

Large subgaleal hematoma as a presentation of parahemophilia

Sir,

Subgaleal hematoma is a potentially life-threatening extracranial bleed that occurs most commonly in neonates after difficult instrumental deliveries.^[1] Its occurrence beyond the neonatal period is rare and is often associated with head trauma involving tangential or radial forces applied to the scalp causing emissary veins traversing the subgaleal space to be ruptured.^[2] Large subgaleal hematoma due to trivial head trauma in a parahemophilic patient with no reported literature is interesting to describe. Parahemophilia, a relatively rare hemorrhagic disorder affecting both sexes, is due to a congenital and frequently familial deficiency of FV. This autosomal recessive disorder varies greatly in severity. The most commonly reported symptoms are bleeding from mucosal surfaces and postoperative hemorrhage. However, hemarthroses, intramuscular, and intracranial hemorrhages can also occur. Subgaleal hematoma associated with parahemophilia, as seen in our case, is yet to be reported in the literature.

A 15-year-old boy presented with progressive head enlargement in a period of 3 days after sustaining trivial head trauma due to assault. He had history of soft tissue and joint bleeding since child hood. There was no history of recent drug intake or fever. On clinical examination, he was fully conscious and alert with no neurological abnormality, but appeared ill with severe

pallor. There were no lymph node enlargements or other manifestations of bleeding tendency such as purpura or ecchymosis. The abdominal examination was normal. Head examination revealed large soft tissue swelling of the entire scalp demonstrated by pressure indentation with normal overlying skin. The head circumference was 67 cm [Figure 1]. Computed tomography (CT) scan of head showed large subgaleal hematoma involving both side with no intracranial hemorrhage, no midline shift, and no skull fractures [Figure 2].

Suspicion of associated coagulopathy arose in our mind. The screening coagulation tests revealed a prolonged activated partial thromboplastin time (aPTT) and marked prolongation of prothrombin time (PT) [Table 1]. Determination of coagulation factor activities yielded normal results, while FV activity was 17% (normal value > 60%). The liver function test was within normal limit. The plasma concentrations of d-dimers, fibrinogen, antithrombin III, protein C, protein S, and plasminogen were found to be normal. There were no antiphospholipid antibodies and no lupus anticoagulant.

The patient received five units of fresh frozen plasma (FFP) transfusions, without complications. A pressure bandage was applied to the scalp and forehead. When aPTT and PT returned to normal range, the hematoma resolved and the head size became normal with improvement of the general condition [Figure 3]. We investigated available members of the family and found the patient's brother with the same deficiency (FV activity, 35%). Other family members were free from same disorder.

Most reported cases of subgaleal hematoma are neonates and the reported incidence of subgaleal hematoma ranges from 1.6 to 3/1000 live births.^[3] Subgaleal hematoma occurring beyond the neonatal period is rare and is often associated with head trauma involving tangential or radial forces applied to the scalp causing emissary veins traversing the subgaleal space to be ruptured.^[2]

Table 1: Laboratory data demonstrating the patient's coagulation profile

Test name	Result value	Reference range
aPTT		
Control	27.2 seconds	
Test	>60.0 seconds	Within 5-7 seconds of control
PT		
Control	11.0 seconds	
Test	>60.0 seconds	Within 3-5 seconds of control
Factor assay		
Factor VIII (%)	106	71-110
Factor IX (%)	104	74-110
Factor V (%)	17	>60



Figure 1: Clinical photograph of the patient at the time of admission showing large soft tissue swelling of the entire scalp and the head circumference measuring 67 cm

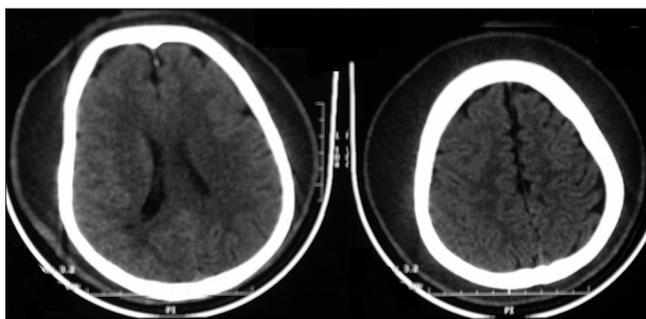


Figure 2: Computed tomography (CT) scan head showing circumferential scalp swelling around skull bones



Figure 3: Clinical photograph of the patient at the time of discharge with complete resolution of scalp soft tissue swelling and normal head size

The diagnosis is generally made clinically, with a fluctuant boggy mass developing gradually over the scalp. The swelling frequently crosses suture lines. Imaging studies such as CT scan head helps to know associated intracranial pathology if any. Detection of

the cause of subgaleal hematoma is very important in its management. It had been reported in children with vitamin K deficiency,^[4] hemophilia,^[5] factor XIII deficiency,^[6] and von Willebrand disease^[7] which can be treated by adequate replacement therapy.

Activated FV (FVa) is the cofactor in the prothrombinase complex that cleaves and activates prothrombin to thrombin, thus participates in the coagulation cascade.^[8,9] Owren first described the deficiency of blood coagulation FV leading to a rare hemorrhagic tendency known as parahemophilia or Owren's disease.^[10] The most commonly reported symptoms are bleeding from mucosal surfaces and postoperative hemorrhage. However, hemarthroses and intramuscular and intracranial hemorrhages can also occur. Subgaleal hematoma associated with parahemophilia, as seen in our case, is yet to be reported in the literature.

Hematological evaluation of a patient with a subgaleal hematoma associated with relatively mild or trivial trauma should include a complete blood cell count, PT, and aPTT. Typically, FV deficiency is first suspected in a patient with bleeding symptoms who has a prolongation of both the PT and aPTT. If a low FV activity is discovered, then FV deficiency must be distinguished from consumptive coagulopathy, liver disease, and combined FV and factor VIII (FVIII) deficiencies. The clinical setting is often sufficient to differentiate FV deficiency from disseminated intravascular coagulation or liver disease, but testing for d-dimers, fibrinogen levels, and liver dysfunction or damage may be useful. A FVIII level is necessary to distinguish isolated FV deficiency from the combined deficiency of FV and FVIII and may help in distinguishing congenital FV deficiency from that owing to liver failure, as FVIII levels are often elevated in liver dysfunction.

Because no FV-specific concentrate is available, FFP is the primary therapeutic option. Most patients are only treated episodically for bleeding and before invasive procedures.^[11] For acute hemorrhage, the goal of therapy is to maintain FV levels above 20%. The half-life of FV is 12-36 hours, and, typically, daily infusions of 15-20 ml/kg of FFP are sufficient.^[12] However, the frequency and dosing should be adjusted empirically to achieve hemostasis.

Most subgaleal hematomas do not require aspiration and drainage, as the risk of introducing infection outweighs the benefit of the procedure. The natural course is for spontaneous resolution in a few days or weeks without complication.^[13] Our patient was managed conservatively with frequent FFP transfusions and a pressure bandage.

No surgical aspiration was done to avoid infection and severe bleeding.

To the best of our knowledge, this is the first case to be reported with large subgaleal hematoma involving the entire scalp caused by a trivial trauma in a parahemophilic boy. Factor V deficiency should be considered in patients who present with large subgaleal hematoma following a trivial head injury. An excellent response to treatment with FFP can be expected. Conservative management with frequent FFP transfusions and a pressure bandage obviates the need for surgical aspiration and avoid the risk of introducing infection and severe bleeding.

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References

1. Benaron DA. Subgaleal hematoma causing hypovolumic shock during delivery after failed vacuum extraction: Case report. *J Perinatol* 1993;13:228-31.
2. Vu TT, Guerrero MF, Hamburger EK, Klein BL. Subgaleal hematoma from hair braiding. Case report and literature review. *Pediatr Emerg Care* 2004;20:821-3.
3. Gebremariam A. Subgaleal hemorrhage: Risk factors and neurological and developmental outcome in survivors. *Ann Trop Paediatr* 1999;19:45-50.
5. Rohyans JA, Miser AW, Miser JS. Subgaleal hemorrhage in infants with hemophilia: Report of two cases and review of the literature. *Pediatrics* 1982;70:306-7.
6. Maruki C, Nakajima M, Tsunoda A, Ebato M, Ikeya F. A case of giant expanding cephalhematoma: Does the administration of blood coagulation factor XIII reverse symptoms? *Surg Neurol* 2003;60:138-41.
7. Rafffini L, Tsarouhas N. Subgaleal hematoma from hair braiding leads to a diagnosis of von Willebrand disease. *Pediatr Emerg Care* 2004;20: 316-8.
8. Kalafatis M. Coagulation factor V: A plethora of anticoagulant molecules. *Curr Opin Hematol* 2005;12:141-8.
9. Asselta R, Tenchini ML, Duga S. Inherited defects of coagulation factor V: The hemorrhagic side. *J Thromb Haemost* 2006;4:26-34.
10. Owren PA. Parahaemophilia; Haemorrhagic diathesis due to absence of previously unknown clotting factor. *Lancet* 1947;1:446-8.
11. Acharya SS, Coughlin A, DiMichele DM; North American Rare Bleeding Disorder Study Group. Rare Bleeding Disorder Registry: Deficiencies of factors II, V, VII, X, XIII, fibrinogen and dysfibrinogenemias. *J Thromb Haemost* 2004;2:248-56.
12. Mannucci PM, Duga S, Peyvandi F. Recessively inherited coagulation disorders. *Blood* 2004;104:1243-52.
13. Kirkpatrick JS, Gower DJ, Chauvenet A, Kelly DL Jr. Subgaleal hematoma in a child, without skull fracture. *Dev Med Child Neurol* 1986;28:506-14.

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