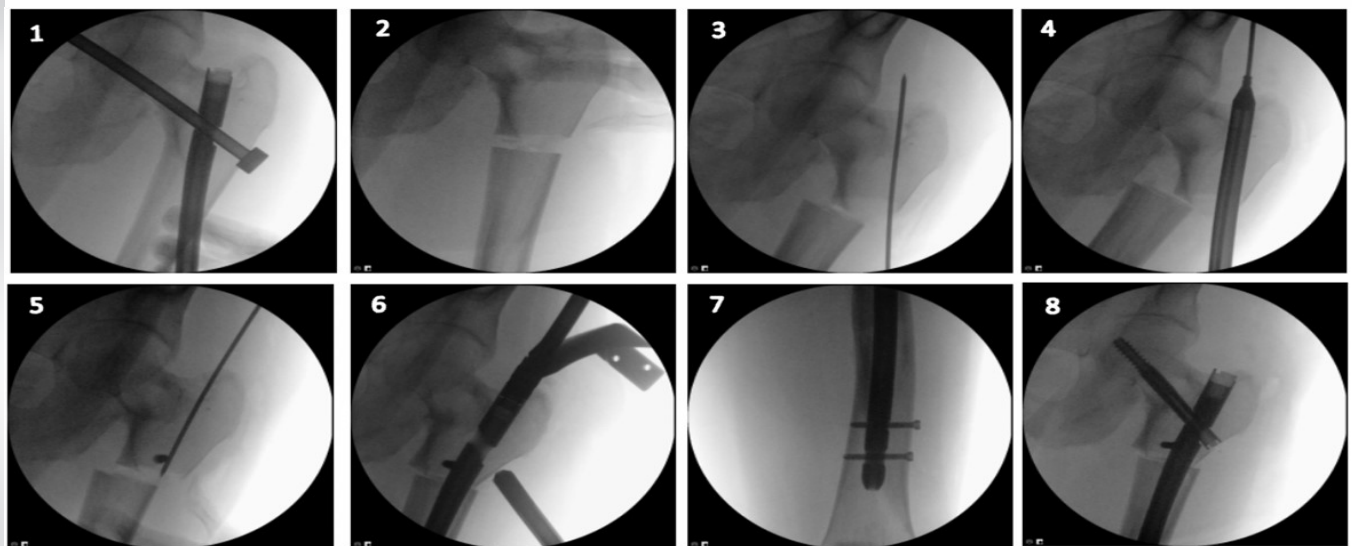


Figure 3: Operation Technique. Cephalomedullary nail left femur for varus correction and fixation of femoral neck stress fracture. (1) Checking the position that the cephalomedullary nail would be applied. An antigrade nail with a proximal bend designed for a trochanteric entry is inserted through the fossa piriformis to ensure proper valgus correction. (2) Osteotomy level allowing for good screw fixation in the proximal fragment. (3 and 4) Retrograde insertion of the guide pin and retrograde reaming with exit at the fossa piriformis. The retrograde preparation of nail canal in the proximal fragment allows for proper nail placement in the patient with a pre-operative coxa vara. (5) Position of the proximal blocking screw. (6) Antigrade insertion of the intramedullary nail with valgus correction of the proximal femur. (7) Distal locking screws. (8) Final correction and position of the cephalomedullary nail.

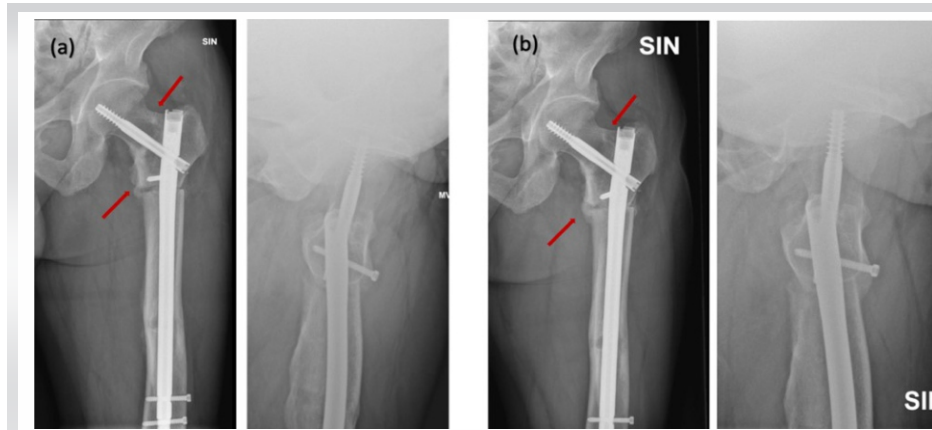


Figure 4: Follow-up in (a) 3 months and (b) 8 months. At 8 months, the hip pain has disappeared, the proximal femoral osteotomy and the femoral neck stress fracture has radiographically healed.

X-rays in 3 months (Fig. 4a) and in 8 months (Fig. 4b) show the bone healing.

Discussion

XLHR is an inheritable disorder that can lead to long bone deformities, requiring multiple surgical correction procedures. Surgical intervention is required for correction of the lower limb deformity in more than half of the patients with XLHR, with the complication rates of these surgeries being high [8].

A stable protection against recurrence of deformities is facilitated by the intramedullary nail, even if the patient develops metabolic decompensation postoperatively. One of the possible complications in the surgical treatment of XLHR adult patients is subsequent loss of correction or recurrence of the deformity. The patient presented in this case report had no complications and no loosening of the intramedullary nail.

In general, when evaluating patients with stress fractures, other metabolic causes should also be ruled out, including eating disorders, hyperparathyroidism, and osteoporosis. In the presented case, the patient had no metabolic disease other than XLHR.

The femoral neck stress fracture of the XLHR patient presented in this report was due to the proximal femoral varus deformity altering the mechanical load of the femoral neck. Coxa vara in hip is defined as the neck-shaft angle being below 120.

Carpintero et al. [9] compared in a retrospective study 22 femoral stress fractures of the femoral neck between patients with coxa vara (12 patients) and without coxa vara (ten patients). Patients sustaining a femoral neck fracture and having coxa vara were younger and had longer duration of symptoms than patients without a coxa vara deformity suggesting that a focal concentration of mechanical stress in the femoral neck may have been produced due to coxa vara. A review of the

conjunction with a varus deformity with the center of rotation of angulation (CORA) at the proximal femoral metaphysis (Fig. 1). The patient was followed for a month without restrictions in daily activities.

After 1 month, there was no improvement of the pain, and no sign of healing was found (Fig. 2). It was then decided to correct the proximal femoral varus deformity to ensure proper mechanical load of the femoral neck. The correction osteotomy was performed with a cephalomedullary nail allowing for screw fixation into the femoral neck and head. The surgical technique is presented in Fig. 3. The patient was discharged 1 week postoperatively with the guidance to bear weight. Phosphate-A and vitamin D treatment was administered during the 3-month post-operative period. At 8 months follow-up, hip pain was relieved, while the femoral neck stress fracture and the proximal femoral osteotomy had radiographically healed. The follow up

Table 1: Literature review of femoral neck fractures fixation due to coxa vara in adult patients.

Study	Country	Age	Sex	Medical history	Femoral neck stress fracture	Pain duration	Treatment	Follow-up	Complications
Naik et al. (2013) [10] (Two cases)	India	38 Years	Female	No metabolic disease	Bilateral	6 months pain in both hip joints and inability to bear weight on the right side for 3 months after a trivial fall	Subtrochanteric valgus osteotomy with DHS fixation on the right side and in situ fixation with three cancellous screws on the left side were done in the same sitting	1 Year	No complications
		48 Years	Female	No metabolic disease	Bilateral	Pain for 4 months	Bilateral subtrochanteric valgus osteotomy and fixation with DHS in two different sittings	6 Months	No complications
Sariyilmaz et al. (2015) [11]	Turkey	26 Years	Female	Osteomalacia	Bilateral	Pain for 7 months, difficulty in walking, no trauma	Pauwels valgus osteotomy with a 135-angled plate	2 Years	No complications
De Oliveira et al. (2016) [12]	Brazil	43 Years	Male	No metabolic disease	Bilateral	Pain for a year	Fixation with two 7-mm cannulated screws, as the femoral neck was too narrow for the placement of three screws or a sliding hip screw	N/A	N/A
Tabrizi and Mehdizadeh (2019) [13]	Iran	35 Years	Male	Obese, Osteomalacia	Bilateral	Lost the ability to walk 2 weeks	Valgus osteotomy with a DHS 135 and anti-rotational partial thread cancellous screw in two different sittings	6 Months	No complications
Rajkumar et al. (2020) [14]	India	30 Years	Female	No metabolic disease	Bilateral	2-month groin pain for which she consulted elsewhere diagnosed with bilateral stress fracture of femoral neck and underwent cancellous screw fixation bilaterally. Pain was not relieved for 6 months and was not able to walk independently, no trauma	THA left hip and no surgery right hip	1 Year	No complications
Tomar et al. (2020) [15]	India	58 Years	Female	Obese, Osteomalacia	Bilateral	Pain with difficulty in walking for the past 3 years, no trauma	Staged bilateral THA	2 Years	No complications

Table 1: Literature review of femoral neck fractures fixation due to coxa vara in adult patients.



literature was performed to identify any case report of femoral neck fractures fixation due to coxa vara in an adult, and the studies are presented in Table 1 [10, 11, 12, 13, 14, 15] highlighting the different treatment options that were used in accordance with patients' age and medical history. Three cases reported on femoral neck stress fractures in patients with coxa vara and osteomalacia. Tomar et al. treated the patient with total hip arthroplasty [15], while two cases report successful healing by valgus correction using either a 135 angulated plate [11] or a dynamic hip screw [13]. We could not find any previous studies demonstrating a combined valgus correction and cephalomedullary nail fixation.

Conclusion

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given the consent for his/ her images and other clinical information to be reported in the journal. The patient understands that his/ her names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

Clinical Message

The possible cause leading to femoral neck stress fracture should always be taken in consideration when planning patient's surgical treatment. Femoral cephalomedullary nail for combined deformity correction and fracture fixation is a treatment option in adult hypophosphatemic patients with coxa vara and a femoral neck stress fracture.

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Consent: The authors confirm that informed consent was obtained from the patient for publication of this case report

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