Atraumatic compartment syndrome of the leg caused by undiagnosed acquired hemophilia A

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Case Presentation

A 63-year-old female with past medical history of diabetes mellitus type 2, tobacco use, hyperlipidemia, and hypertension presented to the emergency department in October of 2018 with complaints of left lower extremity pain and swelling for duration of 3 days. The patient rated the pain as 10/10. The patient stated that over the past few months, she had episodes of similar swelling in her left arm and left thigh, but had resolved spontaneously over a period of one week. She was also experiencing palpitations and shortness of breath. She had been recently admitted to an outside hospital 1 week prior with anemia, hematuria, and epistaxis. Her hemoglobin at that time was 7.0, and she required 4 units of PRBCs during her hospital stay. Hematology was also consulted at that time, and suspected iron deficiency anemia as the cause of her anemia. She was treated with FeSO4, but did not follow up after discharge.

On admission to our facility on October 22, 2018, the patient admitted to non-compliance with her diabetic medications, denied history of trauma, recent travel or history of deep vein thrombosis. 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. 1) and venous duplex ultrasound (Fig. 2) of the left lower extremity. These revealed a large hematoma in the posterior left calf measuring $4.9 \times 7.4 \times 2.6$ 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 were normal, measuring 0–5 mmHg.

The patient was taken to surgery the same day of admission and underwent fasciotomies of the deep and superficial posterior compartments of the left leg. Surgical procedure is described in detail below.

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After induction using general anesthesia, attention was directed to postero-median aspect of the left leg where a longitudinal linear incision was made approximately 12 cm in length along the posterior-medial border of the tibia. The incision was deepened through the subcutaneous layer and the deep fascia was visualized. There was black discoloration to the muscle beneath (Fig. 3). A 10 blade was then utilized to incise the fascia and Metzenbaum scissors were used to release the entire posterior compartment distally and proximally under direct visualization. The muscle was visualized with dark ischemic appearance (Fig. 4). After a period of 10 min, the muscle began to reperfuse (Fig. 5). Deeper dissection was performed into the posterior compartment, and was released as described above. The septum from the posterior aspect of the tibia was then detached to release the deep posterior compartment. Cautery was used to induce contracture of the posterior musculature, demonstrating that the muscle compartments were viable. Wick catheter was utilized to measure the pressures of all compartments of the leg. All compartments were noted to be less than 10 mmHg of pressure. The operative site was then flushed with copious amounts of normal sterile saline. Hemostasis was achieved with bovie cautery. The incision was then dressed with a wound VAC set to 125 mmHg continuous pressure, and a dry sterile dressing. Pt was returned to her inpatient room, with non-weight bearing restrictions to the left lower extremity.

The patient continued to have post-operative 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 cauterization
of the bleeding and reapplication of a wound vac. Due to her continued bleeding, a clotting disorder was suspected and a coagulation panel (Fig. 6) was ordered. Labs revealed an abnormal Partial Thromboplastin Time of 80 and low Hemoglobin of 6. A Von Willebrand panel revealed factor VII level at 2% and factor 8 inhibitor 13 Bethesda units, suggesting an acquired hemophilia. She was successfully treated with intravenous rFVIIa, oral prednisone, and intravenous cyclophosphamide. She was discharged from the hospital in stable condition on October 29, 2018. She was referred to plastic surgery after discharge, and underwent a single skin graft to aid wound closure. Complete healing of the surgical wound was seen at day 50 in December 2018. The patient was seen to have maintained resolution of her acquired hemophilia A at her most recent visit with hematology in December of 2019, with Partial Thromboplastin Time and Hemoglobin within normal limits. At most recent general health follow up in September of 2020, the patient remains pain free with full function to the left leg and foot, without any wounds or other complications the affected left leg.

Discussion

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.

A very similar presentation of atraumatic compartment syndrome of the leg was described by Jentzsch et al., with prompt recognition of the undiagnosed coagulopathy. The patient underwent initial fasciotomy with continued bleeding post operatively leading to the diagnosis of hemophilia. The patient also received initial medical treatment with rFVIIa and corticosteroids, and continued to have uncontrolled bleeding post operatively for several weeks. This led to delayed wound healing, and multiple skin grafts.

Rodriguez-Merchan describes the importance of recognition of hemophilia not only in the setting of soft tissue pathology, but also in the prevention of hemophilic arthropathy. His-review of the literature notes that 90% of hemophilic bleeding occurs in the musculoskeletal system, with 80% occurring within joints. This also emphasizes the need for early diagnosis of hemophilia, to avoid articular destruction and the need for additional surgical management such as joint debridement or replacement.

The differential diagnosis for acute lower limb pain is 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. A working knowledge of coagulation defects may affect post-operative care and prevent additional complications secondary to ongoing bleeding. Literature review would indicate this as only the 3rd case of acquired hemophilia as the cause of non-traumatic compartment syndrome in the leg.

Patient Informed Consent Statement

The authors declare that informed patient consent was taken from all the patients.

Financial Disclosure

None reported

Declaration of Competing Interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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