Case Report **IJCR (2020) 4:187**

International Journal of Case Reports
(ISSN:2572-8776)

Bilateral Distal Humeral Fracture in a patient with Osteogenesis imperfecta During the Covid Pandemic– A Rare Case Report

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ABSTRACT

Introduction: Osteogenesis imperfecta (OI) is a genetic disorder which is characterised by bone deformity, low bone mass, connective tissue manifestations and brittle bones with history of multiple fractures. Although recurrent fractures are known in patient with OI, the occurrence of bilateral simultaneous fracture of humerus is quite rare.

Presenting complaint and investigations: The patient had a fall in the outdoors during a storm during the height of covid pandemic and sustained injury to both arms. Radiographs revealed comminuted fracture of left distal humerus and transverse extraarticular supracondylar fracture of right humerus.

Diagnoses, therapeutics interventions, and outcomes: Although, conservative measures were advised as per the guidelines for orthopaedic surgery during the covid pandemic for most of the fractures, the decision to operate was made considering the morbid obesity and hampered mobility due to bilateral above elbow plaster.The patient underwent staged open reduction and internal fixation of both distal humerus with anatomical locking plates in a span of 2 weeks. She recovered uneventfully with a good functional range of movements and resumed her routine activities.

Conclusion: The treatment for complex and unusual fractures should be individualised and should take into consideration of patient's general condition and safety.

Keywords: osteogenesis imperfecta, brittle bone disease, bilateral, distal humerus fracture

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How to cite this article:
Vaisakh Reghuram, Milind Mehta, Gautam Talawadekar. Bilateral Distal Humeral Fracture in a patient with Osteogenesis imperfecta During the Covid Pandemic– A Rare Case Report. International Journal of Case Reports, 2020; 4:187.

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Website: http://escipub.com/

Introduction

Osteogenesis Imperfecta (OI) or brittle bone disease is the most common heritable disorder of connective tissue [1]. The incidence of osteogenesis imperfecta is approximately 1 in 10000 to 20000 worldwide. It results from the mutations in the gene COL1A1 on chromosome 17 and COL1A2 on chromosome 7, which encodes the synthesis of two pro –alpha 1 chains and pro-alpha 2 chains of type 1 collagen [2,3].

The lack of type 1 collagen affects multiple organs including bones, joints, ears, eyes, skin, and other tissues. This disorder presents with a wide range of clinical spectrum ranging from mild form (multiple fractures and dental involvement) to severe survivable form (fractures at birth and short stature) and perinatal period mortalities [4].

The classification for osteogenesis imperfecta is based on the original publication by Sillence et al (1979) based on the epidemiological and genetic study in 180 OI patients. They were classified into four types based on the primary characteristics and mode of inheritance namely (1) Autosomal dominantly inherited OI with blue sclera (2) Lethally perinatal OI with crumpled femora and beaded ribs (3) progressively deforming OI (4) Autosomal dominantly inherited normal sclera [5].

Bilateral simultaneous humerus fracture is quite unusual and occurs as a result of electrocution, epileptic episodes or extreme trauma [6]. The quality of elbow function after fracture of distal humerus primarily depends on the degree to which normal anatomical relationships are restored. Although these are the basic principles as advocated by AO [7], they may be technically difficult to achieve in the presence of comminution and/or osteoporosis [8]. Hence open reduction and internal fixation using anatomical locking plates are recommended in the treatment of these type of fractures. The work has been reported as per the SCARE criteria [9].

We report a rare case of simultaneous bilateral distal humeral fracture in a patient with osteogenesis imperfecta who underwent open reduction and internal fixation of both humerus during the covid pandemic.

Case Presentation

75-year-old lady was brought to the Emergency department during the covid pandemic following a fall on outstretched hand as she got blown by the wind. She had a history of hypertension, had undergone bilateral total Knee replacements in the past and continued to be independently mobile with occasional use of single stick. She also had a history of multiple fractures in the past that were treated non-operatively.

General examination findings included blue sclera with no ligamentous laxity or dental manifestations. She was morbidly obese with a body mass index of 41. On examination, skin was intact with bilateral elbow swelling and ecchymosis. There was severe tenderness over the distal aspect of both humeri. There were no clinical features suggestive of compartment syndrome and the distal neurovascular examination was normal. Initial management included application of bilateral above elbow back slabs.

Investigation

Radiographs of left elbow showed a comminuted fracture of left distal humerus with posterolateral displacement, whereas radiographs of right elbow showed transverse supracondylar extraarticular fracture of right distal humerus with posterior displacement.

Treatment

The patient underwent open reduction and internal fixation (ORIF) of left distal humerus, under general anaesthesia, couple of days after injury. With patient in lateral decubitus position, with arm over a ‘L’ bar, a posterior midline incision was chosen and fracture was exposed through triceps splitting approach. Following radial nerve

exposure and gentle retraction, the fracture was reduced anatomically and fixed with three 3.5 mm cortical screws using the lag principle. A 8 holed distal humeral extra-articularlocking plate (Synthes) was used as a derotation plate and fixed using combination of cortical and locking 3.5mm screws. The patient was allowed to start gentle mobilisation on day 1 post operatively. This allowed the patient to start using her left arm for purposes of feeding and hygiene, which would not have been the case if the fracture was treated non-operatively.

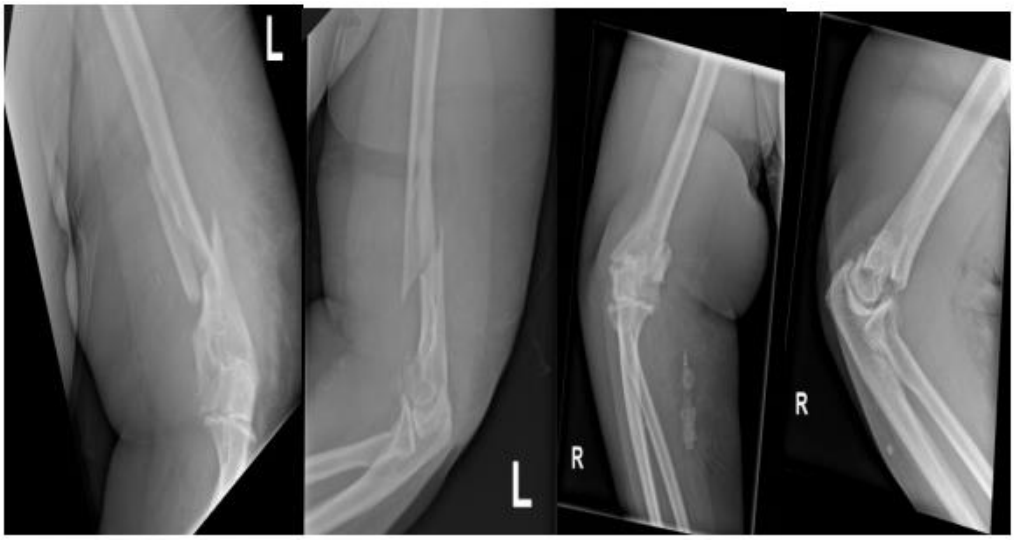

Figure 1: Initial x-rays –AP and lateral view of right and left elbow CT of the both elbows was done for pre-operative planning.

The patient underwent ORIF of right distal humerus 2 weeks following the surgery on left distal humerus, again under general anaesthesia with nerve block and the patient having been positioned in lateral decubitus position with arm over a 'L' bar. Skin incision used was again a posterior midline one however a Para tricipital approach was used for exposing the fracture site on this occasion, due to the distal nature of the fracture. Ulnar nerve was identified and protected throughout the procedure. Lateral and medial pillar were exposed and the fracture was temporarily fixed with K wires, after obtaining anatomical reduction. The columns were fixed using (Synthes) 7 holed medial pillar distal humerus locking plate and 10 holed posterolateral distal humerus plate using combination of 2.7mm and 3.5mm locking screws. After fracture fixation, it was ensured that ulnar nerve was not lying over the plate. Post-operatively, a back slab was applied until wound healing for 2 weeks.

Follow up

The patient was followed up at regular intervals for serial radiographs. She underwent routine physiotherapy as per protocol. This included gentle Range Of Motion (ROM) exercises for 4 weeks followed by active ROM progressing to strengthening exercises over a period of 8 to 10 weeks.

Right elbow radiographs at 2 months suggested radiological union. Left elbow radiographs at 6 weeks suggested that the distal screws along with plate had pulled off from the bone despite use of locking screws. There were no clinical signs of infection. Due to loss of fixation, the patient was put in a above elbow cast for 4 weeks following which the radiographs showed good callus formation at fracture site indicating radiological union.

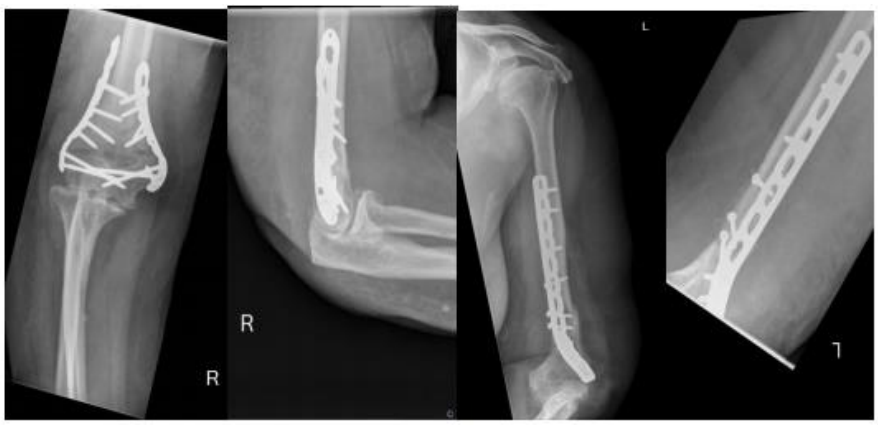

Figure 2: 1-month postop x-rays –AP and lateral view of right and left elbow

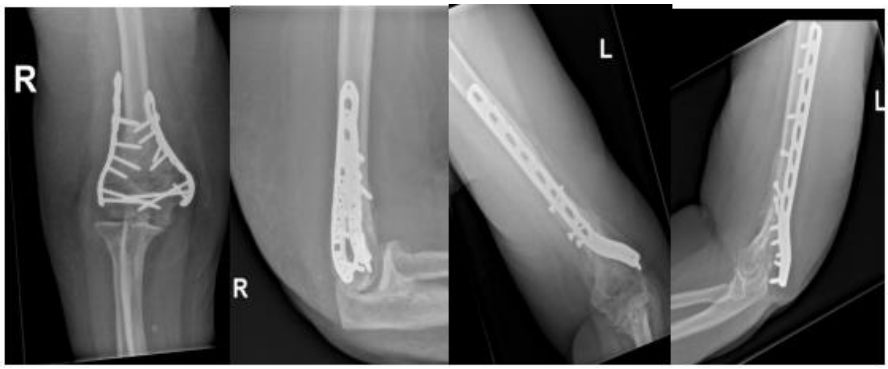

Figure 3: 2-months postop x-rays –AP and lateral view of right and left elbow

At 3 months, the fractures united clinic-radiologically with good range of movements, on both sides. On the left side, however, the plate could be palpable subcutaneously on the lateral aspect of distal arm causing localised pain. Implant removal was suggested but the patient decided against it as the discomfort was minimal.

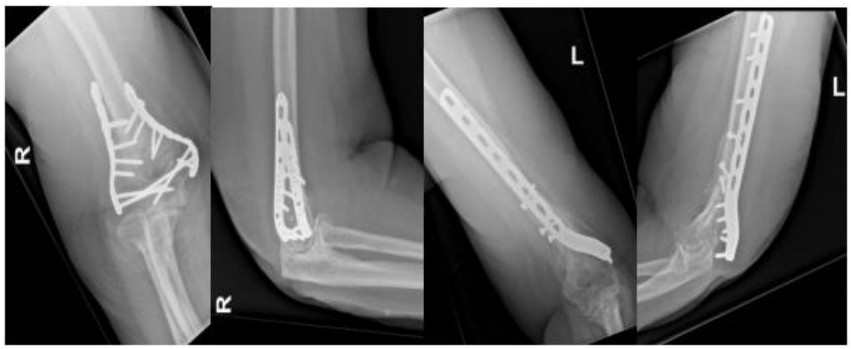

Figure 4: 6-months postop x-rays –AP and lateral view of right and left elbow

The patient is currently able to mobilise indoors without any aid and able to perform her daily household activities and personal care independently. The patient obtained good functional outcome with 80 degree of elbow flexion and 150 degree of elbow extension on the right. Although her elbow range of movements on the left side were slightly less as compared to right one, her functional outcome was similar to that of right arm.

Discussion

Osteogenesis imperfecta is the most common

type of inherited connective tissue disorder associated with bone fragility and extensive molecular and phenotypic heterogenicity [10]. It usually presents with reduced bone mass associated with recurrent fractures due to low energy trauma. The majority of OI patients have autosomal dominant mutations in type 1 collagen genes (COL1A1, COL1A2) resulting in decreased production or aberrant type 1 collagen formation resulting in fragile bone. Although genetic and molecular classification describes the multiplicity of new genes discovered in the field of osteogenesis imperfecta, phenotypic classification is still used commonly as rule [10,11].

Table 1: Current modified phenotypic Sillence classification of osteogenesis imperfecta [11]

OI type	Inheritance	Features
1	AD	**Non deforming type** Fragile bone Presenile hearing loss Blue sclera
2	AD, AR	**Perinatal lethal type** Extremely severe bone fragility
3	AD, AR	**Progressing severe deforming type** Moderate to severe osseous fragility
4	AD, AR	**Moderate type** Generally normal sclera
5	AD, AR	**calcification** of interosseous membrane and/or hypertrophic callus

Clinical features of severe form of OI include fractures and deformities of long bones identified by antenatal ultrasound during the first/second trimester of pregnancy. In moderate cases, the deformities start developing in the first 1-2 years of life when the patient starts weight bearing. In mildly affected cases, the patient suffersfracture only in the periods of rapid growth and osteoporosis [12].

Our literature review revealed several case reports for fractures in osteogenesis imperfecta [13,14] and few articles reporting bilateral proximal humerus fracture [15]. PubMed search could not find any articles describing the treatment for simultaneous bilateral distal humerus fractures. The decision to treat such complex fractures during the covid pandemic needed to be decided carefully as there is higher risk of mortality in elderly patients with fractures [16].

The treatment options considered were conservative treatment for both fractures versus to operate on comminuted fracture and treat the less displaced one conservatively in plaster versus operate on both the fractures. During the covid pandemic, it was recommended to treat most of the fractures conservatively in lieu to surgical treatment considering high risk for the patient and for the surgeon [16]. Although the right distal humeral fracture could have been treated conservatively in above elbow plaster cast, she was struggling with the mobility because of short stature and obesity. Treating the patient in plaster cast may have hampered her mobility causing significant morbidity and risks of deep vein thrombosis and cardiac dysfunction. Hence it was decided to operate on the both distal humeral fractures. Once it was decided to operate on both fractures, the choice was to operate both the humerus fracture in a single sitting or in staged manner. After considering risk factors like obesity, increased operating timing for each fracture and covid infection, it was decided to perform the surgery on the 2 sides in staged manner. The patient underwent ORIF of left distal

humerus first and ORIF of right distal humerus after a gap of 2 weeks. Her follow up radiograph showed evidence of healing. The functional outcome of arm and pain improved with this treatment and she went back to her routine activities.

Conclusion

The case report presents a rare case of bilateral distal humerus fracture in a patient with osteogenesis imperfecta which was treated surgically during the covid pandemic. We recommended to fix bilateral upper limb fractures at the earliest so that the patient regains independence and is able to continue with daily activities and personal hygiene. The general condition and safety of the patient should be considered while treating bilateral upper limb fractures.

Take home message

Simultaneous bilateral humerus fractures in a patient with osteogenesis imperfecta is quite rare.

Operative fixation of upper limb fractures becomes important when it hampers the mobility of the patient.

Early mobilisation of the joints provide good functional outcome and range of movements. Treatment should be catered as per the condition and safety of the patient.

Conflicts of interest

There is no conflict of interest in publishing this case report

Informed consent

Consent was obtained from the patient for making this report. No identity identifiers are present in the manuscript.

Abbreviations

OI – Osteogenesis imperfecta

ORIF – open reduction and internal fixation

AP - anteroposterior

CT – computed tomography

AD – autosomal dominant

AR - autosomal recessive

AO - Arbeitsgemeinschaft für Osteosynthesefragen

References

[1] Elmrini A, Boujraf S, Marzouki A et al. Osteogenesis imperfecta tarda. A case report. Nig J orthop Trauma 2006; 5:61-2

[2] Royce PM, Steinmann B, Byers P. Osteogenesis imperfecta. In: Royce PM, Steinmann B, eds. connective tissue and its heritable disorders. Newyork: Wiley-Liss,1993:317-50

[3] Byers PH, Wallis GA, Willing MC. Osteogenesis imperfecta: translation of mutation to phenotype. J med Genet 1991; 28:433-42

[4] De Paepe A, Nuytinck L, Raes M et al. Homozygosity by descent for a COL1A2 mutation in two sibs with severe osteogenesis imperfecta and mild clinical expression in the heterozygotes. Hum Genet 1997; 99:478-83

[5] FS Van Dijk and DO Sillence. Osteogenesis imperfecta: clinical diagnosis, nomenclature and severity assessment. Am J Med Genet A. 2014 Jun; 164(6): 1470–1481.

[6] Joseph Maalouly, Dany Aouad, Nabil Dib, Antonios Tawk, Georges El Rassi: Simultaneous ORIF for bilateral comminuted proximal humerus fractures: Case report in an elderly patient; https://doi.org/10.1016/j.ijscr.2019.10.061

[7] Thomas P Ruedi, Richard Buckley, Christopher G Moran: AO Principles of Fracture Management; Ann R Coll Surg Engl. 2009 Jul; 91(5): 448–449.

[8] Ring D, Jupiter J.B. Fractures of the distal humerus.Orthop Clin North Am. 2000; 31: 103-113

[9] Riaz A.Agha, Mimi R. Borrelli, Reem Farwana, Kiron Koshy, Alexander J. Fowler, Dennis P. Orgill: The SCARE 2018 statement: Updating consensus Surgical Case Report (SCARE) guidelines; https://doi.org/10.1016/j.ijsu.2018.10.028

[10] Aline Bregou Bourgeois, Bérengère Aubry-Rozier, Luisa Bonafé, Lee Laurent-Applegate, Dominique P Pioletti, Pierre-Yves Zambelli: Osteogenesis imperfecta: from diagnosis and

multidisciplinary treatment to future perspectives; Swiss Med Wkly eCollection 2016 Jun 20; 146: w14322.

[11] Bonafé L, et al. Nosology and Classification of Genetic Skeletal disorders: 2015 Revision. Am J Med Genet A. 2015;167A (12):2869-92 9999 A:1–24).

[12] Heike Hoyer-Kuhn, Christian Netzer & Oliver Semler: Wiener Medizinische Wochenschrift volume 165, pages278–284(2015)

[13] Bayram S, Mert L, Anarat FB, Chodza M, Ergin ÖN: A Newborn with Multiple Fractures in Osteogenesis Imperfecta: A Case Report.; J Orthop Case Rep. 2018 May-Jun;8(3):71-73

[14] Sułko J, Radło W: Operative management of long-bone of the upper limb in children with osteogenesis imperfecta].; Chir Narzadow Ruchu Ortop Pol. 2005;70(3):195-9.

[15] Claro R, Sousa R, Massada M, Ramos J, M Lourenço J: Bilateral posterior fracture-dislocation of the shoulder: Report of two cases. Int J Shoulder Surg. 2009 Apr; 3(2):41-5.

[16] Vijay Kumar Jain, Hitesh Lal, Mohit Kumar Patralekh, Raju Vaishya: Fracture management during COVID-19 pandemic: A systematic review; https: // doi .org /10 .1016 /j.jcot .2020 . 06 .035