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Case Report

Patient with a history of Glanzmann thrombasthenia presented with chronic subdural hematoma: a case report study

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Abstract

Glanzmann thrombasthenia (GT) is a rare plateletdisorder characterized by qualitative/quantitative deficiencies of the platelets' fibrinogen receptor, glycoprotein (GP) IIb/IIIa complex, resulting in impaired platelet aggregation and increased bleeding time. Most cases are hereditary with an autosomal recessive pattern of inheritance, but acquired GT also occurs. We report the surgical management of symptomatic chronic subdural hematoma (CSDH), a rare condition in young individuals, in a 37-year-old man who had GT and a history of mild head trauma approximately one month before admission. Despite hematologic consultation, normal bleeding time and clotting time, and platelet transfusion before surgery, massive hemorrhage during surgery, epidural hematoma, and anisocoria in the ICU occurred that led to craniectomy. This report highlights that CSDH management in patients with GT requires close monitoring of these patients as well as collaboration between neurosurgeons, intensive care physicians, hematologists, and anesthesiologists.

INTRODUCTION

Glanzmann thrombasthenia (GT) is a rare bleeding disorder featured with qualitative or quantitative deficiencies of a fibrinogen receptor (glycoprotein (GP) IIb/IIIa) on the platelets, leading to absence of/impaired platelet aggregation, decreased clot retraction, and increased bleeding time while the platelet count is low to normal [1]. Although GT is mostly an inherited autosomal recessive disorder, acquired GT may also occur due to autoimmunization [1]. GT has a prevalence of one per million individuals and is more frequent in countries with a higher incidence of consanguinity [2].

Chronic subdural hematomas (CSDH) are liquefied hematomas in the subdural space with a characteristic outer membrane that occurs a few weeks after head injury and one of its risk factors is thrombopathy [3]. CSDH are typically seen in the elderly population and are rare in young adults [4]. The most common theory for the cause of CSDH is that minor inertial brain injury causes movement of the brain within the skull and tears bridging veins as they traverse the cell layer of the dural border [3]. Without timely interventions, CSDH may be fatal despite their benign nature [4]. It is consensus that patients with symptoms that can be attributed to radiologically confirmed CSDH should be treated [3].

Platelet transfusion is the standard treatment for patients with GT as a countermeasure for major surgeries. Recombinant activated factor VII (rFVIIa) has also been used in patients with antiplatelet antibodies that render transfusion ineffective [5]. A lack of scientific knowledge of rare diseases and their appropriate management may result in poor treatment outcomes. Thus, this study aimed to report the case of a young male with GT who had symptomatic CSDH.

CASE PRESENTATION

A 37-year-old man was admitted to the emergency department of Taleghani Hospital (Kermanshah, Iran) with complaints of intractable new onset headache, nausea, vomiting, and confusion. Approximately 4 weeks before admission, he had a mild head trauma. Past medical history was remarkable for opioid abuse, intermittent spontaneous otorrhagia controlled by repeated platelet transfusion and GT. However, his laboratory data was unremarkable.

On the examination at admission, he showed normal vital signs. Although the patient's score on the Glasgow Coma Scale (GCS) was 13/15, no other remarkable abnormalities were found by neurological examination. Due to impaired consciousness and a history of head injury, the patient underwent a brain CT scan that revealed a significant hypo-dense left frontoparietal hematoma and the CSDH were diagnosed. Thus, a burr-hole craniostomy was performed.

Although the preoperative testings showed normal bleeding time (BT) and clotting time (CT), the patient received 5 units of platelet preoperatively after consultation with a hematologist. Surgery was started with two burr holes in the frontoparietal area.

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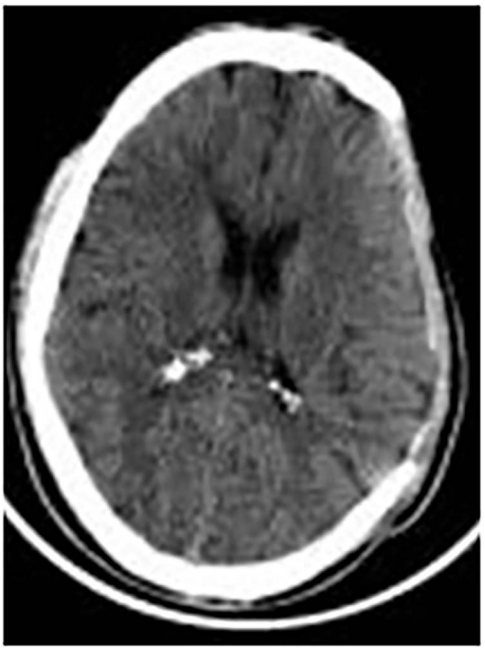

Figure 1. CT scan after the second operation.

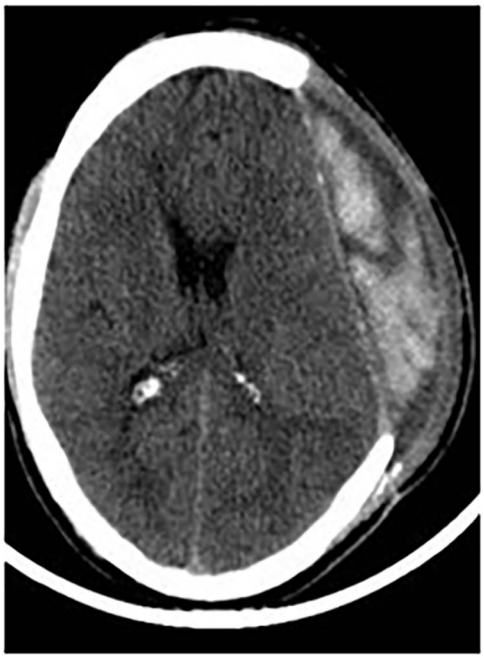

Figure 2. CT scan before the third operation.

The hematoma was evacuated but massive and uncontrollable hemorrhage was seen through the bone and scalp. As possible, partial hemostasis was achieved by bone wax and surgical hemostasis dressing (surgicel) and bipolar cautery, drains were fixed, and the scalp was stitched.

Then, the patient was admitted to the intensive care unit (ICU) by concomitant packed cell transfusion. However, his GCS score began to decrease and he became anisocoric. Immediately, red blood cell and platelet transfusion was done and reoperation was conducted by craniectomy. The epidural and subdural hematoma was evacuated and hemostasis was acceptably achieved by surgicel, bone wax, and bipolar. The dural tack-up stitch was done, the drain was fixed and the scalp was stitched by continuous vertical mattress technique (Fig. 1). Due to the possibility of massive hemorrhage during subcutaneous bone flap storage, the bone flap was discarded. During the craniectomy and until 48 h, the patient was infused with intravenous tranexamic acid 2 gr every 6 h and fibrinogen 1 gr every 8 h. He spent 5 days in the ICU and 5 days on the ward before discharge. From the first surgery until discharge, he received 35 units of pooled platelet, 7 packed cells, and 4 FFP.

One month later, he was admitted because of a headache and the CT scan showed epidural hematomas (Fig. 2). Hence, he was taken to the operating room for hematoma evacuation. In the laboratory examination, increased BT (10 min) was found, and 2 units P.C., 2 units single donor platelet (SDP), and 33 units random donor platelet (RDP) were transfused preoperatively, leading to decreased BT (4 min). After surgery, he received 2 units P.C., 2 units platelet, and 5 units RDP and was discharged home after 5 days.

Two months later, he was admitted for cranioplasty, and a hematology and anesthesiology consult was done. BT and CT were normal but perioperatively he received 35 units RDP, 2 units SDPs, and 5 units P.C. Despite a huge platelet transfusion before and after cranioplasty, bleeding in the ward was reported to the neurosurgery resident by the nurse. It wasn't controlled by overrunning stitches because of bleeding in the needle insertion sites. Finally, bleeding was controlled by tight elastic bandages. After 5 days, the patient was discharged. On follow-up at 2 months, he had no neurologic deficit or residual symptoms.

DISCUSSION

GT is an extremely rare inherited disorder of platelet function with an autosomal recessive mode of inheritance. However, acquired GT due to autoimmunization has also been reported. GT is characterized by qualitative/quantitative deficits of a platelet's fibrinogen receptor, GP IIb/IIIa, resulting in impaired platelet aggregation and increased bleeding time [1]. This rare disorder is more common in populations with a higher rate of consanguinity [2]. Common clinical manifestations of GT include spontaneous mucosal and skin bleeding [6]. There are three subtypes for GT, but no correlation has been found between the bleeding severity and the GT subtype [7].

No therapy is required in patients with GT regularly. However, treatment is always required for these patients during spontaneous bleeding episodes as well as surgical procedures, which are significant challenges due to bleeding probability and a high incidence of alloimmunization induced by repeated platelet transfusion [1, 7]. Local measures (e.g. local pressure, ice therapy, topical thrombin, fibrin sealants, etc.) and/or antifibrinolytic agents (e.g. tranexamic acid) are usually effective in the case of mild to moderate bleeding episodes. Platelet transfusion is the standard treatment for moderate to severe bleeding and the routine prophylaxis for major surgeries in patients with GT [1]. Alternative treatment with rFVIIa has also been recommended for patients who are alloimmunized, but it is expensive and associated with the risk of thrombosis [1, 7, 8].

CSDH are usually caused by slow bleeding from bridging veins on the cerebral surface. However, its pathophysiology has not yet well been understood. While CSDH is one of the most common cases of neurosurgery, it is usually seen in elderly subjects and is rare in young individuals [3]. Its management is based on the presence and severity of hematoma-related symptoms and hematoma size and mass effect. Surgery is required for symptomatic CSDH, hematoma thickness > 10 mm, or midline shift

> 7 mm [9]. The burr-hole craniostomy with/without a drainage system is the most common and most efficient surgical technique used for CSDH management. In the case of hematoma recurrence, burr-hole conversion into a craniotomy should be considered [9]. CSDH is a benign condition, nonetheless, it may be fatal without prompt surgical intervention and hematoma recurrence is still an important concern [4].

To the best of the authors' knowledge, there are no case reports of CSDH management in young patients with GT. As mentioned previously, the classical measures were done in our case, but simple neurosurgery for hematoma evacuation became complicated and nearly fatal despite the teamwork of the neurosurgeon, hematologist, and anesthesiologist. Unfortunately, there are no established recommendations for treating and prophylaxis of surgical bleeding in patients with GT and a combination of functional platelet tests and clinical assessment is used for assessing the response to treatment [10]. However, a useful management algorithm has been proposed by Solh et al. for elective and urgent surgical interventions in these patients [1].

CONCLUSION

We reported a rare case of a young patient with a CSDH who also had rare GT and a history of repeated platelet transfusion to control spontaneous otorrhagia. This report highlights the need for close monitoring in patients with GT when neurosurgery is done for CSDH management. One important point is that despite having normal BT and CT, the patient's bleeding during and after the surgery was difficult to control. Therefore, as previous studies have also shown, BT and CT are poor predictors of clinical bleeding. In light of our experience with this case, we suggest if a patient with CSDH and GT is neurologically and hemodynamically stable, the neurosurgeons should be more prudent and perform more investigations as needed, like what was suggested by Solh et al. Further, close collaboration between neurosurgeons, intensive care physicians, hematologists, and anesthesiologists is critical for CSDH management in patients with GT.

CONFLICT OF INTEREST STATEMENT

None declared.

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There was no funding for this study.

ETHICAL APPROVAL

No ethical approval was needed for this case report.

CONSENT FOR PUBLICATION

The patient gave written informed consent for the publication of this report.

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