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Case Report Open Access

Perioperative Management of Patient with Factor XI Deficiency Undergoing Total Knee Arthroplasty

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Abstract

Factor XI (FXI) deficiency is a rare, inherited bleeding disorder that is typically asymptomatic and presents with unexpected bleeding after surgery or trauma. We present a patient with FXI deficiency that underwent total knee arthroplasty (TKA) under general anesthesia and received peripheral nerve blocks for postoperative analgesia. We also present a review of literature focused on presentation of FXI deficiency in various surgical settings and choices of perioperative anesthetic management.

Keywords: Regional anesthesia; Neuraxial anesthesia; Factor XI deficiency

Introduction

Factor XI (FXI) deficiency is a rare, inherited bleeding disorder (also known as hemophilia C, plasma thromboplastin antecedent deficiency or Rosenthal syndrome) that is typically asymptomatic and initially presents with unexpected bleeding after surgery or trauma. In the general population, the prevalence is exceedingly rare (1:1,000,000), but in the Ashkenazi Jewish population, the reported prevalence varies in the literature [1-6]. Approximately 1:8 individuals are heterozygous and 1:190 are homozygous for a mutation on the F11 gene, the most common gene mutation that results in FXI deficiency in this population [2]. FXI levels <20 U/dL designate a severe deficiency, while levels between 60-80 U/dL and higher are generally accepted as normal FXI levels. Interestingly, FXI levels do not correlate with bleeding propensity and even in individuals with FXI levels <15-20, significant spontaneous hemorrhage is uncommon [2,5]. This is in contradistinction to other bleeding disorders in which lower clotting factor levels result in a proportionately increased risk of bleeding. Possible explanations for this discrepancy include variations in the level of FXI activity in platelets and the relative level of other clotting factors, particularly von Willebrand factor [2,3,5]. As a result, even those with partial or mild deficiency may be at increased risk of postoperative bleeding. A consensus regarding the safety of neuraxial and other regional techniques in patients with FXI deficiency has yet to be established. We present a patient with FXI deficiency that underwent total knee arthroplasty (TKA) under general anesthesia and received peripheral nerve blocks for postoperative analgesia. We also present a review of literature focused on presentation of FXI deficiency in various surgical settings and choices of perioperative anesthetic management. Consent was obtained from the patient for publication of this case.

Case Description

A 55 year-old Jewish, Caucasian male presented to Northwestern Memorial Hospital for a left total knee arthroplasty. He was seen in the preoperative clinic, which revealed a history of hyperlipidemia, arthritis, and FXI deficiency. The patient had been diagnosed with FXI deficiency approximately 30 years prior. His father had been diagnosed with FXI deficiency during a work-up for leukemia and subsequently, the patient and both his brother and sister were screened for a similar deficiency. The patient and his brother were both positive, while his sister was negative. At the time, he was told the deficiency was mild and did not require further follow-up or treatment. The patient denied a history of bleeding problems, including epistaxis or easy bruising. He had previously undergone a knee arthroscopy and extraction of wisdom teeth without any significant bleeding or need for blood transfusion. Preoperatively, the patient had normal basic coagulation studies (aPTT 29.5 sec, INR 1.0) and a FXI level of 40 U/dL, placing the patient in the mild or partial deficiency range. After discussion with the patient, a general anesthetic was performed. The patient preferred to avoid a neuraxial block as he was not willing to accept the risk of spinal hematoma, since the risk was not well established in this population. After confirming 2 units of crossmatched fresh flozen plasma (FFP) were available in the blood bank, the patient successfully underwent a left total knee arthroplasty without complications. Induction of general anesthesia was achieved with fentanyl 100 mcg IV, propofol 180 mg IV and rocuronium 50 mg IV. General anesthesia was maintained with sevoflurane, targeting a MAC of 1.0. Estimated intraoperative blood loss was 150 cc, with the use of a lower extremity tourniquet. Preoperative hemoglobin was 15.7 g/dL, immediately postoperatively was 14.6 g/dL, and a nadir of 11.4 g/dL for the entire hospitalization. No perioperative blood component transfusions or other hemostatic agents were necessary. Postoperatively, the acute pain service placed an ultrasound-guided femoral nerve catheter and performed a single-shot, infragluteal, parabiceps sciatic nerve block, both without complications. The sciatic nerve block was completed by performing a perineural injection of 30 ml of 0.5% ropivacaine with 1:300 K epinephrine, utilizing a 22 g×90 mm stimulating block needle. For the femoral nerve block, a 20 g femoral nerve catheter was placed through an 18 g insulated needle. Both ultrasound guidance and nerve stimulation were used, with strong quadriceps contraction disappearing below 0.5 mA with catheter stimulation. The catheter was advanced 5 cm beyond the tip of the need and after a negative test dose, 10 ml of 0.5% ropivacaine

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was administered through the femoral catheter. A continuous femoral nerve catheter infusion was started at a rate of 4 ml/hr with 0.1% ropivacaine. A patient-controlled demand function was programmed so the patient could receive an additional 5 ml of 0.1% ropivacaine with a 60 minute lock-out. On POD#0, after nerve blocks were performed, the mean VAS score was zero. On both POD#1 and POD#2, the mean VAS score was three. The acute pain service noted that analgesia was adequate at all inpatient follow-up evaluations. The patient was started on warfarin on POD#0 and on a dalteparin bridge on POD#1 for DVT prevention. The patient was closely monitored in the postoperative period. The acute pain service evaluated the patient on POD#1 and POD#2. The patient was evaluated for hematoma at the two nerve block sites. Also, a gross sensory and motor exam was conducted to ensure any deficits were consistent with nerve blockade. On POD#2, the femoral nerve catheter was removed in the early morning. In the afternoon of the same day, prior to patient discharge, the patient was again evaluated to ensure adequate recovery of sensory blockade in the femoral nerve distribution. No doses of dalteparin or warfarin were given on the morning of catheter removal. Platelet count was normal and INR was noted to be subtherapeutic on the morning of femoral catheter removal. The patient was discharged home on POD#2, which is consistent with the typical TKA patient at our facility.

Discussion

Factor XI plays an integral role in the intrinsic pathway of the coagulation cascade (Figure 1) [7]. Initial screening tests for FXI deficiency would reveal an isolated prolonged activated partial thromboplastin time (aPTT). A factor XI level can be obtained if there is a high level of suspicion for FXI deficiency and is also used as a confirmatory test. The presence of a FXI inhibitor, such as lupus anticoagulant, should be excluded. Bleeding in those with FXI deficiency is typically treated with FFP, although factor XI concentrates have been available since the 1980s [2,5]. When compared to the use of FFP, these concentrates have the advantage of smaller volumes, faster infusion times, and do not unnecessarily increase levels of other coagulation factors. FXI concentrates increase FXI levels by 1.8 percent for every unit/kg given, with a half-life of approximately 62 hours [4,8]. When used initially, though, the use of factor XI concentrates resulted in many thrombotic complications. Therefore, current guidelines recommend caution in the elderly and those with thrombotic risk factors [5]. Other therapies have also been investigated including: tranexamic acid, recombinant factor VIIa, and DDVAP. Limited data exists for these

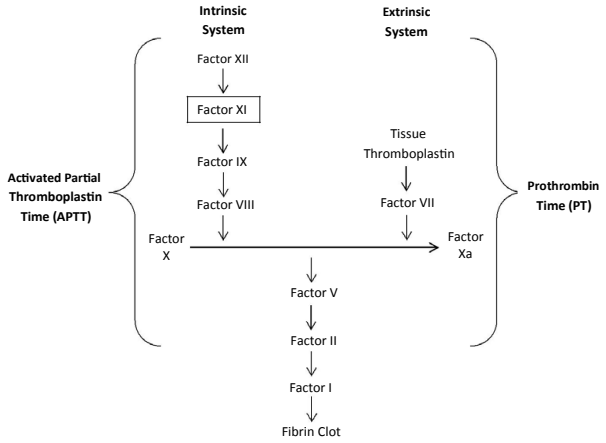

Figure 1: Simplified schematic of coagulation cascade.

therapies and none have direct effects on factor XI. Tranexamic acid has been shown to be useful in patients with mild deficiency undergoing minor procedures and has been studied in women with factor XI deficiency and menorrhagia [4]. Activated recombinant factor VIIa has been successfully used in patients with alloantibodies to factor XI and DDAVP has been studied as a prophylactic treatment for surgical bleeding in those with partial, symptomatic factor XI deficiency [9,10].

Although there are existing recommendations regarding the use of regional techniques in patients taking anticoagulant, antiplatelet, and thrombolytic medications, a consensus regarding the safety of neuraxial and other regional techniques in patients with FXI deficiency has yet to be established and limited data exists in the anesthesia literature [11]. The majority of data related to the safety of neuraxial techniques in FXI deficiency is found in the obstetrical anesthesia literature [12,13].

In a case report by David et al. three patients had peripartum epidurals placed (two for Cesarean section, one for labor analgesia), all without adverse outcome. This group recommends coagulation status is restored to normal prior to placing or removing an epidural catheter in the setting of FXI deficiency [7]. Additionally, a case series by Kadir et al. described two patients that had epidural anesthesia with no complications. This group states that if FXI levels are >50 U/dL during the third trimester and the coagulation screen is normal when the woman is in labor, then regional blocks should be made available to the patient [14]. Finally, a case series by Singh et al. published in Anesthesia and Analgesia in June 2009 analyzed the records of 13 patients with FXI deficiency who presented to labor and delivery between February 2000 and December 2007. Nine of these patients were managed with neuraxial techniques with no complications. Five of these patients were treated with FFP before anesthetic administration, while those that did not receive FFP had mild disease and no clinical bleeding history. This group concluded that it may be safe to administer neuraxial anesthesia to this patient population, especially if factor replacement is performed, but also emphasized the importance of a thorough risk/benefit discussion regarding the various anesthetic approaches [3]. A case report from Christiaens et al. describes a parturient with triplets and FXI deficiency who, despite preoperative transfusion of 8 units of FFP, was provided with a general anesthetic due to the potential risk of epidural hematoma [15].

No reports of spinal hematoma related to neuraxial techniques in those with FXI deficiency were found in the literature. Of note, in 1987, Mustafa et al. reported a case of spontaneous cervical epidural hematoma in a patient who subsequently developed Brown-Séquard syndrome and had a work-up revealing factor XI deficiency. The patient was Jewish, but had no history of bleeding problems, had an uncomplicated hysterectomy in the past, and no family history of bleeding disorders [16].

A thorough review of the literature showed an extremely limited number of studies or reports regarding the use of peripheral nerve blocks in patients with FXI deficiency. A case report published in the Journal of Clinical Anesthesia in May 2011 described an intraneural hematoma that occurred after nerve stimulation-guided femoral block in a patient with undiagnosed factor XI deficiency and a slightly prolonged aPTT (38.9 sec). Several days after surgery, the patient presented with severe paresis of the quadriceps femoris muscle and complete anesthesia to pinprick in the cutaneous distribution of the saphenous nerve. This prompted surgical exploration and removal of the intraneural hematoma. Rodriguez et al. attributed the delayed and progressive onset of neurological symptoms to the increasing anticoagulation produced by the prophylactic low molecular weight

heparin (LMWH) that was started on postoperative day POD#0 [17].

In cases of factor XI deficiency and active bleeding after surgery or trauma, monitoring factor XI levels may help clinicians deliver a more targeted approach to correcting this coagulopathy. In our case, we did not follow this value, since bleeding and the degree of postoperative anemia were similar to that of a non-coagulopathic patient. Coagulation laboratory values remained normal on POD#0 and POD#1, therefore, there was no indication to specifically monitor factor XI levels.

Considering the relatively high prevalence of factor XI deficiency in the Ashkenazi Jewish population and the relatively rare frequency with which we see this clinically, particularly in the perioperative period, it is likely that many cases go undiagnosed. It is important to emphasize that the level of factor XI deficiency does not correlate with bleeding risk, so even a patient with mild or partial deficiency can still have a significant risk of surgical bleeding or spinal hematoma with neuraxial techniques. There are no specific guidelines for anesthetic management in patients with factor XI deficiency, but FFP and factor XI concentrates can be given in cases of severe bleeding. No adverse outcomes are noted in the literature after performing neuraxial blocks in these patients, with only one adverse outcome in a patient with factor XI deficiency who received a femoral nerve block. Clearly, there is a paucity of data regarding the use of neuraxial and other regional techniques in patients with this deficiency and the current literature presents varying perioperative management recommendations. Further research will help guide perioperative management in these patients.

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