Compartment Syndrome from an Insect Bite: An Unusual Presentation of Hemophilia in an Undiagnosed Patient

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Introduction

Development of forearm compartment syndrome in patients with hemophilia A is a rare phenomenon. We discuss an unusual presentation of hemophilia A in a patient presenting for emergent fasciotomy.

Case Presentation

History: A 4-year-old AA male was admitted to the hospital for left forearm swelling secondary to an insect bite two days prior. His past medical history was significant for frequent ER visits for minor traumas and a right arm injury with delayed healing requiring prolonged casting.

Physical and Labs: VS were BP 121/92, HR 144, T 37.1, RR 20 with a SpO2 100% on room air. The right arm was casted from a previous injury. The left forearm was firm and tender with 2 x 2 cm round hyperpigmented lesion with erythema and edema. Lab showed a WBC 13.2, HCT 35, PLT 445,000, CRP <2.9, ESR 30 and coagulation studies pending. An X-Ray showed soft tissue swelling to the proximal medial aspect of forearm.

Hospital course: The patient was started on ceftriaxone, clindamycin and ketorolac. Pain in the arm increased over the next 2 hours and compartment syndrome was diagnosed. The patient was brought urgently to the operating room for fasciotomy. The nurse informed the operating team that the venipuncture site had required a compression bandage to control bleeding.

Case Presentation

In the operating room, the venipuncture site was still bleeding. Coagulation results showed a PTT > 200, PT 10.9, INR 1.05. A rapid sequence induction was performed, the airway secured and surgery proceeded. Intraoperative labs were drawn for repeat coagulation studies, and factor VIII & IX levels. A tourniquet was placed and a forearm fasciotomy was performed with application of wound vac and estimated blood loss of 20 ml. FFP 1 unit was given with repeat PTT of 41.

A hematology work up revealed a Factor VIII level <0.25 consistent with Hemophilia A. Recombinant factor VIII 50 u/kg bolus was given followed by an infusion at 2 u/kg/hr. On POD #1, the wound vac returned 100 ml blood and HCT dropped to 19 for which PRBC were given and the wound evaluated in the OR. By POD #3, labs normalized and the patient returned to OR for wound closure.

Perioperative Course

Discussion

Hemophilia A is an inherited X-linked recessive disorder of the intrinsic coagulation pathway which affects 1 in 5,000 males. Coagulation testing results in an increased PTT in the context of a normal PT and bleeding time. Severe disease is defined as factor VIII activity < 1% of normal, moderate as 1–5%, and mild as > 5% of normal.

Forearm compartment syndrome is a rare phenomenon and a limb-threatening condition. In our patient, a bleeding condition was suspected without a definitive diagnosis. We therefore proceeded with fasciotomy to preserve the limb and FFP was given. Had Hemophilia A been known, recombinant Factor VIII infusion would have been started preoperatively to restore Factor VIII activity to 100% decreasing blood loss and need for reoperation.

A high index of suspicion should always be maintained in a patient with multiple soft tissue injuries, frequent ED visits and an injury more severe than the history would suggest.

References