



Anesthetic management using peripheral nerve block in patients with factor XI deficiency: a case report

SCIENCE

[Case Studies](#)

A 75-year-old woman (46.2 kg, 153.7 cm tall, and American Society for Anesthesiologists physical status II) was scheduled for a partial duodenectomy. She provided written informed consent for participation and publication. Her previous surgical history included ovariectomy under general anesthesia at the age of 38 years, for which the surgical and anesthetic procedures were uneventful. She also had a history of hypertension, which was treated with angiotensin II receptor blockers. She had no other past medical history. At presentation, she was not taking any herbal preparations or anticoagulants. She had no family history of bleeding tendency.

Deficiency of which clotting factor results in difficulty in anesthetic management of patients?

- Factor I
- Factor XI
- Factor IX
- Factor V

Introduction

Factor XI is a plasma glycoprotein that participates in the intrinsic pathway of the blood coagulation cascade. Factor XI deficiency is a rare, autosomal recessive disorder present in 1: 1,000,000 individuals¹. In hemophilia A and B, the bleeding tendency is clearly related to factor level; however, a deficiency of factor XI results in unusual coagulopathy, with a poor correlation between the measured factor XI level and bleeding tendency². The unclear presentation of factor XI deficiency results in difficulties in anesthetic management of patients with this disorder. This report presents a patient with factor XI deficiency who underwent duodenectomy for a duodenal tubulovillous adenoma.

Medical History

The patient had a history of hypertension, treated with angiotensin II receptor blockers and had no other past medical history. At presentation, she was not taking any herbal preparations or anticoagulants. She had no family history of bleeding tendency.

Examination and Laboratory Investigations

Four months prior to surgery, she developed epigastric pain and an endoscopic examination revealed a duodenum tumor. She was scheduled for a partial duodenectomy. Laboratory findings on admission were as follows: normal electrocardiogram and chest X-ray, hemoglobin 13.4 g/dL, platelet count $21.5 \times 10^4 \mu\text{L}$, and coagulation tests showed prolonged activated partial thromboplastin time (APTT 89.7 s); however, the prothrombin time and international normalized ratio of prothrombin time were normal (PT 10.3 s, PT-INR 1.01). Hemophilia A and B are usually considered first, especially in males, whenever a prolonged APTT is noted; however, the patient had no family history of bleeding. Further investigation revealed severely deficient levels of factor XI, and factor XI deficiency was diagnosed. The levels of activity of her factors VIII, IX, XI, and XII are shown in Table 1.

Treatment

Ultrasound-guided subcostal transversus abdominis plane (TAP) block with rectus sheath block was



selected due to the potential risk of epidural hemorrhage. General anesthesia was induced by target-controlled infusion of propofol at a target plasma concentration of 3 µg/mL. Fifty milligrams of rocuronium was administered after loss of response to verbal commands. Remifentanyl was initiated at a rate of 0.3 µg/kg/min, and propofol was adjusted to maintain a bispectral index of 40–60. Subcostal TAP block with rectus sheath block was performed in the supine position. After skin preparation with 0.5 % chlorhexidine, the ultrasound probe was placed parallel to the subcostal margin near the xiphoid process, the needle was advanced to the TAP, and 20 mL of 0.25 % levobupivacaine was administered to the TAP. After subcostal TAP block, the rectus muscle was imaged with the ultrasound probe on the level of the umbilicus and the needle was advanced to the posterior rectus sheath, and 10 mL of levobupivacaine was administered.

The surgical procedure itself was uneventful, and recovery was satisfactory. The patient reported no pain at the surgical site after extubation. Postoperative supplemental analgesics were administered. The patients received intravenous flurbiprofen axetil and acetaminophen on the first postoperative day. After started oral intake, loxoprofen sodium was administered and reported no pain. After the surgery, thin, watery-pink exudate was seen from surgical drain, but on the seventh postoperative day the color of the fluid changed to slightly reddish consistency. Coagulation tests were performed every day after the surgery. The PT and PT-INR levels were normal throughout the perioperative period.

Discussion

Factor XI deficiency, which is relatively common among Ashkenazi Jews, is associated with injury-related bleeding³. This is sometimes called “hemophilia C,” is distinguished from hemophilia A and B by the absence of hemorrhage inside joints and muscles, and has equal incidence in individuals of either gender. The level of the deficiency does not determine the bleeding risk⁴, and bleeding tendency varies among individuals. Patients who have severe factor XI deficiency are at risk of massive hemorrhage; however, some do not have this tendency⁵.

Patients with factor XI deficiency need specific management during surgery. Previously, adverse surgical outcomes of patients with factor XI deficiency have been reported, including cerebral hemorrhage⁶ and spinal epidural hematoma⁷. Different surgical management strategies, each with varying degrees of risk, have been reported, including FFP, factor XI concentrates, and desmopressin⁸.

Four units of FFP were used prior to the surgical procedure. The estimated half-life of factor XI is 45 h, which is similar to standard FFP⁹ therefore, we judged FFP transfusion 1 day prior to surgery which would prevent intraoperative and acute phase massive hemorrhage. APTT levels improved from 89.7 to 36.2 s after FFP transfusion. The transition level of APTT after surgery was investigated. The level of APTT was gradually prolonged to 52.6 s by postoperative day 7. After surgery, the color of the exudate fluid from surgical drain had changed to slightly reddish, which may be related to the change of APTT. Considering the estimated half-life of factor XI, it was anticipated that APTT would rise to the same level as the preoperative value. Fortunately, APTT remained shorter than expected, and additional FFP was not used.

In addition to medication, safe and reliable anesthesia is required to manage perioperative hemorrhage. Epidural anesthesia is one of the most useful methods to alleviate surgical pain during the perioperative period. Although the incidence of neurologic dysfunction resulting from hemorrhagic complication associated with neuraxial blockade, including epidural anesthesia, is unknown¹⁰, the risk of bleeding is increased, especially in presence of coagulopathy. In recent years, peripheral nerve blocks using ultrasound have gained popularity. It serves as an effective alternative analgesia when an epidural is contraindicated or refused. Although guidelines for regional anesthesia in the patient receiving anticoagulants are available, regional anesthesia for patients with a clotting factor deficiency is dependent on the careful decision-making by the attending anesthesiologist. Neurologic dysfunctions after epidural anesthesia are very rare but devastating. Appropriate management for patients with clotting factor deficiency should result in an uneventful recovery.



Learning

Massive hemorrhage occurs unpredictably during perioperative period regardless of the bleeding tendency before surgery. FFP transfusion prior to surgery may help coagulation, and proper anaesthetic management, including peripheral nerve block, may contribute to improved prognosis.

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Exploratory, Propofol, Remifentanyl, Propofol, Factor XI deficiency, Blood, Local Anesthetics, Case report, Coagulation tests, Ultrasound-guided subcostal transversus abdominis plane (TAP), Activated partial thromboplastin time (APTT) levels