CASE REPORT

Intracranial bleeding following soccer-related head trauma in a young student with occult factor VII deficiency

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Abstract

It is important to obtain coagulation tests to assess bleeding risk in trauma patients undergoing emergency surgery when a bleeding disorder may be obscured. Identifying specific clotting factor defects is critical in successful patient management.

KEVWORDS

coagulation testing, emergency trauma surgery, factor VII deficiency, intracranial bleed, soccerrelated trauma

1 | INTRODUCTION

Soccer is the most popular and fastest growing sport, especially among teenage women, and it carries an inherent risk of head injury. Head collision can be even more serious and life-threatening in patients with an undiagnosed underlying bleeding disorder. Therefore, obtaining basic coagulation tests prior to emergency trauma surgery is of great importance. Our aim was to describe an uncommon but important clinical scenario that highlights the benefit of pre-operative testing with prothrombin time (PT) and activated partial thromboplastin time (aPTT) for bleeding risk in trauma patients undergoing emergency surgery despite the absence of past, personal, and family history of bleeding. Abnormal test

results of a coagulation test should raise suspicion for an obscure bleeding disorder that could be inherited or acquired (such as occult factor VII or acquired factor deficiency). Identifying specific types of clotting factor defects is critical in successful management of patients.

2 | CASE PRESENTATION

A 17-year-old woman without significant past medical history experienced a head-on collision with another player of the opposing team during a heading dual of a high school soccer match. She complained of severe pain in the left fronto-temporal region without loss of consciousness.

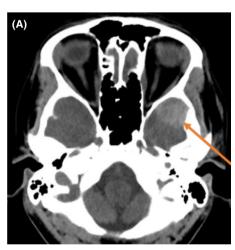
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After evaluation by the team coach and the staff who followed the concussion protocol, she resumed to play, but soon had to leave the field due to symptoms of dizziness, worsening headache on the left side, blurry vision, and nausea. She reported symptoms of generalized numbness and had two episodes of projectile vomiting during transfer to emergency room in an ambulance. Patient denied any bleeding diathesis in childhood, while family history and social histories were noncontributory. Clinical examination showed a young, thinly built high school student of average height reporting severe dull headache of left side. Her vital signs were significant for a pulse rate of 57/minute, and physical examination showing multiple ecchymosis limited to shins of both legs that patient attributed to be soccer-related trauma. Her systemic examination was unremarkable as she was fully alert and oriented to person, place, and time. The speech and cranial nerves II-XII were normal, and there was no focal motor, sensory, or cerebellar deficits. Routine laboratory tests were unremarkable, while a computed tomography (CT) scan of the head without contrast revealed a lenticular hyperdensity in the left



Axial plane:



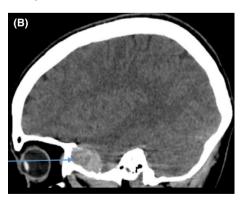
Sagittal plane:

FIGURE 1 CT head without IV contrast. Initial. Axial section displays left middle cranial fossa hyperdensity measuring 24×19 mm (orange arrow in A). Sagittal section reveals lentiform shape, typical of epidural hematoma (orange arrow in B)

middle cranial fossa (Figure 1) suspected to be a localized bleeding versus a meningioma. While closely monitoring her symptoms in the next 24 hours, patient experienced worsening headache and developed new-onset right upper and lower extremity weakness (muscle strength 4 out of 5). A repeat CT scan of the head showed an increase in size of the lentiform lesion in the left middle cranial fossa suggesting an enlarging epidural hematoma (Figure 2). In preparation for surgical evacuation of the hematoma, a pre-operative workup of bleeding risk was assessed that showed elevated prothrombin time (PT) and a normal activated partial thromboplastin time (APTT). Factor VII deficiency was suspected in the setting of an intracranial bleed despite no known previous personal or family history of bleeding. Pre-operative peripheral blood specimen was sent for coagulation tests, and patient was infused activated prothrombin complex concentrate (APCC) and vitamin K



Axial plane:



Sagittal plane:

FIGURE 2 CT head without IV contrast. Subsequent. Left middle cranial fossa hyperdensity has now increased to 30×24 mm. Lentiform hyperdensity has increased to 29×18 mm indicating epidural hematoma expansion, a surgical emergency (blue arrow)

prior and during craniotomy. Operative findings revealed lacerations of the left middle meningeal artery at multiple points along with extradural collection of a large size hematoma that was then successfully evacuated.

The pre-operative coagulation test results revealed correction of PT following mixing with normal plasma, consistent with a diagnosis of factor VII deficiency, estimated at 36% of normal (normal range 60%-140%). The levels of factor II, IX, and X were normal. Patient underwent an uneventful postoperative recovery during which she received recombinant factor VIIa (rVIIa) infusions with normalization of hemostasis.

3 | DISCUSSION

A diagnosis of inherited bleeding disorder is typically made early in childhood from bleeding symptoms and a positive family history. However, a small proportion of inherited bleeding disorders may remain occult and undiagnosed until later in childhood when investigations into excessive bleeding episode in diverse clinical settings are triggered. Fc VII deficiency is estimated to affect 1 in 300 000 to 1 in 500 000 people. ²

In our patient, the diagnosis of factor VII deficiency was made following emergent pre-operative workup of soccer-related head trauma, a risk associated with a fast growing sport in schools and communities.^{3,4} Although patient reported no bleeding history in the past, unusual ecchymosis of both legs was attributed to soccer-related trauma, confounding the clinical possibility of an underlying bleeding disorder. Nevertheless, testing for bleeding risk before an emergent surgical procedure led to the diagnosis of an occult factor VII deficiency and successful perioperative management with intraoperative infusion of APCC/vitamin K and postoperative recombinant factor VII infusion.

A panel of blood tests containing PT and APTT serves to screen patients for underlying coagulation defects in those undergoing surgery. Some studies indicate that routine pre-operative hemostatic tests are unreliable and poor predictors of perioperative bleeding and therefore generally not recommended. As per general guidelines for testing of bleeding risk in patients undergoing emergent surgery, PT and APTT testing is not routinely recommended if a structured history for spontaneous or excessive bleeding in response to trauma is absent, both in patient and his/her family. As

However, our patient's detection of factor VII deficiency was possible by pre-operative testing of bleeding diathesis in the setting of head trauma despite lack of past or family history of bleeding. Our case exemplifies the value of performing pre-operative testing of bleeding risk in a patient undergoing trauma-related emergency surgery. Our case

also suggests that guidelines for pre-operative testing of bleeding risk following traumatic injury distinguish itself from bleeding risk for emergency surgery not provoked by trauma.

Factor VII deficiency is suspected when PT is prolonged and APTT is normal, in association with normal liver function tests. However, applying general guidelines could miss an underlying rare bleeding disorder such as occult factor VII or acquired factor deficiency, due to their presentation in patients later in life and absence of family history given an autosomal recessive mode of inheritance.

A great deal of work is being done in diagnosing and achieving successful management of patients with factor VII deficiency. ^{10,11} Although the principles of management of this disorder are similar to those of commonly encountered inherited defects of coagulation, perioperative management of factor VII deficiency is conspicuous by lack of correlation between factor VII level and the risk of bleeding. In a study involving patients with factor VII deficiency undergoing surgery and receiving perioperative replacement therapy with recombinant factor VII, previous history of major bleeding and not factor VII level or type of surgery correlated with amount, frequency, and duration of replacement therapy. ¹² This information is important in the patient management since overtreatment with replacement therapy leads to a high risk of clotting complications.

Our patient's motivation to continue to play soccer necessitated extrapolation of management principles developed in children diagnosed with hemophilia A, B, and von Willebrand disease. Such efforts translate into improved self-esteem, motivation, and physical and emotional development of a child.

Our case represents an uncommon but important clinical scenario that highlights the importance to consider obtaining coagulation tests to assess the bleeding risk in trauma patients undergoing emergency surgery when a bleeding disorder may be obscured as seen in our case. Identifying specific types of clotting factor defects is critical in successful management of patients.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTION

NLL: MD involved in idea formulation, preparation, creation, and/or presentation of the published work, specifically writing the initial draft. AL: MD assisted in critical review of initial draft. DSA: MBBS assisted in critical review of initial draft. SS: MBBS assisted in critical review of initial draft. LW: MD assisted in critical review of initial draft. UH: MD involved in Idea formulation, preparation, creation, and/or presentation of the published work, assisted in critical review of initial and subsequent drafts.

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