Compartment Syndrome Caused by Undiagnosed Acquired Hemophilia

Case Report

Physical exam revealed an elevated blood pressure and heart rate of 151/76 and 117, respectively. All other vital signs were stable. She had palpable pedal pulses with no open wounds. She was unable to perform active dorsiflexion or plantarflexion of the left lower extremity due to pain. She also had extensive tenderness to palpation and tense edema to the left lower extremity. Her sensation was intact to the left leg and foot.

Further workup included CT angiogram (Fig. A) and venous duplex ultrasound (Fig. B) of the left lower extremity. These revealed a large hematoma in the posterior left calf measuring 4.9 x 7.4 x 2.6 cm and edema of the posterior compartment musculature, with heterogeneous mixed hyper/hypointense mass involving the medial head of the gastrocnemius muscle. It was at this time that she was suspected to have compartment syndrome, prompting bedside compartment pressure testing using a Wick Catheter. Testing revealed elevated compartment pressures in the superficial posterior compartment of 80 mmHg, and deep posterior compartment of 40 mmHg. Anterior and lateral leg compartment pressures measured 0-5 mmHg.

The patient was taken to surgery and underwent fasciotomy of the deep and superficial posterior compartments of the left leg. The patient continued to have postoperative bleeding after the fasciotomy. Patient’s wound vac clotted off and had to be removed bedside. She was taken back to the operating room 2 days later for cautery of the bleeding and reapplication of a wound vac. Due to her continued bleeding, a clotting disorder was suspected and a coagulation panel was ordered (Fig. F).

The differential diagnoses for acute lower limb pain are very broad, and during limb threatening conditions, a timely diagnosis is vital. Many factors can confuse or delay diagnosis, including a delayed presentation, lack of trauma, an unusual anatomic location and a lack of known coagulopathy or use of anticoagulation medications. The diagnosis of non-traumatic compartment syndrome requires high index of suspicion along with identifying significant physical and clinical features. Underlying coagulopathy should be investigated in patients with compartment syndrome because there is a high incidence of bleeding disorders in this population (5).

Diagnosis of coagulation disorders is critical for surgical optimization. Acquired hemophilia A is a rare bleeding disorder caused by the development of autoantibodies against clotting factor VIII and is autoimmune in nature. An incidence of only 1,500,000 are symptomatic and it is usually diagnosed by unexplained bleeding after surgical procedures (5). In conclusion, our literature review would indicate this one of only a few recorded cases of acquired hemophilia as the cause of compartment syndrome. A working knowledge of coagulation defects may affect post operative care and prevent additional complications secondary to ongoing bleeding (6).

References: