Acute Epidural Hematoma in a Patient With Glanzmann’s Thrombasthenia
—Case Report—

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Abstract
A 7-year-old girl with Glanzmann’s thrombasthenia (GT) fell and hit her head against a table. Within 2 hours she began to vomit and became drowsy. On admission to our hospital her Glasgow Coma Scale score was 13. Computed tomography (CT) on admission showed acute epidural hematoma in the left posterior fossa. We administered platelets, performed emergent lateral suboccipital craniotomy, and totally removed the epidural hematoma. Postoperative CT showed no evidence of hematoma or re-bleeding. She was discharged without neurological deficits 14 days after the operation. GT is a platelet aggregation disorder due to a functional loss of platelet membrane glycoprotein IIb/IIIa. The present patient with GT underwent successful emergency craniotomy after platelet transfusion.

Key words: Glanzmann’s thrombasthenia, acute epidural hematoma, head injury, management, platelet transfusion

Introduction
Glanzmann’s thrombasthenia (GT), a rare inherited platelet disorder, is characterized by a complete lack of platelet aggregation due to a defect in the platelet membrane receptor complex (IIb/IIIa) for fibrinogen.1,2,7) Epistaxis, purplish-type bleeding, gum bleeding, and menorrhagia are common clinical manifestations of GT. Spontaneous bleeding is uncommon, but posttraumatic and postoperative hemorrhage may be particularly serious in these patients.2,7) Acute epidural hematoma is common after head injury and frequently requires emergent surgical intervention. However, management strategies must take into account the surgical risks in GT patients. There are few reports on the treatment of GT patients with head injury.2,7)

We treated a GT patient who sustained acute epidural hematoma after a head injury and was successfully treated after platelet transfusion.

Case Report
A 7-year-old girl with GT fell and hit her occiput against the corner of a table. She weighed 20 kg. She had suffered several episodes of subcutaneous bleeding and was diagnosed with GT in early childhood. Two hours after her fall she began to vomit and was taken to our hospital. On admission, she was drowsy with Glasgow Coma Scale (GCS) score of 13 (E3V4M6). Computed tomography (CT) showed acute epidural hematoma in the left posterior fossa under a fracture of the occipital bone (Fig. 1). We decided that immediate surgical intervention was necessary and delivered 10 units of platelets by transfusion to prevent intra- and postoperative hemorrhage. Her platelet count was 199,000/mm³ before and 221,000/mm³ after the transfusion. Left lateral occipital and suboccipital craniotomy was then performed without removing the occipital bone covering the transverse sinus. Intraoperative findings revealed fresh epidural clot mixed with uncoagulated blood covering the transverse sinus. We identified the left transverse sinus as the bleeding source. Meticulous hemostasis was applied throughout the surgical procedure. Her total blood loss was 20 ml. Postoperative CT demonstrated that the hematoma had been successfully removed, with no evidence of rebleeding (Fig. 2). Her consciousness level improved postoperatively, but refractory oozing from the wound appeared on the day after the operation. She required administration of an additional 10 units of platelets to stop the oozing. There were no further complications. She was discharged without neurological deficits 14 days after the operation.
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Discussion

Our patient with GT was treated for traumatic epidural hematoma after a fall. GT is attributable to abnormalities in platelet membrane glycoprotein IIb/IIIa resulting in the absence of platelet aggregation ability. Laboratory studies showed prolonged bleeding time with no or decreased retraction and a normal platelet count. Coagulation studies were normal. The lack of platelet aggregation in response to all physiologic stimuli is diagnostic of GT. Flow cytometry is the method of choice for diagnostic confirmation because procedures now exist for the quantitation of the residual glycoprotein IIb/IIIa complex content in platelets.

Spontaneous, unprovoked bleeding is rare in patients with GT, but profuse bleeding may occur after trauma or surgical procedures. Although no definitive management strategies have been developed for GT patients undergoing surgery or suffering trauma, platelet transfusion is the standard treatment for severe bleeding and for surgical support. An intracerebral cavernoma was surgically excised in a GT patient. Platelets were transfused perioperatively and the clinical course was uneventful. One of 2 GT patients with intracerebral hematoma after head injury required surgical evacuation and platelet transfusion. Our patient manifested an acceptable response to platelet transfusion because intraoperative hemostasis was obtained without difficulty.

Repeated platelet transfusions may elicit alloimmunization to human leukocyte antigens and/or platelet membrane glycoprotein IIb/IIIa, rendering future transfusions ineffective. Platelet transfusions also raise the risk for adverse reactions including virus transmission and may not be readily available to patients in remote areas.

The first successful use of recombinant factor VIIa to address severe epistaxis in a boy with GT was reported in 1996. Additional case reports have suggested the usefulness of this agent in patients with traumatic bleeding and for surgical prophylaxis in patients with congenital functional platelet disorders including GT. The mechanism underlying its action is still not completely understood. Thrombin generation is impaired in GT patients. The ability of high-dose recombinant factor VIIa to improve thrombin generation via direct binding to activated platelets and/or by overcoming the inhibitory effect of zymogen factor VII may contribute to its therapeutic efficacy in GT patients.

GT is very rare, but patients with GT may experience a traumatic emergency. Platelet transfusion remains the first-line therapy in GT patients with bleeding and as prophylaxis in GT patients scheduled for surgery unless they harbor antibodies to platelets. The administration of recombinant factor VIIa may be a potent management alternative in thrombasthenic patients with antiplatelet antibodies.

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References


Fig. 1 Computed tomography scans on admission showing supra- and infratentorial epidural hematoma in the left posterior fossa. Bone image revealing a fracture of the left occipital bone (arrow).

Fig. 2 Postoperative computed tomography scans showing the epidural hematoma in the posterior fossa has been completely removed.
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