

Haemophilia-A-related haematoma: management in resource constraint settings

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DESCRIPTION

Haemophilia is a X-linked recessive benign haematological disorder characterised by bleeding manifestations and coagulation abnormalities. Haemophilia-A (HA) is more common form of the disease and is associated with deficiency of factor VIII (FVIII). Here we present an interesting case of a 16-year-old boy from the low socioeconomic strata, who was diagnosed to have severe HA (FVIII activity <1%) in early childhood.

By early pubertal age, he developed osteoarthropathy of left elbow joint secondary to recurrent episodes of haemarthrosis and due to non-affordability of FVIII therapy. This time, he presented with acute onset, unprovoked, painful, gradually progressive swelling over left side of the back. On examination, there was a large, subcutaneous, mildly tender, non-pitting, non-pulsatile, diffuse swelling over the left side of the back extending

inferiorly up to the inferior angle of left scapula of ~16 cm×12 cm (figure 1).

The standard treatment in such case would be weight-based FVIII concentrate.¹ However, this treatment is very costly and in a resource-limited country, it is usually not feasible or realistic in many cases. Cryoprecipitate and fresh frozen plasma (FFP) are acceptable ways of supplementing FVIII. However, the use of FFP is limited by the volume overload, and it can increase FVIII to a maximum concentration of 15–20% only.^{2 3}

As the patient could not afford FVIII concentrate and bleeding was located in a non-vital region, patient was managed with FFP as a cheap, easily available alternative therapy. He was transfused FFP at the rate of 15 mL/kg/day for 7 days. Excellent response was noted in the form of significant reduction in the swelling and pain, along with improvement in joint mobility in 7 days (figures 1 and 2).



Figure 1 Clinical examination showing large subcutaneous haematoma in the back in the initial period followed by complete resolution of the swelling after fresh frozen plasma transfusion.

Learning points

- ▶ In a resource-constrained situation, management of a non-fatal bleed in haemophilia should be promptly started with alternative therapy, in order to decrease the debility of haemophiliacs.
- ▶ Fresh frozen plasma (FFP) is an effective alternative therapy for non-fatal haematoma in haemophilia-A.
- ▶ Fluid overload and minimal recovery of factor VIII level are major limitations for FFP transfusion.

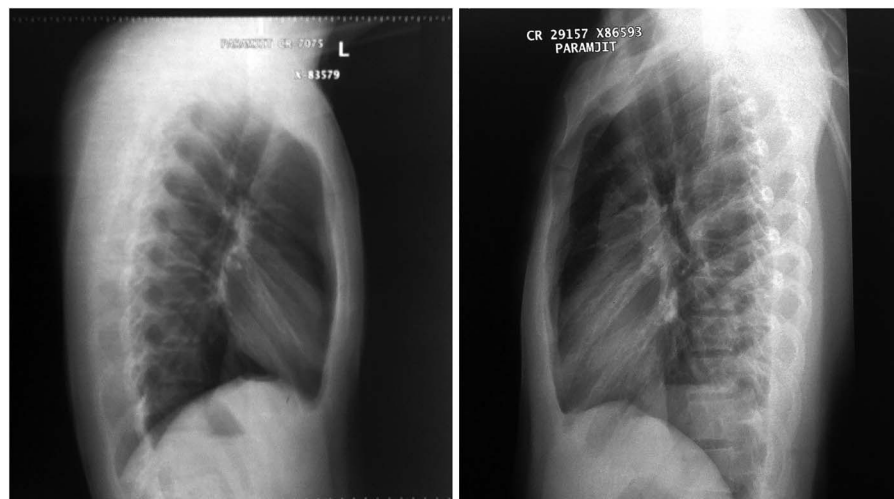


Figure 2 Chest X-ray (lateral view) showing soft tissue haematoma in the back, followed by resolution of soft tissue collection after fresh frozen plasma transfusion.



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