Percutaneous screw fixation of fractured neck of femur in a teenage girl with osteogenesis imperfecta. A case report

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A R T I C L E   I N F O

Article history:
Received 3 May 2018
Accepted 28 June 2018
Available online 9 July 2018

Keywords:
Osteogenesis imperfecta
Fractured femur neck
Fragility fracture
Surgical fixation
Percutaneous screw fixation
Case report

A B S T R A C T

INTRODUCTION: Neck of femur fracture in osteogenesis imperfecta is rarely reported. The management is uncertain and always reported as difficult. Clinical outcomes are not well described.

CASE PRESENTATION: A teenage girl, who is known to have osteogenesis imperfecta, sustained neck of femur fracture in the presence of a previously inserted implant. She was treated by percutaneous screw fixation. In both immediate and long term follow up, our patient had satisfactory outcome and return to pre-fracture functional status.

DISCUSSION: Dealing with fragility fracture in small abnormal hips is challenging especially, in the presence of a retained implant in situ. Intra-operative difficulty were encountered during positioning, reduction, and screw insertion attempt.

CONCLUSION: This rare case illustrates the anticipated difficulties in managing neck of femur fracture in osteogenesis imperfect patients. It also emphasize on the importance of surgical fixation to control pain and allow for healing in anatomically functional position.

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1. Introduction

Osteogenesis imperfecta (OI) is a genetic disorder of connective tissue that mainly affect bone [1]. It is associated with four genes mutations, primarily affecting the dominantly inherited type I collagen genes COL1A1 and COL1A2 producing abnormal bone matrix that does not respond well to mechanical loads [2]. Recessive transmission involving other genes has been reported [1]. OI occurs worldwide with variable incidence and no gender preference affecting approximately 25,000–50,000 individuals in the United States [1].

Four distinct types were initially described by Silence [3] according to clinical and genetic characteristics, varying from the most severe perinatal lethal form to the mildest, nearly asymptomatic form. Three additional types (Type V–VII) with different genetic abnormalities, and specific associated clinical findings were later described, representing only 5% of all patients [4,5]. Currently, there is no specific treatment to correct the genetic defect. However, advancements have been made in medical symptomatic treatment [1,2,6].

Repeated long bone fractures is the main feature of OI. Lower extremity are more affected than upper extremity, especially the femur [1,7–9]. Fractures occur after trivial injury, with no particular fracture pattern diagnostic to the disease. They are thought to be the result of combination of disuse osteopenia, progressive long bone deformity, and joint stiffness from immobilization [10]. They are usually non displaced [8] and heal at a normal rate [1]. Frequency of fractures is expected to decline significantly after adolescence [1]. The resultant deformsities following asymmetric growth disturbance due to multiple physal fractures lead to short stature [10]. Severe bowing deformities of lower extremity is typically corrected by surgical realignment and intramedullary rodding [11].

Neck of femur (NOF) fractures in OI is rarely described in literature [7,8]. It is usually undisplaced, associated with other fractures, and therefore, easily missed on standard imaging [7]. It is difficult to treat NOF fracture in OI especially when there is an intramedullary (IM) rod in situ in the femur shaft [7–9]. There are only few who reported the difficulty of surgical fixation [7–9]. Most patients had previous implant in the femur at the time of NOF fracture. All cases except for one were treated by surgical fixation, in which the implant was retained, and fracture was fixed with either dynamic hip screw, or percutaneous screw fixation. The clinical outcomes were not consistently reported in all cases.

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https://doi.org/10.1016/j.ijscr.2018.06.039
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We present our experience in managing NOF fracture in a teenage girl with osteogenesis imperfecta in the presence of a previously inserted implant. In both, immediate and long term follow up, our patient had satisfactory outcome and return to pre-fracture functional status. This case report was written according to the recently published SCARE criteria [12].

2. Case presentation

A 19 year-old girl wheelchair dependent, known to have scoliosis, and osteogenesis imperfecta (Sillence Type III) [3] presented to our emergency department complaining of right hip pain after a twisting mechanism to her right lower extremity while she was lifted from car to wheelchair by a caregiver. Her pain was localized to hip and groin area of the right side. Patient was previously managed for multiple long bone fractures with a telescoping growing rod of her right femur. She was on medical treatment; she started her first dose of intravenous bisphosphonate two months prior this presentation.

On examination, patient was lying in bed with the right lower extremity externally rotated. Right hip was tender to palpate with limited painful range of motion. X-ray revealed right neck of femur non displaced trans-cervical fracture with an Intra-medullary rod (Fassier-Duval IM rod) in situ (Fig. 1A). Operative fixation was decided for the patient and computed tomography was obtained to evaluate for bone stock, screw insertion site, and direction (Fig. 1B).

Patient was taken to the operating theater in less than 24 h. Under general anesthesia, she was placed supine on fracture table with both lower extremity suspended in scissoring position, but not under traction. Under C-arm guidance, fracture was stabilized with percutaneous pins. Entry point and path of the guide pin was continuously checked in two views; anteroposterior and lateral. Obtaining the lateral view was difficult because while the patient was in scissoring position, the beam angle was directed very cephaled trying to obtain lateral projection and image was

Fig. 1. Patient presentation: A. X-ray image shows right neck of femur non displaced trans-cervical fracture with the presence of an Intra-medullary (Fassier-Duval telescoping rod) in the femur shaft. B. Computed tomography was obtained for pre-operative planning.
obstructed due to presence of IM rod. However, we managed to place two cannulated screws (size 6.5 mm) in either ends of the rod, one anterior and another posterior to the rod, with no possibility of inserting a third screw due to the narrow neck (Fig. 2). Post operatively, she was immobilized in a hip spica cast for six weeks. Her hospital stay was uneventful. The reduction was maintained during early post-operative period.

Five month follow up X-ray and computed tomography confirmed healing of the neck of femur fracture in anatomic position (Fig. 3). Two and a half year following the surgical fixation, patient had satisfactory result of complete union with no signs of head vascular compromise (Fig. 4). She returned to her pre-fracture functional status.

3. Discussion

Osteogenesis imperfect is a life long disease characterized by multiple pathological fractures of long bones resulting in skeletal deformities, functional impairment, and disability [1–5]. Little is known about NOF fracture due to rarity of reported cases [7,8]. NOF fracture typically occurs just proximal to a previously inserted implant. This is explained by increased stiffness of the femoral shaft following implant insertion, producing high stress concentration at the NOF [13]. The fracture itself follows a trivial injury resulting in undisplaced fracture line. Blood supply is likely to be reserved and therefore, it is managed by internal fixation [7,8]. Surgical fixation of such a fracture is always difficult. Only
few, described in details the intra-operative challenges and clinical outcomes [7–9].

The largest series in literature reported by Chow et al. [7] consisted of five hips with NOF fracture in four patients, two children and two adults with one having bilateral NOF fracture. All cases except for one had previous implant in situ; typically an IM rod which was retained in all cases, or dynamic hip screw (DHS) which was removed prior to NOF fracture fixation. All cases were surgically treated by percutaneous screw fixation except for one which had an ipsilateral inter-trochanteric fracture and therefore, was fixed with pediatric dynamic hip screw. Three cases had an associated ipsilateral distal femur fracture that was managed by cross k-wiring. Within early post-operative period, one case was revised with open reduction and internal fixation with cancellous screws due to improper reduction and fixation. Another case, which was fixed with two screws anterior to the IM rod, lost the reduction at the third week, and resulted in coxa vara.

Another series by Tsang et al. [8] consisted of two pediatric patients. Both cases had previous IM rod which was retained, closely reduced and fixed with percutaneous screws. One case was fixed with two screws adjacent to the IM rod and a third screw with washer. The other case had too narrow neck for a third screw. Both cases resulted in union with no signs of avascular necrosis. The use of post fixation spica cast were consistently mentioned in all cases in both series.

Intraoperative difficulties were encountered in all reported cases during both positioning of the patient and fixation of the fracture. It is recommended to avoid the use of traction and instead rely on freehand technique [7]. The presence of IM rod in anatomically deformed short narrow neck makes it difficult to properly reduce the fracture. However, removal of the IM rod would risk displacing the NOF fracture. Moreover, it might predispose the patient to sustain future femur shaft fractures [9]. Assessment of orientation of the fracture line and direction of the screw with narrow margin of screw insertion attempt is another difficulty [7–9]. Chung et al.
ability. NOF fracture in OI patients is rare, associated with other fractures, and therefore, could be easily missed [7–9]. Surgical fixation of such a fracture is always difficult due to deformed anatomy, bone fragility, and presence of a previously inserted implant. Management outcomes are not well described due to lack of rich experience [8]. Control of pain, and patient return to pre-fracture functional status are the goals of surgical intervention. Goals are achieved by optimal fracture fixation, allowing healing in anatomically functional position.

Conflict of interest

No conflict of interest.

Funding

None.

Ethical approval

Ethical approval was obtained from the Institutional Review Board at the Royal Medical Services of the Bahrain Defence Force (file attached).

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Jawaher Alkhatteeb: literature review, collecting data, writing and submitting the manuscript.

Abdulla Aljawder: literature review, collecting data, reviewing and editing the manuscript.

Fawaz Alabbasi: collecting data, editing the manuscript.

Registration of research studies

Research registry 4036.

Guarantor

Jawaher Alkhatteeb.

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