CASE REPORT

Recurrent Post-Traumatic Epidural Hematoma Revealing Factor XIII Deficiency: A Case Report

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Abstract

Trauma is a leading cause of intracranial bleeding. When these traumatic brain injuries are paired with rare bleeding disorders, management may be challenging. A 37-year-old male was brought to the emergency room with an acute subdural hematoma. He underwent craniotomy and evacuation of the hematoma. He developed recurrent postoperative epidural hematomas on consecutive days despite normal routine coagulation studies. FXIII deficiency was confirmed, and empirical cryoprecipitate and platelet transfusion stabilized his condition. He underwent successful cranioplasty under factor support and recovered without neurological deficits. Mortality is high in patients with traumatic brain injury. Outcomes of surgical management depend on GCS on arrival and the time since injury. The presence of coagulation disorders worsens these outcomes. Unusual bleeding should prompt early evaluation and multidisciplinary input.

Keywords: *Subdural hematoma, trauma, factor XIII, traumatic brain injury, and coagulation disorders.*

INTRODUCTION

Acute or chronic subdural hematomas (SDH) and epidural hematomas are the two most common classifications of traumatic brain injuries (TBI). Acute SDH (ASDH) is typically accompanied by underlying parenchymal brain injury when it occurs in conjunction with severe traumatic brain injury. The rupture of a surface or bridging vessel, which usually occurs after an acceleration-deceleration motion, results in hemorrhage in the subdural region [1]. The most common cause of acute SDH is head trauma brought on by attacks, falls, or accidents involving motor vehicles [2, 3]. According to one study, acute SDH in the context of significant head trauma is projected to have a hospitalization mortality rate of 11.8% [4].

Factor XIII (F XIII) deficiency is a relatively infrequent coagulation disorder, and it is classified into acquired and congenital forms [5]. Henoch-Schönlein purpura, inflammatory bowel disease, disseminated intravascular coagulopathy, liver disease, and sepsis are the primary causes of the acquired form of F XIII deficiency [5, 6]. The congenital form is an autosomal recessive condition with an estimated incidence of 1 in 3-5 million people [7].

In such coagulation disorders, the assessment of possible risks and benefits of surgery in these individuals is particularly challenging. We report here recurrences of an epidural hematoma after a surgery in a patient with congenital Factor XIII deficiency.

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CASE PRESENTATION

A 37-year-old male with no prior comorbidities presented to the emergency department after a road traffic accident with an isolated head injury. On arrival, his Glasgow Coma Scale (GCS) was E1V1M4 = 6/15. He was managed according to ATLS protocols and intubated. CT brain revealed an acute subdural hematoma in the right frontoparietal and temporal convexity (width 1 cm) with 1.1 cm leftward midline shift (**Fig. 1**). A linear occipital bone fracture extending to the foramen magnum was also noted. Urgent craniotomy and evacuation were performed, and he was kept intubated in the ICU.

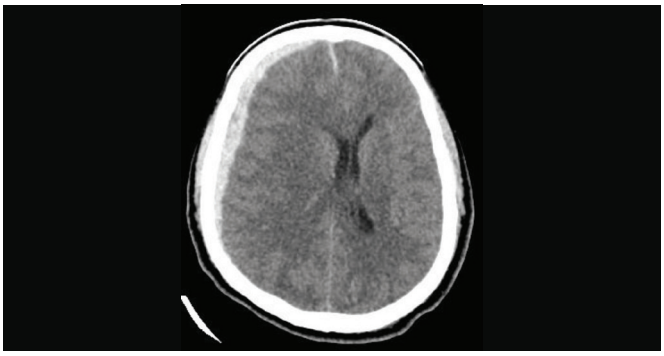

Fig. (1): Acute subdural hematoma in the right frontoparietal and temporal convexity (width 1 cm) with 1.1 cm leftward midline shift.

Postoperative CT showed a right frontoparietal epidural hematoma (width 2.1 cm, shift 1.1 cm) (**Fig. 2**). A repeat craniotomy with dural hitch sutures was done, during which excessive scalp bleeding was observed. He remained intubated postoperatively.

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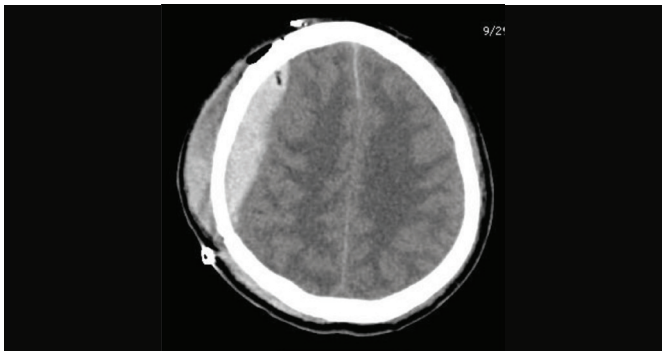

Fig. (2): Epidural hematoma in the right frontoparietal convexity (width 2.1 cm) with 1.1 cm contralateral shift.

The following day, CT demonstrated a recurrent epidural hematoma (width 1.5 cm, shift 1.5 cm) (**Fig. 3**). Coagulation profile (PT, APTT, platelets) remained normal. Hematology consultation was obtained, and factor XIII levels were checked. The patient was empirically transfused with platelets and cryoprecipitate, followed by craniectomy with bone flap placement in the abdominal wall. Intraoperative bleeding was less than before.

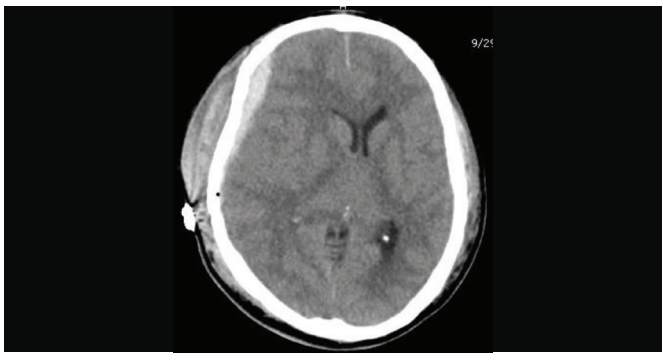

Fig. (3): Recurrent epidural hematoma in the same region (width 1.5 cm) with 1.5 cm contralateral shift.

A subsequent CT showed near-complete resolution of the hematoma with improved midline shift (**Fig. 4**). The patient's GCS normalized, he was extubated, and regained independent ambulation within two days. Oral intake was tolerated, and he was discharged on the fourth postoperative day. Factor XIII level returned at 54% (normal: 70-140%).

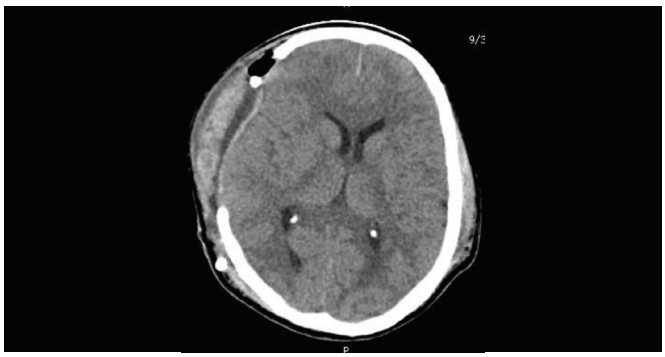

Fig. (4): Near-complete resolution of epidural hematoma with marked improvement in midline shift.

One month later, he was readmitted for elective cranioplasty (**Fig. 5**). Preoperatively, six cryoprecipitates were administered. Intraoperatively, persistent scalp oozing prompted repeat factor XIII assessment, and additional platelets were transfused. Recovery was uneventful, and he was discharged the following day. Post-transfusion factor XIII was 117%.

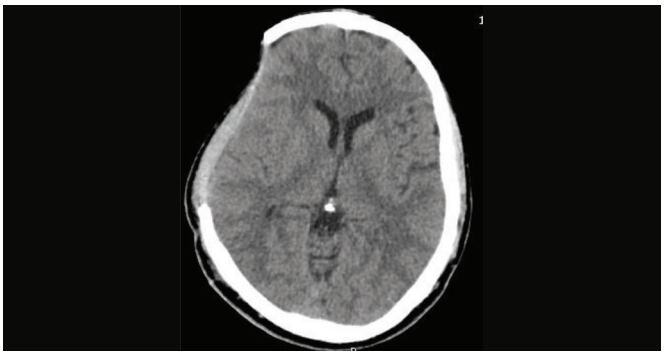

Fig. (5): Craniotomy defect.

At one-year follow-up, he remained well with no deficits or spontaneous bleeding episodes (**Fig. 6**).

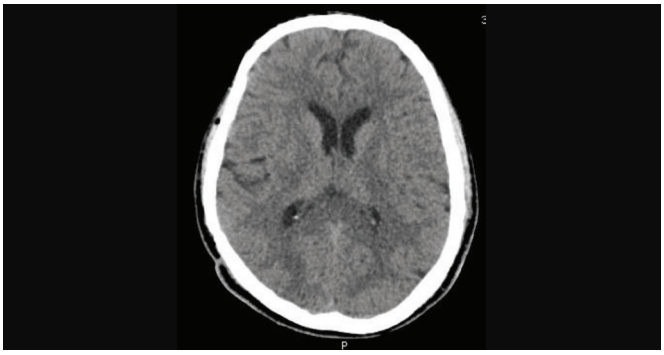

Fig. (6): One-year follow-up CT scan.

DISCUSSION

Traumatic SDH with brainstem compression typically presents with altered mental status, pupillary asymmetry, and abnormal posturing [2]. According to studies, assaults, falls, and traffic-related mishaps make for 53% (range: 30-73%), 30% (range: 7-52%), and 8% (range: 1-19%), respectively, of all intracranial bleeds [2, 8].

GCS at presentation is the strongest predictor of surgical outcome in intracranial bleeds; patients with GCS 3-5 in acute SDH have a 74% mortality rate [9]. Prognosis in intracranial bleeding is also influenced by age, pupillary changes, associated lesions, delay to surgery, and intracranial pressure. (ICP) [2].

Surgical decisions in ASDH rely on hematoma thickness and midline shift; evacuation is recommended if thickness >10 mm or shift >5 mm, regardless of GCS [2, 10, 11]. Early, aggressive evacuation of ASDH lowers mortality; comatose patients operated within 4 hours show ~30% mortality *versus* ~90% when delayed [12, 13]. Surgical

treatments for acute SDH are craniotomy, craniotomy with dural grafting, or decompressive craniectomy [12].

Factor XIII is one of the rarest coagulation disorders [14]. FXIII is a protransglutaminase vital for coagulation, circulating as a plasma tetramer (FXIIIA2B2) with catalytic A subunits from bone marrow cells and carrier B subunits from the liver [15, 16]. It may be autosomal recessive or sex-linked [17]. In most situations, the clinical characteristics and bleeding propensity in hereditary FXIII with activity <3% can be severe. Following trauma, persistent bleeding, ecchymosis, hematomas, and intracranial bleeding are common [18, 19].

Factor XIII deficiency is treated with plasma-derived (Corifact/Fibrogammin P) or recombinant FXIII-A2 concentrate [20, 21]. Replacement therapy includes FFP (10 ml/kg every 4-6 weeks), cryoprecipitate (1 bag/10-20 kg every 3-4 weeks), or recombinant FXIII [22, 23]. Recombinant FXIII (rFXIII) is also available [24-26]. For surgery, FXIII levels should be >10-20%; give 20-30 U/kg daily for 2-3 days preoperatively [24, 27]. To maintain FXIII >10% and prevent worsening hemorrhage, it is recommended to administer 10-20 U/kg of fibrogammin every other day for 2 weeks [18, 28, 29].

CONCLUSION

TBI commonly presents with epidural or subdural hematomas, but management is challenging with coagulation disorders. In patients with recurrent intracranial hematomas and routine coagulation tests, rare clotting factor deficiencies, such as factor XIII deficiency, should be considered. Early testing and targeted replacement therapy can significantly improve outcomes.

CONSENT FOR PUBLICATION

Written informed consent was taken from the patient.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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Declared none.

AUTHORS' CONTRIBUTION

AS: Conception of study, Manuscript writing, revision, final approval, and agreement of accountability for all aspects of the work.

HH: Design of study, Manuscript writing, revision, final approval, and agreement of accountability for all aspects of the work.

YR: Conception of study, Critical review of manuscript, revision, final approval, and agreement of accountability for all aspects of the work.

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