

INTRAVENOUS REGIONAL ANALGESIA IN A PATIENT WITH GLANZMANN THROMBASTENIA

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Abstract

Glanzmann thrombastenia (GT) is a rare condition of an inherited autosomal recessive gene characterized with bleeding tendency. The condition is rarely met in the OR. and therefore it is essential that anesthesiologist be cognizant of the risk involved and be prepared with all necessary precautionary measures. We present a GT case in a 27 year old male with a mass in the anticubital region of right wrist that was successfully excised using the non-invasive intravenous regional analgesia (IVRA).

The use of platelet transfusion and the recombinant factor VIIa, are stressed.

Key words: Glanzmann thrombastenia, Intravenous regional anesthesia, Recombinant factor VIIa, surgery.

Introduction

Glanzmann thrombastenia (GT) is a rare condition associated with fatal bleeding. Three hundred such cases have been reported in the literature¹. It is a trombocyte aggregation deficiency inherited autosomal recessive, characterized with bleeding tendency because of the deficiency of number or function of glycoprotein IIb/IIIa complex found on trombocyte membrane². Bleeding time is significantly increased although platelet count, morphology, prothrombine time (PT) and activated partial thromboplastin time (aPTT) values are normal^{2,3}. Most commonly seen bleeding symptoms in these patients are purpura, epistaxis, gingival bleeding and menorrhagia³. Any surgical or invasive procedures can be high bleeding risk in these patients. Platelet transfusion is the only way to stop bleeding. It is, ironic however, that excess platelet transfusion may also increase the bleeding risk due to alloimmunisation⁴.

We present our experience with intravenous regional anesthesia (IVRA) and share information on Glanzmann thrombastenia (GT).

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Case Report

A 27 years-old and 78 kg male, was admitted for excision of a mass in his right antecubital wrist region. For the previous 22 years, patient had been diagnosed as Glanzmann thrombastenia. Except for thrombastenia, the preanesthetic evaluation was not contributory. Preoperative laboratory findings revealed: Hct 45%, Plt 161000/mm³, PT 16.1 sec, INR 1.29, aPTT 32.6 sec, AST 15 U/L, ALT 11 U/L, Glucose 99 mg/dl, Urea 22 mg/dl, Creatinine 0.82 mg/dl, Factor IX 98% (normal range 60-150%), Factor VIII 99% (normal range 60-150%). Radiological examination showed a 32 × 20 mm solid mass with regular borders in the right wrist antecubital region. A Hematological consultation emphasized that anesthetic and surgical interventions could be highly risky because of the diagnosis of GT and the antibodies formed against platelets in the patient.

In the operating room (OR), monitoring consisted of ECG, NIBP and pulse oximetry. An intravenous (iv) line, 20-gauge cannula, was done at his left arm. NaCl 0.9% infusion at 3 ml/kg/hr started. Premedication consisted of Midazolam (Dormicum ® roche) 1 mg. To start the IVRA, a 22-gauge cannula was introduced in dorsum of right hand. Following exanguination of blood from right arm using the Esmarch bandage and double layered tourniquet inflated, 3 mg/kg lidocaine (Aritmal ® Biosel) (0.9% NaCl solution was added to make up a total volume of 40 mL), was given intravenously.

Vital signs remained normal during surgery. 2400 units rFVIIa (NovoSeven ® Novo Nordisk) was given 1 hour before and at the beginning of the operation. Bleeding amount was 100 ml approximately. One apheresis platelet suspension was given during operation. After tourniquet deflation, there was some blood leakage from the incision site. After bleeding stopped, patient was transferred to the service ward without any problems. A total 7200 units rFVIIa was given at 2, 6, and 12 hrs postoperatively.

Discussion

Life threatening difficult to stop hemorrhages mostly develop secondary to trauma or surgical procedures in Glanzmann thrombastenic (GT) patients.

The administration of recombinant activated factor VIIa is safe and effective in such cases. Blood and blood products (platelet rich plasma, platelet concentrates) can also be given^{4,5}.

The early diagnosis of GT is most important, because only platelet transfusion can be life saving. Platelet agrometria, thromboelastographic techniques, bleeding time measurements, are the methods for follow up of platelet function. However, the platelet agrometria and thromboelastographic techniques are not commonly used⁴⁻⁷.

Anesthesiologists rarely meet GT in the OR. All preparation and precautionary measures must be taken. Without the adequate preparation of patient, all anesthetics carry risk factors. Tracheal intubation for one, can cause intractable bleeding and difficulty in airway management. Nasogastric catheters and other oral interventions, can cause bleeding⁵. Central and peripheral blocks also have bleeding risk.. It is therefore essential that the advantages and disadvantages of a central block should be weighed carefully before being done.

In situations such as the GT, minimally invasive analgesic techniques should be the procedure of choice and an IVRA technique should be highly considered.

Literature review revealed that surgery of GT patients has been done both under general anesthesia and local anesthesia^{2,3,5,8,9}. However, no IVRA technique has been reported in such cases.

The IVRA technique used in our case is minimally invasive and produces bloodless field of the surgical area. Bleeding risk, however, after tourniquet deflation is a disadvantage. Due to the high pressure produced by the IVRA in the tourniquet region, tissue damage may occur.

No serious bleeding or tissue damage was observed during and after operation in our patient. It is probable that having given platelet suspension and rFVIIa preoperatively, may have a salutary effect in the prevention of bleeding.. The rFVIIa transfusion is approved in intractable life threatening bleeding situation. It must also be realized that although platelet transfusion is the only accepted treatment in GT, this platelet transfusion may cause antibody formation against Glicoprotein IIb/IIIa, and this can inactivate

succeeding transfusions⁴.

It can be concluded that intravenous regional analgesia (IVRA) provides a good non-invasive

analgesic technique in the management of patients with Glanzmann thrombasthenia.

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