Subgaleal Hematoma with Bilateral Proptosis as a Manifestation of Fibrinogen and Factor X Deficiency - A Case Report

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INTRODUCTION

Subgaleal hematomas are usually seen after high-velocity head trauma; however, their presence following trivial injury should raise suspicion regarding an associated clotting disorder. Here we describe an interesting patient who presented to us with bilateral proptosis associated with a subgaleal hematoma due to fibrinogen and factor X deficiency. This condition can result in serious bleeding diathesis often necessitating multiple surgeries to control haemorrhage.

Our patient, an 11-year-old male child presented to us 4 weeks after a minor head injury. He had a gradually progressive soft swelling on his forehead with proptosis of both eyes. Aspiration of the swelling at a nearby private hospital had revealed blood. The swelling recurred over a span of 2 weeks.

On neurological examination, there were no positive findings. On local examination, he had a large, non-tender subgaleal hematoma over the bilateral fronto-temporo-parieto-occipital region of his head extending to both eyes. There was associated bilateral subconjunctival haemorrhage, peri orbital ecchymosis and gross proptosis. The vision was affected on the right side, finger counting at 1 foot. Left side vision was intact. Extraocular movements were restricted in both eyes in all planes. CT scan of the brain showed subgaleal hematoma over the bilateral fronto-temporo-parieto-occipital region with an extension over the supraorbital ridge into the bilateral orbit beneath the orbital roof, there were bilateral relatively symmetrical areas of subperiosteal haemorrhages involving superior aspect of orbit causing proptosis and inferior displacement of the globes. On haematological examination, his haemoglobin was 5 gm/dl. Platelet count was 240,000/cumm. On coagulation profile, prothrombin time was 45 seconds (normal range 11.1-14.5 seconds) with an activated partial thromboplastin time of 39.5 seconds (normal range 28.1-39.7 seconds), and thrombin time was 58 seconds. The plasma fibrinogen levels were < 100 mg/dl (normal range- 200-400 mg/dl). D-dimer was strongly positive. The patient also had factor X deficiency. The peripheral blood smear had a normocytic normochromic picture.
CLINICAL DIAGNOSIS

Subgaleal haematoma due to coagulation factor deficiency.

DISCUSSION OF MANAGEMENT

As the patient was anaemic, he was given adequate packed red cell transfusion according to age and weight. Fibrinogen and Factor X deficiency was corrected by adequate FFP transfusion. After correcting the coagulation profile, Subgaleal hematoma was aspirated by an 18 G IV cannula under aseptic precautions. 500 cc dark brown motor-oil like fluid was aspirated and a tight crepe bandage was applied covering the entire surface of the head. Repeat CT was s/o no residual subgaleal collection. The compressive dressing was continued for 4 weeks. Ophthalmologists performed the aspiration of a subperiosteal hematoma under local anaesthesia. Proptosis gradually decreased over two weeks.

PATHOLOGICAL DISCUSSION

The subgaleal space contains emissary veins that connect the veins of the scalp to the intracranial dural venous sinuses. High velocity, shearing injury on these vessels results in subgaleal hematomas i.e., haemorrhage between the epicranial aponeurosis and the pericranium. The orbital septum acts as a barrier between the facial and orbital structures. The anterior layer of the levator palpebral aponeurosis blends with the posterior part of the orbital septum except in the lateral canthal region where there is continuity of the cranial subgaleal space with the potential space between the levator palpebrae aponeurosis and orbital periosteum. Thus the subgaleal space communicates anteriorly over the superior orbital ridge with the superior orbit, allowing blood to track into the periorbital space as shown in Fig. 5. This anatomical feature, recognized by Gioia et al. in 1987 and elaborated on by Kim and Taragin, explains the delayed occurrence of proptosis in patients with large subgaleal hematomas.

Orbital Hematomas and Proptosis

Although trauma is the most common cause of subgaleal hematomas and proptosis, haematological diseases, including congenital hypofibrinogenemia, heterozygous factor VII deficiency, and sickle cell anaemia (due to marrow hyperplasia), are known to be etiological factors too. Subgaleal hematomas usually occur 1–14 days after trauma and spontaneously resolve within 1–3 weeks; the majority not requiring any intervention. Subperiosteal hematomas of the orbit and extradural hematomas from orbital fractures can mimic subgaleal hematomas, as all three can cause proptosis and visual deficits.

Congenital dysfibrinogenemia is an abnormality of fibrinogen molecules which includes defects in steps of fibrin formation and stabilization. In a normal coagulation cascade, thrombin cleaves fibrinogen to release fibrinopeptide-A and fibrinopeptide-B. The fibrin monomer thus formed, gets polymerized to form an insoluble fibrin clot which is stabilized by Factor XIIIa. Subsequent degradation of the clot takes place with the concerted activity of plasminogen and its activator. Defects in any of these steps can lead to dysfibrinogenaemia. Hence, its presentation is variable ranging from abnormal clinical bleeding thrombotic tendency and defective wound healing to no clinically apparent disease. However, the most common defect occurs in the polymerisation of the fibrin clot. Congenital dysfibrinogenemia is inherited as an autosomal dominant. Most of these patients are heterozygotes, few are homozygotes and rare are compound heterozygotes. About 40% of these patients are asymptomatic, 45–50% of these have bleeding disorders and the remaining 10–15% have thrombotic tendencies. The bleeding associated with dysfibrinogenaemia is generally mild and includes soft tissue haemorrhage, easy bruising and menorrhagia. Intraoperative and postoperative bleeding have also been reported but none were found to be life-threatening. Acquired dysfibrinogenaemia occurs mainly in patients with chronic liver disease. Other causes include hepatocellular carcinoma, obstructive biliary disease, and immune disorders (e.g., multiple myeloma and systemic lupus erythematosus). In our case, the patient had prolonged prothrombin time, activated partial thromboplastin time and thrombin time, with decreased fibrinogen level and normal factor VIII-C level. The laboratory results observed in this case are consistent with other studies of congenital dysfibrinogenemia. Platelet count was found to be within normal limits which helped to exclude the possibility of disseminated intravascular coagulation. Liver function test and liver enzymes were normal, excluding acquired dysfibrinogenaemia. The patient was not on any anticoagulants. Thus, considering all the above findings, a diagnosis of congenital dysfibrinogenemia was given.

Among coagulopathies, Von Willebrand disease is the most common inherited bleeding disorder. Our patient happened to have the rarest form of coagulopathy, caused by a deficiency of Factor X. The incidence of Factor X deficiency is estimated at 1 in 1000000 births. It is inherited in an autosomal recessive fashion.
This case report highlights anatomical communication between subgaleal space and the lateral aspect of the roof of the orbit. Thus, large subgaleal hematomas can extend into this potential space, resulting in severe proptosis and visual disturbances.

Hence, in a young patient with large subgaleal hematoma due to trivial injury associated with an altered coagulation profile, a possibility of rare disorders like congenital dysfibrinogenemia and factor X deficiency should always be thought of, as they can be fatal if missed.

REFERENCES


