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Case Report

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Successful Ultrasound-Guided Spinal Anesthesia in a Patient With Severe Hemophilia A Undergoing Total Hip Arthroplasty

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Abstract

Neuraxial anesthesia is the preferred technique for total joint arthroplasties. However, the absolute safety of neuraxial anesthesia in hemophilia patients has not been established. We describe a case of an adult male with severe hemophilia A, who presented for primary hip replacement due to severe hemophilic arthropathy and was managed with ultrasound-facilitated neuraxial anesthesia. Due to bleeding risks, additional considerations were necessary to minimize development of postoperative spinal hematoma. There were no perioperative adverse events. Careful preoperative multidisciplinary planning, perioperative management of neuraxial anesthesia (including the use of spinal ultrasound), and hemostasis were instrumental to successfully accomplish this. Following these principles, we demonstrate that neuraxial techniques may be a safe option for managing patients with severe hemophilia A.

Keywords: Hemophilia; Neuraxial; Spinal; Ultrasound; Anesthesia; Arthroplasty

Introduction

Hemophilia A is one of the most common serious congenital coagulation disorders resulting from a deficiency of factor VIII. Hemophilia A is an inherited X-linked recessive disorder that occurs in 1:5,000 male births with no geographic or racial predilection [1]. Patients often experience recurrent joint hemarthroses which can lead to the development of hemophilic arthropathy requiring early surgical intervention [2]. Providers must consider the risks with surgical and procedural sites that can result in morbidity and mortality, secondary to bleeding. As such, there may be hesitancy to utilize techniques that can be catastrophic because of bleeding tendency, such as neuraxial anesthesia. However, we present a case demonstrating success with this technique in a patient with severe hemophilia A, presenting for primary total hip arthroplasty.

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Case Report

Investigations

A 56-year-old male (body mass index (BMI) 23.3 kg/m^2) with a history of severe hemophilia A (baseline factor VIII levels < 0.01 IU/mL) and no history of inhibitor, suffering from severe hemophilic arthropathy of his right hip presented for primary joint arthroplasty. He was seen in the pre-admission clinic to discuss anesthetic management for his upcoming procedure. Given his history of hemophilia A, it was imperative to include his long-term hematologist and an orthopedic surgeon, who is experienced in hemophilia in regard to perioperative management of hemostasis and perform the major surgery in a setting with optimal coagulation laboratory and blood bank support. Other significant past medical history included prior hepatitis C infection (negative viral load), positive human immunodeficiency virus (HIV) status, and chronic pain secondary to hemophilic arthropathy involving multiple joints, particularly his right hip. Of note, his pain regimen consisted of gabapentin 300 mg daily and oxycodone 1,160 mg daily.

Diagnosis

The patient was presented with the options of general anesthesia versus neuraxial (spinal) anesthesia. In-depth conversations took place in the preadmission clinic regarding benefits of spinal anesthesia in the context of his chronic pain and opioid dependency. Despite limited epidemiological data surrounding risks of neuraxial anesthesia and hemophilia, specifically spinal hematoma, the patient preferred spinal anesthesia for his upcoming surgery. Once operative planning was complete, the patient's hematologist gave specific perioperative instructions

for maintenance of hemostasis by targeting a preoperative factor VIII level between 0.8 and 1.0 IU/mL immediately and to ensure safe factor VIII trough levels postoperatively, in accordance with the World Federation of Hemophilia guidelines [3]. While continuous infusion of coagulation factor VIII is preferred over bolus administrations for major surgeries, the specifically designated infusion pumps are not available at our institution.

Treatment

In the operating room, the patient was given recombinant factor VIII 40 - 50 U/kg and tranexamic acid 1 g IV push, exactly 30 min prior to skin puncture for the spinal anesthesia attempt. The patient was in the sitting position, and a midline approach was planned. Spinal ultrasound (Clarius Mobile Health Corp., BC, Canada) was utilized to identify the desired intervertebral space and to minimize spinal anesthesia attempts. Using the parasagittal oblique view, the L3-4 space was identified and marked. The interspace was then confirmed with the transverse view of the probe, which also identified the L-spine midline and assisted in the planned trajectory for spinal needle insertion. The ligamentum flavum was visualized approximately 4 cm below skin. Following aseptic preparation, a 25-G Whitacre needle with a 20-G introducer needle for stabilization was used. One attempt was required for dural puncture, and there was return of clear cerebrospinal fluid. A combination 14 mg of 0.5% isobaric bupivacaine and 10 µg fentanyl was injected in the intrathecal space. No blood was noted either on the needle tips or through the skin puncture upon withdrawal of the needles. Once the patient was positioned lateral, another 1 g of tranexamic acid IV was injected. Adequate anesthesia was confirmed with a forceps pinch-test, and the patient was then adequately sedated with propofol IV (40 - 50 µg/kg/min) and was administered oxygen via nasal prongs at 4 L/min for the duration of the surgery. At the conclusion of surgery, fibrin glue was applied from intracapsular to superficial sites, and a periarticular injection of a combination of 200 mg of 1% ropivacaine and hydromorphone 2 mg was administered. Blood loss during the surgery was average and estimated at 200 mL.

Follow-up and outcomes

The patient was given recombinant factor VIII 25 U/kg IV every 6 h (q12h) (for a total of five doses) starting 8 h after the initial preoperative dose. On postoperative days 3 - 7, factor VIII 25 U/kg IV was administered daily in the morning, then every other day on postoperative days 8 - 14. The patient was also instructed to continue tranexamic acid 2,000 mg orally (po), three times a day (tid) for a total of 10 days. Avoidance of aspirin and non-steroidal anti-inflammatory agents, as well as the use of mechanical thromboprophylaxis were implemented to mitigate the bleeding risk. In the morning of postoperative day 1, a factor VIII trough level was drawn and was found to be 0.87 U/mL (normal range 0.5 - 1.5 U/mL). A peak factor VIII level was drawn immediately after factor VIII administration and was found to be 1.64 U/mL. Twenty-four hours postoperatively, the patient showed complete block regression with resumption of lower extremity motor function and sensation. The patient continued with his postoperative rehabilitation regime in hospital and was ultimately discharged home on postoperative day 4 with no complications, including no immediate or delayed postoperative bleeding.

Discussion

This case highlights two important principles in managing patients with hemophilia undergoing total hip arthroplasty and receiving a neuraxial anesthetic: 1) Careful multidisciplinary preoperative planning with the patient's hematologist, surgeon, anesthesiologist, coagulation laboratory and blood bank should be utilized to reduce perioperative bleeding risk; 2) Emphasis on minimizing trauma using visualization techniques to reduce the risk of spinal hematoma. In this case, a small gauge pencil-point spinal needle and the assistance with spinal ultrasound helped minimize attempts for a successful spinal anesthetic. This strategy was used to mitigate the significant risk factors such as large needle gauge, multiple needle passes, and difficult needle placement for the development of spinal hematoma [4].

A literature review completed by Choi et al [5] demonstrated six articles related to hemophilia and neuraxial techniques; they comment that there is a paucity of published data regarding the provision and safety of neuraxial techniques in patients with hemophilia, and as such, evidence-based recommendations cannot be offered. A recent single-center retrospective study of 116 patients (54 of whom received neuraxial anesthesia) however suggests that neuraxial anesthesia can be as safe as general anesthesia in hemophilia patients undergoing joint arthroplasty, but general anesthesia should still be favored when length of surgical time is an issue [6]. A systematic review of neuraxial techniques in patients with hemophilia identified 13 articles (n = 134 patients), with a risk of neuraxial hematoma in three (2%) of the individuals, all of whom had no factor replacement prior to lumbar punctures, due to previously undiagnosed moderately severe hemophilia [7]. In our case, the patient's perioperative hemostasis regimen was guided by the World Federation of Hemophilia guidelines. Therefore, although standardized evidence-based recommendations do not exist, individualizing treatment for the patient based on previous experience may be warranted in these circumstances.

It is widely understood that individuals with increased bleeding risk from coagulopathies such as hemophilia are at an increased risk of complications from neuraxial anesthesia. Spinal hematomas require immediate surgical decompression to avoid permanent loss of neurological function secondary to cord compression. Due to this bleeding risk, there is uncertainty surrounding the usage of neuraxial anesthesia as an optimal anesthetic in patients undergoing joint arthroplasty [8]. However, evidence suggests that by using a factor VIII threshold of > 0.5 IU/mL to correct coagulation defects, neuraxial anesthesia can be safely used, at least in obstetrical populations [7]. A recent French observational study evaluating hemophilia carriers reported no spinal hematoma associated with the use

of neuraxial anesthesia, including in three women who underwent neuraxial anesthesia without hemostatic replacement for low factor VIII < 0.5 IU/mL [9]. However, one should be cautious when applying expert-based recommendations from the obstetric population to other surgical populations, due to a lower risk of spinal hematoma following neuraxial anesthesia in the former.

Previous meta-analyses [10-13] describe benefits of spinal anesthesia over general anesthesia for total hip arthroplasties, including lower incidence of deep venous thrombosis, intraoperative blood loss, likelihood of blood transfusion, hospital length of stay, and 30-day mortality. Importantly, neuraxial anesthesia is recognized as the preferred technique for primary unilateral hip replacement based on lower complication risk [14]. Additionally, neuraxial anesthesia is associated with decreased pain scores and usage of rescue opioids postoperatively [15, 16]. This aspect was beneficial in our case given the patient's chronic pain and high levels of opioid use for daily function. As such, spinal anesthesia was likely the best option for our patient's surgery.

Historically, patients with hemophilia are treated with on-demand factor concentrates and are at risk of developing end-stage arthropathy at an early age. Orthopedic interventions such as joint arthroplasties are often offered at a younger age than the general population. Significant advances in hemophilia care have reduced the annual rates of joint hemarthroses to near-zero, thereby reducing the risk of hemophilic arthropathy. US data from 130 hemophilia treatment centers and the Center for Disease Control and Prevention (CDC) demonstrated a 5.6% decrease in invasive orthopedic interventions in patients with hemophilia over an 11-year period (2000 - 2010) [17]. The decision to proceed with total joint arthroplasty in any patient is made when the probable benefits of the procedure outweigh the likely risks. Total joint replacement in patients with hemophilia involves a greater degree of joint destruction, deformity, and bone loss, as well as soft tissue contracture [18], which increases the complexity of the surgery. Postoperative rehabilitation is very important to achieve a good result and is more difficult in these patients [19]. This is further complicated by difficulty in controlling postoperative pain in patients who have higher preoperative pain medication requirements. Our patient was treated with large doses of opioids preoperatively and had severe joint destruction and soft tissue contracture. Given his preoperative pain and limited mobility from his right hip, our patient's likelihood of benefit outweighed the risk, and he decided to proceed with total hip arthroplasty. Using the methods described above as well as intraoperative fibrin sealant at the end of the procedure, his perioperative bleeding was within normal limits and his postoperative pain control allowed for early mobilization on the same day of surgery. The patient continued with his rehabilitation regime in hospital and was ultimately discharged home on postoperative day 4 with no complications. Prior to discharge, we discussed the patient's experiences with the anesthetic and the surgery, and although he had some difficulties with the rehabilitation process primarily due to challenging perioperative analgesia, he overall was satisfied with his perioperative management.

The successful outcome in our patient supports the idea that hemophilia is not an absolute contraindication to neuraxial anesthesia. Rather, this technique should be considered as a potential strategy in patients where general anesthesia is suboptimal for perioperative management or the presence of comorbidities favors a neuraxial technique. However, it is important to acknowledge that we are limited in this single case, and large controlled trials demonstrating widespread safety have not been conducted. As such, there are no formal criteria outlining when this approach is completely warranted, nor criteria for safety. Ultimately, neuraxial approaches for patients with hemophilia are not a typical recommendation. Therefore, the decision should only be made in appropriate cases after informed discussions between the patient and care team to consider the risks, benefits, patient preference, and comfort level of the anesthesiologist.

Learning points

Neuraxial (spinal) anesthesia is the preferred method of anesthesia management in patients undergoing elective total joint arthroplasties and can lead to decreased postoperative morbidity and mortality. The absolute safety of neuraxial anesthesia in hemophilia patients has not been established, in particular with potential adverse events of spinal hematoma resulting from neuraxial anesthesia. By using a multidisciplinary approach, including perioperative planning with the team consisting of the anesthesiologist, hematologist, and surgeon, these risks can be minimized. Specifically, with proper perioperative clotting factor management, surgical methods to reduce bleeding risk, and anesthesia utilizing methods to reduce trauma from neuraxial anesthesia (i.e., ultrasound), total joint arthroplasty in hemophilia patients can be safely managed with these principles described above.

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None to declare.

Conflict of Interest

The authors declare they have no conflict of interest.

Informed Consent

The patient gave written consent to publish the circumstances surrounding his surgery.

Author Contributions

All authors contributed to the writing and editing of the manuscript and approved the submitted version.

Data Availability

Any inquiries regarding supporting data availability of this study should be directed to the corresponding author.

Abbreviations

HIV: human immunodeficiency virus; IV: intravenous; CDC: Center for Disease Control and Prevention

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