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ANESTHETIC CONSIDERATIONS IN A CHILD WITH GLANZMANN'S THROMBASTHENIA: A CASE REPORT

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ABSTRACT

Glanzmann's thrombasthenia (GT) is a rare inherited platelet disorder causing defective aggregation and severe bleeding risk. We report a seven-year-old boy with GT who underwent circumcision and dental extraction under sevoflurane sedation. A single intravenous dose of recombinant activated factor VII (rFVIIa) 90 µg/kg was given preoperatively. The procedure and recovery were uneventful, with no bleeding or transfusion required. rFVIIa proved effective in achieving hemostasis by bypassing the platelet defect and promoting thrombin generation. Anesthetic management should avoid invasive or bleeding-prone techniques, emphasizing multidisciplinary coordination to ensure patient safety.

KEYWORDS

Glanzmann's thrombasthenia, rFVIIa, anesthesia, pediatric, bleeding disorder

MAIN ARTICLE

INTRODUCTION

Glanzmann's thrombasthenia (GT) is a rare hereditary bleeding disorder with autosomal recessive transmission, characterized by a qualitative or quantitative defect of the platelet receptor integrin αIIbβ3 (glycoprotein IIb/IIIa), which is essential for platelet aggregation [1]. First described by Eduard Glanzmann in 1918, the disease manifests as mucocutaneous bleeding such as epistaxis, gingival bleeding, bruising, and gastrointestinal or gynecological hemorrhages often appearing in early childhood [2].

The diagnosis is established by a normal platelet count with absent or severely impaired platelet aggregation, as demonstrated by platelet aggregation studies and flow cytometry [3]. Management includes local hemostatic measures, antifibrinolytics, platelet transfusions, and, when ineffective or contraindicated, administration of recombinant activated factor VII (rFVIIa) [4].

Surgical and anesthetic procedures pose a major hemorrhagic risk and require meticulous preparation and multidisciplinary coordination [5,6].

This report describes the anesthetic management of a 7-year-old boy with Glanzmann's thrombasthenia who underwent sedation for circumcision and dental extraction, successfully managed with rFVIIa.

CASE REPORT

A 7-year-old boy weighing 20 kg, born to consanguineous parents, had been followed since the age of 18 months for Glanzmann's thrombasthenia confirmed by platelet aggregometry. He was admitted for elective circumcision combined with dental extraction, both considered procedures with high bleeding risk.

The child had no history of massive transfusion or known anti-HLA alloimmunization. Physical examination showed no cutaneous bleeding signs and no predictors of difficult intubation or ventilation.

Preoperative laboratory tests revealed a normal platelet count of 259,000/µL and a hemoglobin level of 10.4 g/dL.

Sedation was performed with sevoflurane under continuous monitoring of vital signs (oxygen saturation, heart rate, and blood pressure).

Induction was smooth and atraumatic. After insertion of a 22-gauge peripheral venous catheter, a slow intravenous bolus of recombinant activated factor VII (rFVIIa) at a dose of 90 µg/kg was administered.

No intraoperative bleeding occurred. Sedation was maintained until completion of the procedure, and the child was transferred to the post-anesthesia care unit for observation. Postoperative recovery was uneventful, with no secondary bleeding or anemia. The clinical course was favorable, and the patient was discharged after 24 hours of observation.

DISCUSSION

Anesthetic management of patients with Glanzmann's thrombasthenia remains a major challenge due to the high risk of severe bleeding and possible platelet transfusion refractoriness related to anti-HLA or anti-αIIbβ3 alloimmunization [7].

According to the recommendations of the French Reference Center for Inherited Platelet Disorders, close collaboration between anesthesiologists, surgeons, and hematologists is essential to establish a tailored perioperative protocol. [1].

Use of HLA-compatible platelet transfusions and rFVIIa therapy (90–120 µg/kg every 2 hours) is recommended for surgical procedures or in cases of transfusion failure [1].

rFVIIa acts by bypassing the platelet defect, directly activating factor X on the platelet surface, which results in thrombin generation and stable clot formation [8].

Several studies have demonstrated its efficacy and safety in minor surgical procedures particularly dental interventions with success rates exceeding 94% and no significant thromboembolic events [2].

In the present case, a single 90 µg/kg dose of rFVIIa, effectively prevented hemorrhagic complications.

This approach illustrates the value of an individualized treatment plan based on the type and severity of surgery.

From an anesthetic standpoint, the literature emphasizes the importance of avoiding invasive or bleeding-prone techniques such as spinal anesthesia, deep nerve blocks, or traumatic intubation [9].

Use of thromboelastography (TEG), can be helpful in guiding platelet transfusion and assessing clot quality [3].

This case aligns with current data highlighting the effectiveness of rFVIIa and the critical importance of multidisciplinary coordination.

CONCLUSION

Glanzmann's thrombasthenia requires highly specialized anesthetic management, even for minor surgical procedures.

The use of rFVIIa, combined with platelet transfusion and antifibrinolytic therapy, provides an effective strategy to prevent bleeding complications.

This case underscores the importance of an anticipatory protocol, multidisciplinary planning, and rigorous monitoring, particularly in pediatric patients.

Current international guidelines support individualized perioperative approaches tailored to the type of procedure and the patient's transfusion history.

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