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Letters to the editor

Recombinant activated factor VII and epsilon aminocaproic acid treatment of a patient with Glanzmann's thrombasthenia for nasal polipectomy

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To the editor: We hereby present an anesthesia course consisting of a regimen starting with epsilon-aminocaproic acid (tranexamic acid) prophylaxis and continuing with recombinant activated factor VII (rFVIIa) administration pre- and postoperatively, supplemented by platelet transfusion, in a patient with Glanzmann's thrombasthenia (GT), a congenital bleeding disorder caused by the lack of platelet surface glycoprotein (GP) IIb/IIIa receptor. Platelets are normal in number and size, and adhesion is normal, but aggregation occurs only in response to ristocetin [1]. Two articles, published by Poon et al. [2], and Poon and d'Oiron [3], encouraged us to register our case at the data collection website www.glanzmann-reg.org and to share this case with our colleagues in this issue of the *Journal of Anesthesia*.

A 24-year old male patient with a diagnosis of GT was scheduled to undergo nasal polipectomy by functional endoscopic sinus surgery under general anesthesia. In the patient's medical history, he had been diagnosed with essential thrombocytopenia at the age of 5 years and when he underwent circumcision he received one pack of platelets.

According to the GT treatment strategy indicated by our institution's hematologists, intravenous (i.v.) tranexamic acid (Transamine ampule, 10%, 250 mg; Fako, Ankara, Turkey) four times 750 mg was ordered within 48–72 h preoperatively and 7 to 10 days postoperatively. Additionally, an i.v. bolus dose of 90 $\mu g \cdot kg^{-1}$ of rFVIIa (NovoSeven flacon, 2.4 mg (120 KIU; Novo Nordisk, Istanbul, Turkey) was ordered 5 min before the start of surgery and repeated twice, at 2-h intervals, followed by another two such doses in 4 h. Two packs of platelets collected by apheresis were prepared in advance.

The patient received i.v. tranexamic acid 72 h before the operation. After the initial i.v. dose of 480 µg rFVIIa was administered, thiopentone sodium 5 $mg \cdot kg^{-1}$ and 0.1 $mg \cdot kg^{-1}$of vecuronium bromide were administered to the 55-kg patient for the induction of anesthesia; anesthesia maintenance was provided by desflurane in an oxygen-nitrous oxide mixture. The operation and anesthesia were completed uneventfully in approximately 2 h. The total amount of blood loss was approximately 300 ml. It has been recommended that, in patients with GT, mucocutaneous bleeding that is considered as minor bleeding could be controlled by local measures such as compression or the use of a gelatin sponge or gauze alone or dipped in tranexamic acid or topical thrombin, and it has also been reported that there was a possibility of the development of a vicious cyle of rebleeding [4]. Therefore, we administered a second dose of rFVIIa in order to stop bleeding when we observed apparent nosebleed from the gauzes in the recovery room.

Because the preoperative hemoglobin decreased from 16.7 to 7.7 $g \cdot dl^{-1}$ and the platelet count decreased from 249 to 130 $\times$ $10^9 \cdot l^{-1}$, third and fourth doses were given at 2-h intervals, and a fifth dose was given simultaneously with one pack of platelets in the ward. When hemoglobin and platelets returned to normal baseline values, the patient was discharged.

Anesthesiologists are rarely faced with patients having GT undergoing surgery. Vaginal hysterectomy, pediatric open cardiac surgery, and herniorrhaphy were some of the reported procedures requiring anesthesia in such patients, and were treated with similar immediate pre- and postoperative effective therapeutic regimens [5–7]. Although platelet transfusion therapy has been an accepted treatment of GT bleeding for years, it may result in the development of antibodies to GP IIb/IIIa, rendering further transfusions ineffective [1,2].

The optimal dose and mode of rFVIIa administration are still under investigation at the present time, and bolus infusions of 80–85 $\mu g \cdot kg^{-1}$ or more at 2-h intervals have been recommended, if one to three doses were not sufficient to arrest the bleeding, more doses were given according to the current treatment strategies [4].

In conclusion, in patients with GT, the present therapy protocol, including rFVIIa and tranexamic, acid could be safely used, without adverse effects (particularly thrombotic events), in combination with platelet transfusion, based on the case reported here.

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