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Case Report

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Malunited supracondylar femur fracture in haemophilia A: a case report on evaluation and management

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ABSTRACT

Haemophilia is a rare inherited X-linked bleeding disorder associated with various levels of coagulation factor VIII (type A) or IX (type B) deficit. Hemophilic patients can be affected by trauma and fractures just like the general population. But due to their bleeding disorder, such patients need specific multidisciplinary management, from the emergency room to the operating theatre, in order to limit severe complications. The goal of modern fracture treatment is to obtain an optimal outcome, with the patient's return to full activity as soon as possible. If a fracture is correctly treated in a haemophilic patient, it will progress to consolidation in a similar time-frame to fractures occurring in the general population. Worldwide, there are only a few specialized orthopaedic centres dedicated to the management of hemophilia. Management of supracondylar femur fracture in patients with haemophilia is no different from general population if an adequate haemostasis is achieved. The purpose of this paper is to report our experience on the supracondylar fracture of the femur in hemophilia by a multidisciplinary team at a single institution.

Keywords: Haemophilia, Coagulation factor, Fracture, Multidisciplinary management

INTRODUCTION

Haemophilia is X-linked recessive coagulation disorder in which deficiencies of certain clotting factors are the cause of haemorrhage with spontaneous or trivial trauma. The three types of haemophilia are haemophilia A, B and C. The most common form, haemophilia A, occurs in 1 in 5000 male births. The deficient factor (f) is factor VIII. Haemophilia B affects only 1 in 50 000, where there is factor IX deficiency. Although bleeding can occur at almost any age, it usually comes to clinical attention by the age of one year with unexpected bruising over the extremities or haemarthrosis as the most common clinical manifestations.[1] The incidence of fracture in patients with haemophilia is rarely seen, as these individuals are less ambulant due to the gravity of their illness. Their daily activities are reduced because of associated arthropathy and contractures. On the other hand, poor musculature, osteoporosis and haemophilic changes in the bone may predispose them to risk of fractures. In patients with haemophilia the fracture can occur after a trivial trauma, especially if associated factors of haemophilic arthropathy, muscle wasting and osteoporosis render the bone more fragile and prone to fracture.[2]

CASE REPORT

A 24 year old male patient with alleged history of road traffic accident and was given primary care at a local primary health centre with above knee POP slab application, right side (Figure 1 A). After 3 months, he came to emergency department of our tertiary health care centre with chief complaints of pain and swelling in right knee with inability to bear body weight over the affected lower limb. Patient was admitted in emergency ward and later shifted to general ward for further management. He was a known case of hemophilia A and was under

treatment in hematology and medical genetics department of our centre.

On the day of admission, patient's vitals were within normal limits. For local examination, POP slab was cut and the overlying skin was found to be normal. A diffuse soft to firm swelling of approximately 20 x 8cm in size was seen in upper part of right knee. Vascularity was preserved as distal pulse was palpable.

Hematological and serological investigations were within normal limits (Table 1), except for mild thrombocytopenia (120×10^3/cumm). Coagulation profile was deranged. Serum factor VIII levels were reduced (2.7%). Viral markers were non-reactive.

In radiological investigation, X-ray of right hip with thigh with knee showed a malunited (procurvatum deformity) extra-articular fracture of right distal femur (Figure 1 B).

Surgery was planned for the deformity correction. In the pre-operative period, factor VIII was transfused as advised from hematology department and serum factor VIII and APTT monitoring was done. Extension osteotomy of distal femur was done and osteotomy was fixed with anatomical distal femur locking compressive plate under tourniquet.

Post operative period was uneventful. Patient was allowed protected weight bearing until fracture healed radiologically (Figure 1 C and D).

Follow up was done for 1 year 2 months apart and was unremarkable with no signs of fixation failure or infection. Range of active motion showed 70 degree flexion with full extension at knee joint with normal gait.

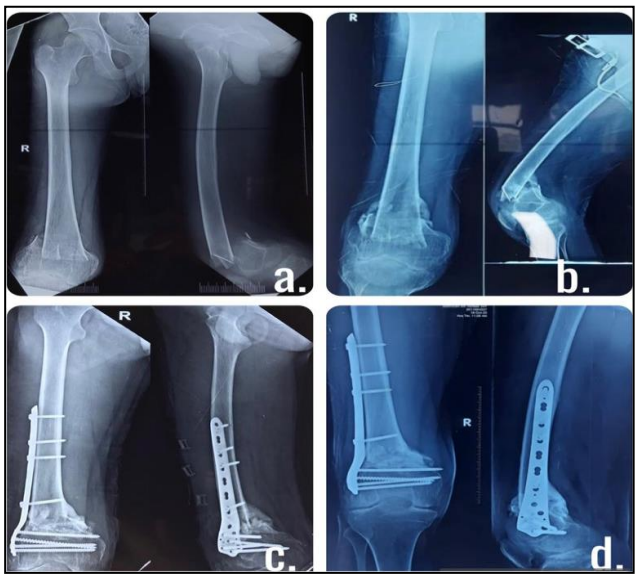

Figure 1 (A-D): X-ray right knee with thigh (AP and lateral view): On the day of trauma, malunion at fracture site, post operative day 1 and one year after surgery during follow-up.

Figure 2: Clinical picture during rehabilitation period.

Table 1: Routine investigations on day 1 of admission.

Parameters	Value	Units
Hematological		
Total leucocyte count	7.7	$\times10^3$ /cumm
Hemoglobin	14.2	gm/dl
Platelet count	120	$\times10^3$ /cumm
Biochemical		
Albumin	5.0	g/dl
Alkaline phosphatase	78	µ/L
SGPT	19	µ/L
SGOT	22	µ/L
Bilirubin-conjugated	0.34	mg/dl
Bilirubin-total	0.72	mg/dl
Creatinine	0.85	mg/dl
Sodium	142	mmol/L
Potassium	4.4	mmol/L
Total Calcium	8.6	mg/dl
Urea BUN	26.2/12.25	mg/dl
Coagulation profile		
APTT	64.1	Seconds
PT/INR	16.3/1.22	Seconds

DISCUSSION

Haemophilia is a rare inherited X-linked bleeding disorder associated with various levels of coagulative factor VIII (type A) or IX (type B) deficit.[3] A third rare form is haemophilia C, which is associated with a deficiency of clotting factor XI. Patients with hemophilia rarely engage in weight-bearing activities due to the fear of bleeding and chronic pain caused by haemophilic arthropathy, resulting in failing to achieve adequate bone mass and affecting the relative risk of fracture. The absence of FVIII or FIX and the failure to activate FX results in a deficient production of thrombin, the latter being able to cleave osteopontin,

which is required for osteoclast anchoring to the mineralized matrix. Furthermore, thrombin inhibits osteoblast apoptosis and osteoclast differentiation and stimulates osteoblastic cell proliferation, thereby improving bone formation and reducing bone resorption. FVIII is a biological regulator of bone metabolism.[4]

The rate of fractures is significantly greater in hemophilia patients than in the general population due to reduced bone mineral density. People with severe factor deficiency have an increased risk of fractures.[5] Fractures may be challenging conditions to treat in hemophilic patients, particularly cases with severe displacement, exposure and periprosthetic fractures. Displaced fractures as well as long-bone or periprosthetic fractures can show a high tendency to bleed and be associated with significant rates of complications. In such cases, immediate or urgent management is required.[3]

The presence of target joints next to the fracture site alters the final clinical outcome. Indeed, even though the fracture has healed, several patients still need further joint surgeries to correct the potential new imbalance caused by even minimal malalignment or arising during the functional recovery period itself.[6]

Surgical interventions in hemophilic patients are associated with an increased risk of intra- and postoperative major bleeding, wound healing disorders, and postoperative infections, but this was not seen in our case due to proper preoperative management and aseptic surgical precautions.[7]

Caviglia et al reported the largest series of fractures in PWH published to date. They treated 151 fractures in 141 patients: 121 subjects by a conservative strategy and the rest by internal fixation. In the follow-up period, they found 40 cases of mal alignment in the non-operated group, and 3 cases in the operated group. Moreover, they recorded a shorter fracture consolidation time and better anatomical alignment in patients treated by internal fixation than in those receiving conservative treatment.[8]

CONCLUSION

Despite the improvements in the management of haemophilia and its related clinical issues, a severe or displaced fracture in hemophilia is still challenging. This specific category of orthopaedic patients should receive urgent surgical treatment in dedicated facilities in order to ensure good outcomes and low rates of complications. Given the frequent condition of osteoporosis in hemophilic patients, further exploration of their low levels of bone density is required in order to achieve better knowledge of their present low risk of fractures.

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