Spontaneous Spinal Epidural Hematoma in Von Willebrand Disease

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Abstract—Spontaneous spinal epidural hematoma (SSEH) is a rare neurologic emergency with accumulation of blood in the spinal epidural space without trauma or iatrogenic cause. Coagulopathy accounts for 20–30% of all cases with no reported cases associated with von Willebrand disease despite its common occurrence in the general population (1%). The mechanism of hematoma formation remains unclear and may differ depending on the underlying risk for spontaneous hemorrhage. We report a patient with type 1 von Willebrand disease presenting with spontaneous spinal epidural hematoma requiring urgent decompression laminectomy with hematoma evacuation and pretreatment with desmopressin, fresh frozen platelets and anti-hemophilic factor/von Willebrand factor. Von Willebrand disease is not often associated with spontaneous spinal epidural hematoma and when they occur concurrently, an arterial cause such as underlying spinal arteriovenous malformation is more likely.

Keywords — epidural hematoma, von Willebrand disease, spontaneous epidural hematoma, spinal cord compression, coagulopathy

I. INTRODUCTION

Spontaneous spinal epidural hematoma (SSEH) is a relatively rare neurologic emergency with accumulation of blood in the spinal epidural space without obvious traumatic or iatrogenic cause. Early diagnosis and intervention is often hampered by atypical presentation and rapid progression of neurologic deficits resulting in significant morbidity and mortality. Incidence of SSEH is estimated at 0.1/100,000 patients per year with coagulopathy as a cause in 20–30% of all cases (1, 2). Von Willebrand disease (VWD) is a common inherited coagulopathy affecting up to 1% of the population with bleeding into the central nervous system typically associated with trauma (3). There are no reports of SSEH in patients with VWD in the literature despite common occurrence of VWD in the general population. We report a case of rapidly progressive SSEH in a patient with type 1 VWD and discuss pathophysiology and treatment options.

II. CASE REPORT

A 77-year-old woman presented with sudden severe radiating neck pain without neurologic deficits at presentation. Her neck pain began spontaneously without any history of trauma, strenuous exercise, hyperextension injury or notable inciting event. She had a history of bruising but was not on antiplatelets or anticoagulant therapy. Her neck pain initially improved with oral analgesics but she developed rapidly progressive asymmetric weakness, numbness and paresthesias over the next several hours after admission. Repeat motor examination revealed strength of (Medical Research Council Grading) 4/5 in the right arm, 0/5 in the right leg, 4/5 in the left arm and leg. Sensory examination revealed absent light touch, pinprick and temperature sensation both legs with proprioception and vibration decreased in the right leg and a sensory level at the right T3 dermatome. Deep tendon reflexes were depressed on the right and increased on the left with Babinski responses bilaterally.

Her medical history included remote brain aneurysm with surgical clipping and type 1 von-Willebrand disease (VWD). Initial laboratory results were unremarkable without any evidence of hematologic abnormalities. Prothrombin time (PT) was 11.1 seconds (International normalized ratio 1.0), partial thromboplastin time (PIT) was 25.6 seconds and platelet count was 280,000/mm³. Aneurysm clip was not compatible with magnetic resonance imaging. Conventional myelography followed by cervical computed tomography revealed a contrast filling defect extending from C5 through T1 levels consistent with severe cervicothoracic cord compression with impingement of the posterior epidural space (Figure 1, 2). She was diagnosed with spontaneous spinal epidural hematoma (SSEH) and was treated with desmopressin, fresh frozen platelets and anti-hemophilic factor/von Willebrand factor complex. She underwent surgical decompression/laminectomy with hematoma evacuation. Follow-up at one month revealed recovery of sensation in the left leg with persistent deficits in the right leg with strength of 4/5 in the right arm, 2/5 in the right leg and 5/5 on the left side.

III. DISCUSSION

Spontaneous spinal epidural hematoma is a rare neurologic emergency with accumulation of blood in the spinal epidural space without obvious traumatic or iatrogenic cause. Early intervention correlates with better outcomes but diagnosis is
often hampered by atypical presentation and rapid progression. Coagulopathies account for 20–30% of cases with idiopathic cases reported as high as 40% (1). Coagulopathies were reported include hemophilia A (factor VIII deficiency), hemophilia B (factor IX deficiency) or factor XI deficiency with no reported cases involving von Willebrand disease. SSEH commonly involves the cervicothoracic region usually dorsal to the spinal cord spanning greater than three vertebral segments. Presentation involves severe radicular pain with variable signs and symptoms of spinal cord compression with deficits developing and progressing rapidly after onset of pain. Diagnosis is confirmed with imaging modalities including magnetic resonance imaging or computed tomography myelography.

The pathologic mechanism of SSEH remains unclear. One proposed mechanism suggests undulate pressure from thoracic and abdominal cavities causing venous congestion resulting in rupture of the epidural venous plexus into the epidural space due to lack of venous valves within the plexus. Valves typically regulate pressure within the venous system and its absence may account for risk of rupture in situations involving wide fluctuations in pressure such as those caused by lifting or squatting (1).

An arterial mechanism has also been proposed since venous pressure is typically less than intrathecal pressure and cord compression from low-pressure venous hemorrhage is disproportionate to SSEH symptoms (4). In this model, SSEH may be a result of spontaneous rupture of underlying arteriovenous malformations (AVM), which has been reported in 14–18% of SSEH cases. Detection rates for spinal AVMs in SSEH are low due to presence of hematoma and rare use of spinal angiography and may be underreported in SSEH cases. Von Willebrand disease (VWD) is a common inherited bleeding disorder affecting up to 1% of the population with 75% of VWD patients classified as partial quantitative deficiency (type 1) (2). Von Willebrand factor (VWF) plays an important role in coagulation and is involved in several steps in the coagulation cascade. Von Willebrand factor is suspected to be most efficient at areas of high shear stress particularly in narrow arteries and is responsible for decelerating platelets, formation of thrombus, and endothelium repair (5). Von Willebrand disease patients typically experience bleeding in the skin and mucous membrane with symptoms of epistaxis and gastrointestinal hemorrhage. Rare neurologic complications typically involve hemorrhage into the central nervous system caused by trauma.

In the case of our patient, SSEH may be due to an arterial cause since the lack of VWF would predispose to bleeding in high shear stress vessels. Epidural artery rupture and hematoma formation is unusual in VWD since it is typically associated with small vessel disease exhibited by epistaxis and gastrointestinal hemorrhage. Since our patient has a history of cerebral aneurysm, it is possible she may have had an underlying spinal aneurysm or spinal AVM which predisposed her to SSEH in combination with her underlying VWD.

Early surgical intervention with hematoma evacuation remains the mainstay of treatment for symptomatic SSEH. Surgical decompression is particularly recommended for cervical SSEH due to the high morbidity of cases treated conservatively (6). Delay in surgical decompression results in worsening morbidity/mortality with poor recovery in cases beyond 12 hours of onset (7).

In patients with VWD or Hemophilia, pretreatment with desmopressin (ddAVP) and Antihemophilic factor/von Willebrand factor complex reduces the risk of additional hemorrhage perioperatively in patients undergoing surgery. Patients with type 1 VWD receiving ddAVP have the same incidence of hematoma perioperatively as patients without coagulopathy (8). Replacement therapy with vWF concentrate is recommended for significant hemorrhages in patients with type 1 VWD in whom either DDAVP is ineffective or contraindicated. There is no data on treatment with desmopressin and antihemophilic factor/von Willebrand in...
either conservative treatment or surgical decompression of SSEH in the setting of VWD.

In summary, spontaneous spinal epidural hematoma is a neurologic emergency requiring quick diagnosis and urgent treatment. While coagulopathy is comorbid in nearly one third of patients with SSEH, VWD is not commonly associated likely due to the specific nature of its coagulopathy effect. An arterial cause for SSEH is most likely in cases where VWD is present.

References

**Questions (please choose one single answer):**

1. What percentage of spontaneous spinal epidural hematomas are associated with underlying coagulopathies?
   A. 10%
   B. 20%
   C. 50%
   D. 70%

2. What is the most common presenting symptom in patients with spontaneous spinal epidural hematoma?
   A. Urinary incontinence
   B. Severe radicular pain
   C. Paresthesias
   D. Ataxia

3. What is the mainstay of treatment in spontaneous spinal epidural hematoma?
   A. Corticosteroids
   B. Pain management alone
   C. Early surgical intervention with evacuation
   D. A & B

Correct answer: 1: B; 2: B; 3 C.