Pelvic Ring and Peritrochanteric Fractures in a Patient with Osteogenesis Imperfecta Treated Surgically: Case Report with a 5-Year Follow-Up

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Patient: Male, 52-year-old
Final Diagnosis: Osteogenesis imperfecta • pelvic ring fracture • peritrochanteric fracture
Symptoms: Limited mobility • pain
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Congenital defects/diseases
Background: Osteogenesis imperfecta (OI) is a rare disorder associated with brittle bones, skeletal deformities, short stature, and conductive hearing loss. It is caused by mutations in genes encoding collagen type I production and is associated with multiple fractures occurring during a patient’s lifetime. Atypical fractures can occur without a history of previous injury or diagnosis.

Case Report: A 52-year old man sustained a fall from his own height. He experienced pelvic pain but could bear weight. He was referred to the hospital by his primary care physician and was admitted to the Orthopedic Department with a pelvic ring fracture. We performed open reduction and internal fixation with a reconstruction plate via an intrapelvic approach. At 12 weeks after discharge, he sustained a peritrochanteric fracture. It was surgically treated with a dynamic hip screw. The patient commenced weight-bearing 20 weeks after the initial procedure. Bone union was achieved clinically and radiographically. He did not have any revision procedures. At the time of this report, 5 years after the described period, he felt no discomfort around the hips, sacroiliac joints, and pubic symphysis.

Conclusions: OI poses a difficult treatment challenge, but, achieving a good clinical and functional result is possible with a precise surgical technique and meticulous preoperative planning. Clinicians should always consider the possibility of a simultaneous fracture occurrence in different parts of the body. Some patients present without a previous diagnosis, and we should always have some suspicion of OI in cases of young patients presenting with low-energy fractures.

Keywords: Hip Fractures • Osteogenesis Imperfecta • Pelvic Bones

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Background

Osteogenesis Imperfecta (OI) is mostly known as a brittle bone disease [1,2]. In essence, it is a disorder affecting the production of type I collagen and therefore affects all of the tissues in which it is present [3-6]. Thus, in addition to a lower mechanical resistance of bones, its characteristic features are deformities and short stature (due to involvement of epiphyseal plates), conductive hearing loss, pulmonary ventilation abnormalities, and hemorrhagic diathesis [1,2,4,6-8].

Type I collagen consists of 2 alpha 1 chains and 1 alpha 2 chain wrapped around each other in an alpha-helix [5,6]. The structure of the chains is encoded in COL1A1 and COL1A2 genes [5,6]. OI mutations affect COL1 genes or genes responsible for post-translational modifications of collagen chains [6]. Depending on the type of mutation, there is either a qualitative or quantitative disruption of type I collagen production [6]. To date, 20 OI variants have been described in the literature [4]. Most OI types are inherited in an autosomal recessive manner, but there are also variants that are inherited in an autosomal dominant manner or linked to chromosome X [4].

In the 1970s, Silent et al described the first 4 OI variants, dividing them according to their clinical presentations [3,9]. The mildest is type I OI. With this type, the fracture risk is higher than in a general population, but it decreases with age and typically does not lead to complete disability; the characteristic traits of these patients are blue eyes and conductive hearing loss. Short stature and bone deformities such as scoliosis are uncommon [4,5]. Type II is prenatally lethal [4-6,9]. In patients surviving infancy, the most crippling is type III. It is associated with short stature, bone deformities, respiratory system disorders, and a high degree of motor disability [4,5]. Type IV is associated with a higher fracture risk than type I and more frequently leads to severe deformations [3,9].

Case Report

A 52-year old man sustained injury during orthostatic syncope, when he got up from the chair. Immediately after the trauma, he could bear weight but had pelvic pain. He reported to his primary care physician. A pelvic fracture was suspected after an examination that revealed bruising around the umbilicus and a painful pelvis on palpation. He was referred to the Emergency Department of the nearest hospital (which happened to be ours) for further evaluation. Diagnosis was established, and he was admitted to the Orthopedic Department due to a fracture of both pubic bones. A physical examination revealed typical features of OI, namely blue eyes and conductive hearing loss. Also, obturator nerve injury was recognized as a sequela of the fracture. A pelvic ring fracture was diagnosed based on radiographs (Figure 1). It was classified as type 61B according to AO classification [10]. Three-dimensional reconstruction in computed tomography (CT) showed a high degree of displacement and raised concerns of a bladder wall injury. After we obtained informed consent from the patient, he was prepared for surgery. Two units of packed red cells were cross-matched, enoxaparin was administered as an antithrombotic prophylaxis, and extended antibiotic prophylaxis was prescribed.

The surgery was performed on the tenth day after admission. The time elapsed from admission was used to prepare the patient and operating staff for the procedure. This report’s first author is a pelvic trauma specialist who is not a regular employee at our hospital. The procedure needed to be scheduled ahead of time. Based on a pelvic fracture protocol established at our institution, the patient was evaluated for possible sites of infection prior to surgery: daily changes in white blood cell count and C-reactive protein levels were observed. Also, the patient was closely monitored for any signs of respiratory or genitourinary infection. Before the procedure, deep vein thrombosis needed to be ruled out with ultrasonography. Based on radiographic studies in which 3D CT reconstruction is mandatory, our first author selected which systems, instruments, and implants should be ordered, because they are not available at all times at the hospital. The patient was placed supine on the operating table, and after standard surgical site preparation and draping, an intrapelvic approach was performed. The skin incision was made 3 cm distally to the umbilicus. Deeper, subcutaneous layers were incised in the plane of the skin incision. The rectus abdominis muscle was dissected bluntly in the linea alba. The pubic bone fractures were reduced by using bone forceps and were fixated with a 12-hole modeled pelvic reconstruction 3.5-mm plate and cortical screws (Figure 2). The plate and screws were placed, limiting manipulation and over-drilling. The quality of bone was extremely poor, and the main goal of the fixation was proper cancellous bone engagement. On the right side of the pelvis, it was possible to place...
screws to cross over the acetabulum. On the left side, we decided that 3 screws provided satisfactory stabilization and that trying to change the plate position or the plate itself would pose more risk than provide benefit. Based on the fracture location and its posterior displacement, the earlier suspicion of bladder injury was sustained and exploration was made by a general surgeon. No injury to the bladder structures was identified. A Spongostan sponge and surgical drain were placed in the operative field. Due to the inability to stitch fascia with sutures only, the reconstruction of the abdominal walls was made with a surgical mesh (Ethicon 6×11). The blood loss was estimated at below 500 mL.

The patient was transferred to the orthopedic ward after the procedure. Because of a decreasing hemoglobin level, 2 units of packed red blood cells were transfused. The patient was rehabilitated in bed. Weight-bearing was prohibited until the bone fusion was achieved. Follow-up visits were made in the outpatient clinic at our hospital. For 8 weeks, the patient was rehabilitated in bed. He could flex his hip joints up to 50 degrees, abduction and external rotation was limited, and he was encouraged to change position between supine and on his sides, with the prone position prohibited. After that period, he was allowed partial weight-bearing, using crutches for another 4 weeks.

Twelve weeks after the initial procedure, the patient sustained a low-energy peritrochanteric fracture of the left femur during another orthostatic hypotension episode. Closed reduction and fixation with a dynamic hip screw was performed. Four days after the procedure, he was discharged from the hospital. The postoperative protocol was similar to the previous one, and weight-bearing with a walking aid was commenced 8 weeks after the femur fracture.

At the time of this report, 5 years from the described period, he has not had any revision procedures; bone union was achieved after 4 months, and the skin and deeper structures healed without complications (Figure 3). He did not have any pain around the hips, sacroiliac joints, or pubic symphysis. His level of activity was decreased, but, as he stated, it was not because of the pelvic or femur injury but rather due to severe knee osteoarthritis. We could not justify performing a postoperative X-ray after 5 years, since the patient had not reported any symptoms from around the pelvis. An X-ray would have posed unnecessary radiation exposure and would have been associated with a marked discomfort for the patient, given his level of mobility. No neurologic or cardiologic diagnosis was made regarding the syncope episodes. After receiving education about orthostatic hypotension prevention, the patient had not experienced any similar episodes.

**Discussion**

Treatment of fractures in OI is very difficult due to the poor bone stock [1]. It is pivotal to handle the fracture fragments gently to prevent further comminution.

In OI there is a higher risk of malunion and nonunion, the callus quality is worse compared with that of otherwise healthy individuals, and there is a higher risk of fixation destabilization due to poor bony support [4].

Soft tissues can be hypoplastic and have structural defects, and their mechanical strength is significantly impaired [1,2,8]. This is associated with difficult wound closure. We encountered such difficulty during this case. A reconstructive mesh had to be used to close the abdominal walls.
Also, OI poses a significant challenge for the anesthetic team, owing to hemorrhagic diastasis, which significantly increases the risk of severe blood loss [1,2,8]. In the present case, the blood loss was relatively small, proving that with a careful and meticulous surgical technique, it is possible to reduce the blood loss to a volume that does not threaten hemodynamic stability.

To date, 5 cases of operative treatment of pelvic fractures in patients with OI have been described, all in acetabular fractures [1-3,8]. To the best of our knowledge, there is no described case of pelvic ring fracture treated surgically in a patient with OI. Darmis et al described treatment of T-type acetabular fractures in 2 patients. In both cases, the fixation was performed with a 3.5-mm reconstruction plate. In the first case, the blood loss was estimated at 1500 mL, and in the second, at 1800 mL. Both patients were made touch-down weight-bearing for 6 weeks, with progression to partial weight-bearing thereafter [8]. Liporace et al performed a total hip arthroplasty combined with fracture fixation in a female patient with type I OI. She sustained a fracture of both acetabular columns. Fixation was made with 2 modeled 3.5-mm reconstruction plates: a 12-hole plate was used for the column fixation and a 6-hole plate was used for the quadrilateral surface fixation. Intraoperative blood loss was estimated at around 1200 mL. The patient was made touch-down weight-bearing for 8 weeks, with gradual progression to full weight-bearing [2]. Medici et al published a case of bilateral acetabular fracture treated in 2 stages. The first stage involved posterior column fixation. The second stage was performed 10 days from the initial procedure and involved fixation of the anterior columns and quadrilateral surface. The blood loss volume was below 400 mL, and there were no indications for blood transfusion after the procedure. The patient was mobilized with crutches, with touch-down weight-bearing for 6 weeks and partial weight-bearing for another 6 weeks [1]. Pesch et al described treatment of an acetabular fracture (type 62 B3 according to AO) and femoral neck fracture in a patient with type IV OI. The femoral neck fracture was treated with a dynamic hip screw, and the pelvic fracture was treated with 2 3.5-mm reconstruction plates (4-hole and 12-hole). Stabilization of all the fractures was supported photodynamically [3].

It is difficult to compare our treatment to these previous cases, since only intraarticular fractures were described, which require a different strategy. Given the fracture location and its displacement, there was no other option than operative treatment for our patient. Conservative treatment would have inevitably failed and resulted in the patient having a disability. Also, using a single plate with an intrapelvic approach limited the injury to hypoplastic soft tissues and subsequent detrimental effects on wound closure and wound healing. In the present case, the intraoperative blood loss was one of the lowest described in the literature. The weight-bearing protocol of our patient differed from that of other cases, but our decision was based on an unexplained syncope mechanism, extremely poor bone stock, and not fully anatomical reduction. We did not want to risk destabilization of the fixation.

Conclusions

Every patient with suspected or confirmed OI should be thoroughly evaluated after every trauma, even minor trauma, and when having pain or loss of function. Physical examination and radiographic studies are mandatory because multiple fractures can occur simultaneously. Furthermore, clinicians should remember OI when a young patient sustaining low-energy fracture is presented. The suspicion should be raised when the patient has other distinctive features, such as bone deformities, conductive hearing loss, and blue eye color.

Pelvic fractures in patients with OI pose a significant challenge for the entire medical staff. Our case shows that it is possible to achieve good clinical and radiographic results with surgical treatment. During surgery, soft tissue injury and bone manipulation should be minimized as much as possible. The postoperative protocol should be designed on a case by case basis.

Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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