APPROACHING AND MANAGING GLANZMANN THROMBASTHENIA: A CASE REPORT

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BACKGROUND

Glanzmann Thrombasthenia (GT) is a rare inherited autosomal recessive platelet function disorder. It is caused by mutations of its surface glycoproteins (GP) IIb/IIIa that act as a fibrinogen receptor, important for mediating platelet aggregation induced by physiologic agonists and for fibrin clot retraction¹. Patients frequently have severe hemorrhagic conditions and, although platelet transfusion remains the standard treatment, it can be associated with anaphylaxis and refractoriness.

CASE REPORT

- 22-year-old female patient with GT proposed for correction of thoracolumbar scoliosis.
- 8 months old - Recurrent episodes of epistaxis and post-traumatic hemorrhages which became progressively more sporadic and self-limited until the age of 7.
- 5 years old - Specific blood screening was performed leading to GT diagnosis as levels of GPIIb/IIIa were below 1%, GPIb was normal and antiplatelet antibodies were not found.
- 8 years old - Platelet transfusion was performed before a tooth extraction without any intercurrent.
- 11 years old – Platelet transfusion was performed before appendectomy and she suffered an preoperative anaphylactic reaction to platelets.
- Preoperative approach - Due to her history and presence of mild anemia, it was decided to start iron supplementation 2 weeks before surgery.
- Intra / Postoperative approach - Tranexamic acid (TXA) was administered at the induction and recombinant factor VII (rFVIIa) during the procedure and the following 3 days and a controlled hypotension was maintained. For postoperative analgesia, an endovenous patient-controlled device was considered safer in order to avoid the possible risks of a spinal technique. She was discharged 6 days after surgery.

DISCUSSION

Anaphylactic reactions to platelets occur in less than 20% of the transfusions but are highly heterogeneous². Epidural analgesia is an aid in controlling the intense postoperative pain of this surgical procedure but, as a platelet disorder, GT requires a focused approach to minimize adverse outcomes³. Regarding the lack of studies on its management, these patients should be viewed as the ones taking antiplatelet drugs and the ideal scenario should include a pro-coagulation status allowing the manipulation of the neuraxis.

CONCLUSION

Albeit rare, these platelet disorders can be clinically severe and a correct evaluation is mandatory both for diagnosis and treatment. Their anesthetic management can also be challenging but still needs research to be clarified and a protocol for reliable methods of coagulation study should be created.

REFERENCES

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