Fracture neck of femur in Factor XIII deficiency: Was better outcome possible?

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Abstract

The fracture in a Factor XIII deficiency patient is being reported for the first time in the literature. We report a displaced fracture neck of femur in a 17-year-old boy with Factor XIII deficiency. Open reduction and internal fixation was done 8 days after the following the initial injury. Two units each of fresh frozen plasma and cryoprecipitate were given perioperatively to prevent excessive bleeding. No perioperative bleeding complications were encountered. At 18-months follow-up, the fracture had united with evidence of avascular necrosis. The fracture neck of femur in a child or young adult needs to be reduced and stabilized at the earliest to prevent devastating complications. Its occurrence in a patient with Factor XIII deficiency is to be managed like in a normal patient, but with extra perioperative care. Undue delay in fixation as happened in this case should be avoided for a better outcome.

Keywords: Bleeding diathesis, clotting factors, cryoprecipitate, femur, fracture, hemophilia

Introduction

Hereditary Factor XIII deficiency is a rare bleeding disorder (1 in 5 million). Factor XIII deficiency is usually attributed to mutation in the Factor XIII A gene located in chromosome 6. Factor XIII deficiency is associated with increased mucosal bleeding, umbilical cord bleeding, menorrhagia, prolonged posttraumatic bleeding, repeated miscarriage, and life-threatening intracranial hemorrhages. To our knowledge, there is no literature on the management of fracture in patients with Factor XIII deficiency. We describe displaced fracture neck of femur in a boy with this rare bleeding disorder.

Case Report

A 17-year-old boy presented with pain in the right hip and inability to bear weight on the right lower limb following a trivial fall 6 days back. He was diagnosed to have Factor XIII deficiency at the age of two by clot lysis test. Earlier, he had multiple intramuscular bleeds (iliopectos twice, left quadriceps once) and intracranial bleed once. His right lower limb was in a flexed, abducted, and externally rotated attitude, and there was shortening. There was no contusion on the lower limb, and all movements of the right hip were painfully restricted. There were no signs of bleeding from any other site. Radiographs showed a displaced Delbet's Type 2 fracture neck of the right femur [Figure 1a and b].

Meanwhile, his bleeding and coagulation profiles were checked. Two days after admission, after obtaining clearance from his treating pediatricians, he underwent open reduction and multiple cannulated cancellous screw fixation of the fracture [Figure 2a and b], with evacuation of the intracapsular hematoma. On the preoperative night, he was given two units of fresh frozen plasma (FFP).

In the postoperative period, he received two units of cryoprecipitate infusion. Intra- and post-operative periods were...
uneventful, without any bleeding or wound complications. He started nonweight bearing crutch walking.

Radiographs revealed maintenance of the reduction and progression in the union of the fracture after 6 weeks [Figure 3a and b]. He was advised to continue nonweight bearing. However, he was noncompliant in follow-up. Incidentally, he presented with hemarthrosis of the right knee after 1 year. Radiographs of the hip showed features of avascular necrosis and loss of sphericity of the head of the right femur with backing out of implants [Figure 4a and b].

He underwent removal of the implants without any bleeding complications [Figure 5a and b]. He received four units of cryoprecipitate preoperatively and two units postoperatively during the second surgery.

At final follow-up, he was walking full weightbearing. He had 10° of fixed external rotation deformity and a painless short limb gait.

**Discussion**

Even though musculoskeletal complications such as hemarthrosis, intramuscular bleeding, pseudotumor, fractures, compartment syndrome, joint contracture, and secondary degenerative arthritis are well described in hemophilia,[3] long bone fractures have never been reported in Factor XIII deficiency. Internal stabilization for long bone fractures and external fixation for associated severe soft tissue injury are recommended in hemophilic patients.[4]

The principles to prevent and control bleeding during surgery are same in any bleeding disorder. Factor replacement is the key to the management of these patients.[4] With early fixation and mobilization, the rate of fracture healing and consolidation is same as the general population.[4]

Fractures of the hip in children are rare injuries (less that 1% of all pediatric fractures).[3] The principle of the management of fracture neck femur in young children is an urgent reduction, decompression of hematoma, and stable internal fixation. The frequency of complications is more than in other fractures. Osteonecrosis, coxa vara, nonunion, loss of fixation, premature physeal closure, and limb length discrepancy are common complications.[3] Delbet reported 16–78% prevalence of osteonecrosis after Type II pediatric neck femur fractures, and the incidence is more in displaced fractures. To reduce the risk of complications, type II fractures are usually treated with anatomic reduction and stable fixation with multiple cannulated cancellous screws.

Fracture neck of femur in children or adolescents with bleeding disorder is rarely reported in literature. High-energy trauma is needed to fracture neck of femur in children, but in children with bleeding disorders, as age progresses, because of repeated musculoskeletal problems and avoidance of contact sports, bones might be osteoporotic, and trivial trauma can lead to fracture.

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**Figure 1:** Anteroposterior (a) and lateral (b) radiographs of the hip joint showing Delbet’s Type 2 displaced fracture of the neck of the right femur

**Figure 2:** Immediate postoperative anteroposterior (a) and lateral (b) radiographs

**Figure 3:** Anteroposterior (a) and frog-lateral (b) radiographs at 6 weeks after surgery showing progressive union at the fracture site

**Figure 4:** Anteroposterior (a) and lateral (b) radiographs at 1-year follow-up showing avascular necrosis of the head of the right femur with coxa irregularis and backing out of screws
Osteonecrosis is a common complication in skeletally immature patients after fracture neck of femur due to vulnerable blood supply. Initial high-energy injury, degree of displacement, timing of operation, intracapsular hematoma evacuation, accurate reduction, and compliance with nonweight bearing are the determinants of osteonecrosis. Beaty reported Pforringer and Rosemeyer reported a better outcome in children and adolescents who underwent early operative treatment (within 36 h) than in those whose treatment was delayed. Many authors have reported the usefulness of decompression of the joint to reduce the risk of osteonecrosis. There were several reasons for osteonecrosis in our patient such as late referral and presentation, late clearance for surgery, poor compliance of patient, and probable intra-articular hematoma due to bleeding disorder despite getting a good reduction and stable fixation. We did not encounter any external or internal bleeding problems before or after both surgeries.

Lee et al. described the management of fracture neck of femur in hemophilia patients with trivial trauma. Fracture neck of femur is almost two decades earlier in patients with hemophilia compared to the general population. With a moderate dose of factor infusion, the end result of the treatment of fracture neck of femur in these patients is comparable to the general population in terms of average time of union and complication rates. Hemostasis in Factor XIII deficiency may be achieved with levels as low as 2–3%. Since the half-life of Factor XIII is 11–14 days, bleeding can be effectively controlled by giving FFP, cryoprecipitate, or plasma-derived Factor XIII concentrate, and these patients can be taken up for emergency surgeries if indicated. One to five ml per kg of cryoprecipitate is sufficient to maintain the hemostatic Factor XIII levels for 2–6 weeks. Since Factor XIII also has a role in angiogenesis, adequate supplementation can reduce wound healing complications. Ten to fifteen ml per kg of FFP is usually adequate for hemostasis, but as it has a short half-life, 6 to 8 hourly dosing may be required in the perioperative period.

Thus, we feel that in patients with Factor XIII deficiency, when urgent surgery is required, there is no need to delay the procedure due to fear of perioperative bleeding if cryoprecipitate or FFP is available. A fracture of necessity such as fracture neck of femur in Factor XIII deficiency patient must be managed early to avoid long-term disabling complications.

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Conflicts of interest
There are no conflicts of interest.

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