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BALANCED GENERAL ANESTHESIA IN A PATIENT WITH HEMOPHILIC ARTHROPATHY: A CASE REPORT

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ABSTRACT

Introduction: One of the main complications observed in patients diagnosed with Hemophilia B is hematrosis, a chronic joint condition that causes pain and limited movement, often requiring total arthroplasty of the affected joint. **Case report:** Patient MGS, male, 19 years old, with Hemophilia B, diagnosed with hemophilic arthropathy of the left knee. General anesthesia was chosen for the case, unlike neuraxial blockade due to a higher risk of spinal canal hematoma. The procedure was uneventful. **Discussion:** Hemophilic arthropathy is characterized by a chronic and progressive inflammatory process that leads to stiffness and limited joint movement. Treatment includes conservative management, which may evolve to the need for synovectomy, arthrodesis and, in more severe cases, arthroplasty. However, since hemophilic patients have a higher risk of significant bleeding, some considerations are relevant, such as avoiding unnecessary airway management, avoiding mucosal trauma and avoiding neuraxial blockade due to the risk of causing spinal canal hematoma. Therefore, general anesthesia would be the safest option for such patients. It is also worth mentioning that optimizing coagulation factor levels in the preoperative period can reduce complications, as can the administration of activated recombinant factor VII. Therefore, this article presents a case report of a hemophiliac patient with a high risk of bleeding, requiring specific anesthetic considerations to minimize the chance of major hemorrhagic complications.

Keywords: Hemophilia B, Hemartrhosis, General anesthesia, Arthroplasty, Spinal anesthesia.

INTRODUCTION

Characterized as a coagulation disorder caused by a deficiency of factor IX, Hemophilia B results from an X-linked recessive disorder, with no predilection for geographic area or ethnicity, occurring in approximately 1 in 30,000 male births.[1]

One of the main characteristics of the disease is hemarthrosis, especially in the joints of the knees, elbows, and ankles, causing pain and limited mobility. Intra-articular bleeding leads to synovitis, where proteolytic enzymes cause cartilage degeneration, characterizing hemophilic arthropathy,

which may also present with arthritis, restricted movement, and chronic pain.[1]

From the anesthetic perspective, in many cases that are indicated and eligible for surgical treatment of such complications—such as total knee arthroplasty, a common procedure among patients with hemophilic arthropathy—one would expect the use of neuraxial blockade as the anesthetic technique for this approach, given its low complication rate, shorter hospital stay, and fewer admissions to the intensive care unit (ICU). However, the possibility of bleeding and the formation of a spinal epidural hematoma (SEH) can lead to permanent neurological complications, especially if not diagnosed and treated in a timely manner.[1,2]

Thus, the aim of this case report is to address the particularities of anesthetic management in a patient with Hemophilia B and the challenges involved in formulating an anesthesia plan that ensures greater safety and minimizes risks during the procedures performed.

CASE REPORT

Patient M.G.S., male, 19 years old, 160 cm tall, weighing 62 kg. Diagnosed with Hemophilia B at 8 months of age, with hemophilic arthropathy in the left knee, previously undergoing synovectomy and arthroplasty of that joint (in 2018), and currently presenting with a pathological fracture of the distal femur and proximal tibia.

The patient was evaluated by the hematology team during the pre-anesthetic assessment, with the recommendation to administer factor VIIa at a dose of 6 mg every 4 hours, and every 3 hours on the day of surgery. In addition, the use of tranexamic acid was authorized, with a recommended dose of 500 mg every 6 hours. The use of red blood cell transfusion was also approved if necessary, using leukocyte-filtered and washed units. The use of nonsteroidal anti-inflammatory drugs (NSAIDs) was to be avoided due to their platelet inhibition effect.

The patient was then taken to the operating room, where multiparametric monitoring was established (ECG, SpO_2, Non-Invasive Blood Pressure – NIBP, capnography, Bispectral Index (BIS) cerebral monitor); prior central venous access was established in the right internal jugular vein; pre-oxygenation was performed with 100% O_2. Anesthesia induction was carried out intravenously with 120 mg of propofol, 20 micrograms of sufentanil, 25 mg of ketamine, and 100 mg of rocuronium. Orotracheal intubation (OTI) was performed using a 7.0 cuffed tube without complications, confirmed by capnography on the monitor. Anesthetic maintenance was done with 2% sevoflurane and remifentanil via target-controlled infusion (TCI). The patient was then positioned in the horizontal dorsal decubitus position, with thermal and ocular protection, the tube properly secured, and protection for joint and pressure points. During the procedure, administration of factor VIIa was carried out as recommended by the hematology service, along with 1 g of tranexamic acid and adjunct medications (4 mg dexamethasone, 2 g dipyrone). After approximately 60 minutes of surgery, the surgeon reported severe diffuse bleeding in the surgical field, difficult to control hemostasis, associated with visualization of severe fractures of the distal femur—supracondylar and intercondylar—and proximal tibia, showing signs of marked osteoporosis. Therefore, revision arthroplasty was chosen because surgical reduction with osteosynthesis was not feasible for the condition. The surgeon applied 3 g of Arista® hemostatic powder with good bleeding control. Additionally, prothrombin complex concentrate and two units of leukocyte-filtered packed red blood cells were administered. During the intraoperative period, the patient experienced sustained sinus tachycardia without

hemodynamic compromise, which improved after administration of 2 mg of metoprolol.

The procedure proceeded without major complications. Postoperative pain prevention was provided with 10 mg of morphine, remifentanil was discontinued, neuromuscular blockade was reversed with 200 mg of sugammadex, and the patient awoke and was extubated in the operating room without any adverse events. The patient was then transferred to the ICU for close monitoring and immediate postoperative care.

DISCUSSION

Hemophilic arthropathy is characterized by recurrent intra-articular bleeding, leading to the deposition of hemosiderin in the synovial tissues, which induces hypertrophy, neovascularization, and fibrosis of these tissues. These processes can cause inflammatory reactions and cartilage degeneration, which manifest as frequent pain, joint deformity, stiffness, and limited range of motion.[3]

Initially, treatment is based on conservative management. As the condition progresses, surgical procedures may become necessary, such as synovectomy (previously performed on the patient in this case), arthrodesis, and, finally, arthroplasty, as in the case presented.[3]

Since hemophilic patients have a higher risk of bleeding, it is recommended that their surgical procedures be scheduled for the first slot of the day, as well as at the beginning of the week, to ensure better support from the blood bank if needed. Anesthesiologists managing such cases should preferably have experience in handling hematological disorders.[3,4] Regarding anesthetic management, it is important to recognize that even airway manipulation can cause trauma with significant mucosal bleeding, as can more invasive interventions such as intubation, deep venous access, peripheral nerve blocks, and neuraxial blocks. To minimize these risks, measures are recommended, including lubrication of airway management equipment, the use of videolaryngoscopy to reduce the chance of trauma, avoiding nasotracheal intubation when possible, padding of joints at pressure points, and even avoiding the use of succinylcholine due to the theoretical risk of worsening intramuscular hematomas and hemarthrosis caused by the drug-induced fasciculations.[3,4]

Due to its low complication rate, regional anesthesia is an excellent option for patients requiring orthopedic procedures on the lower limbs, such as knee surgeries. This anesthetic modality reduces hospital stay duration and decreases the number of ICU admissions. However, a feared and potentially catastrophic complication when performing neuraxial anesthesia (subdural or epidural) in patients at increased risk of bleeding is spinal epidural hematoma (SEH), which must be diagnosed and treated within 8 to 12 hours of its formation. If not managed within this timeframe, it can lead to neurological complications due to spinal cord compression.[2] Despite this possible complication, SEH is a rare condition, with increased incidence in elderly patients, females, those using anticoagulants, and in the presence of vertebral canal diseases (such as stenosis, scoliosis, among others).[2]

Thus, some authors recommend general anesthesia for surgical procedures in these patients to reduce the risk of spinal epidural hematoma (SEH).[3] One of the key points in the preoperative management of these patients, aimed at minimizing postoperative complications such as infections and surgical failure, is the optimization of coagulation factor levels: maintaining factor IX levels between 60-80 IU/dL in patients with Hemophilia B undergoing major surgery, gradually

decreasing to approximately 50% until the surgical wound is fully healed. Administration is typically guided by clinical estimates of intraoperative blood loss, preferably kept below 25%.[3,4] These values may vary depending on the literature consulted, as there is no well-defined protocol yet for preoperative coagulation factor levels in this population.[3,4] Therefore, this remains a controversial topic, as demonstrated in a systematic review by Togioka et al., which showed low evidence supporting the need for factor IX levels above 50% for neuraxial anesthesia approaches.[5]

Another possible approach for these patients, both as prophylaxis and treatment for intraoperative bleeding, is the administration of recombinant activated factor VII (rFVIIa), even after the use of factor IX (in the case of Hemophilia B), as was done with the patient in the clinical case, under the guidance of the hematology team. This practice directly activates factor X, leading to thrombin formation independently of factors VIII and IX in hemophilic patients.[6] A review article by Vince and Brandão in 2009 showed that the use of rFVIIa began in the 1980s and started gaining popularity from 1999 onward, with promising results. However, despite many studies demonstrating a reduction in the need for blood transfusions in these patients, the lack of consensus in the literature regarding the ideal dosing regimen and the potential complications associated with rFVIIa use (such as thromboembolic effects) highlight the need for further research on the topic.[6]

Furthermore, Marrone et al.[7] reported a case in which a peripheral nerve block (PNB), specifically an adductor canal block and sacral erector spinae plane block, was performed for a total knee arthroplasty in a hemophilic patient, in order to avoid neuraxial blockade. This approach provided satisfactory analgesia during surgery and up to 72 hours postoperatively, with pain levels up to 4/10, using only periodic acetaminophen. Despite this relevant result, PNBs in patients with a high risk of bleeding should be considered as having a high probability of hemorrhagic complications and may require compression of the puncture site, similar to neuraxial approaches.[2]

CONCLUSION

Because they have a higher risk of hemorrhagic complications during anesthetic and surgical procedures, patients with a history of hemophilia require special attention regarding such adverse events, with important considerations aimed at reducing the risk of difficult-to-control bleeding and its possible catastrophic consequences. In this context, the complexity and diversity of anesthetic management options for these patients pose a significant challenge for the anesthesiologist, who must work closely with the hematology team to ensure the greatest perioperative benefit and safety, as was done in the patient in this clinical case.

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EDITORIAL AND REVIEW

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