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Perioperative Management of a Patient with Factor XI Deficiency Undergoing Cardiac Surgery

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Abstract

Background: Factor XI deficiency (FXID), is a rare disorder of the coagulation system and the incidence of FXID is estimated to be one in a million. It is claimed to be associated with prominent bleeding in case of trauma and surgery irrelevant to the FXI level. The treatment modality of FXID varies from Fresh frozen plasma (FFP) to the plasma-derived FXI concentrate depending on country and medical center. In this case report, we present a patient with FXID who underwent coronary artery bypass grafting (CABG), and his perioperative management.

Case report: A 52-year-old, male patient was admitted to the cardiovascular surgery department and prepared for surgery. He was prepared in accordance with hematologist's recommendation and underwent CABG successfully. He was discharged from hospital on the seventh postoperative day without any complication.

Conclusion: The diagnosis of FXID is mainly based upon suspicion in cases of elevated aPTT or unusual bleeding after or during the surgery or trauma, as well as family history like in our case. Suspicion and proper history taking warned us to take precautions and manage the case successfully. In case of cardiovascular surgery of FXID patients, a team-based approach including a cardiologist, surgeon, intensivist and a hematologist with an on-site coagulation laboratory is essential.

Keywords

Perioperative Management, Factor XI deficiency, Cardiac Surgery

Introduction

Factor XI deficiency (FXID), is a rare disorder of the coagulation system and the incidence of FXID is estimated to be one in a million. It is claimed to be associated with prominent bleeding in case of trauma and surgery irrelevant to the FXI level [1]. The inheritance of FXID follows an autosomal recessive pattern -caused by changes (mutations) in the F11 gene- and affects both genders equally. Spontaneous bleeding is rare and may stop without treatment so uniform prophylaxis is not needed [2]. Unlike its siblings -Hemophilia A and B- FXID, is not well known and even in case of the severe illness, evidence-based guidelines are scant. Bleeding, especially from the mucosal surfaces during and after the surgery is the major apprehension in patients with FXID [3] and the risk is higher in case of cardiac surgery. Although FXI activity levels do not correspond directly to surgical bleeding, plasma levels < 30 IU/mL (the normal blood level of FXI is 70-120 U/dL) is generally felt to be associated with increased hemorrhage in an unpredictable fashion. The treatment modality of HC varies from fresh frozen plasma (FFP) to the plasma-derived FXI concentrate depending on country and medical center [4]. In this case report, we present a patient with FXID, underwent coronary artery bypass grafting (CABG), and his perioperative management.

Case Report

A 52-year-old, male patient was diagnosed with ischemic heart disease and admitted to the cardiovascular surgery department. The cardiac catheterization revealed

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coronary artery disease and he was prepared for surgery. A transthoracic echocardiogram (TTE) was done preoperatively and ejection fraction was found 60%. The left ventricular end-diastolic diameter was 4.8 cm without any abnormality.

His family history revealed FXI deficiency in his sisters. He had not undergone any surgical procedure, except tooth extraction and spontaneous bleeding or any other hematologic complication was not described. The hematologic evaluation showed a factor XI level of 30%, with an activated partial thromboplastin time (aPTT) of 38.3 seconds. The screening test for Lupus anticoagulant was negative. There was no bleeding complication during or after cardiac catheterization which was performed initially before surgery.

He was referred to the hematology department and in accordance with hematologist's recommendation, he was treated with FFP loading dose of 10 ml/kg. The infusion was tolerated well without volume overload. In the peri- and post-operative period he was treated with FFP infusion dose of 5 ml/kg. He underwent CABG successfully and transferred to the intensive care unit. During this period neither recombinant Factor VIIa nor oral aminocaproic acid was required. Posttransfusion coagulation studies were normal and his post-operative period was uneventful. He was discharged from hospital on the seventh postoperative day.

Discussion

FXID is the rarest group of hemophilia patients and it is stated that less than 30% of the FXI plasma level increases the tendency to bleed. Bleeding severity is not necessarily correlates with factor XI level and patients with mild deficiency may bleed severely. It is stated that bleeding history of the patient is more predictive of oncoming bleeding [1]. Rugeri, et al. [5] proposes the thrombin generation test which evaluates the overall tendency of a plasma sample to form thrombin after coagulation induction to predict bleeding risk without any relation to the FXI level.

Brunken, et al. [6] described the first cardiac surgery in an HC patient in 1984 and since then FXI concentrate, FFP, transexamic acid, factor VII, and desmopressin were used to prevent bleeding in these patients [3]. The management of a patient with HC with FFP was described by Hennaux and colleagues [7] for different surgical procedures. Instead of routine prophylaxis, replacement of FXI 2 days before the surgery and continued infusion 7 days after the surgery is recommended since FXI has a half-life of almost 50 hours [4]. Some countries utilize factor XI concentrates which carries thrombosis risk, whereas in some countries-like USA and Turkey- only FFP is used. It has to be remembered that FFP treatment may be complicated by volume overload and precautions must be taken [8]. In case of cardiovascular surgery of HC patients, a team-based approach including a cardiologist, surgeon, intensivist and a hematologist with an on-site coagulation laboratory is essential.

Conclusion

The diagnosis of FXID is mainly based upon suspicion in cases of elevated aPTT or unusual bleeding after or during the surgery or trauma, as well as family history like in our case. Suspicion and proper history taking warned us to take precautions and manage the case successfully.

Conflict of Interest Statement

All of the authors declare no conflict of interest concerning this manuscript.

Authorship Statement

All authors had full access to the data and participated in the data collection, design and writing of the manuscript. Each author has seen and approved the submitted version.

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Disclosures

None.

References

1. Wheeler AP, Gailani D (2016) Why Factor XI deficiency is a clinical concern. Expert Rev Hematol 9: 629-637.
2. Fitzsimons MG, Leaf RK, Mack J, et al. (2018) Perioperative management of a redo aortic root replacement in a patient with severe factor XI deficiency. Journal of Cardiac Surgery 33: 86-89.
3. Petroulaki A, Lazopoulos G, Chaniotaki F, et al. (2017) Factor XI deficiency and aortic valve replacement: Perioperative management. Asian Cardiovasc Thorac Ann 25: 450-452.
4. Bauduer F, de Raucourt E, Boyer-Neumann C, et al. (2015) Factor XI replacement for inherited factor XI deficiency in routine clinical practice: Results of the HEMOLEVEN prospective 3-year postmarketing study. Haemophilia 21: 481-489.
5. Rugeri L, Quélin F, Chatard B, et al. (2010) Thrombin generation in patients with factor XI deficiency and clinical bleeding risk. Haemophilia 16: 771-777.
6. Brunken R, Follette D, Wittig J (1984) Coronary artery bypass in hereditary factor XI deficiency. Ann Thorac Surg 38: 406-408.
7. Hennaux V, d'Otreppe S, Guillaumie B, et al. (2016) Successful major surgical procedures in a severe FXI deficient patient using fresh frozen plasma: A case report. J Hematol Transfus 4: 1042.
8. Leff JD, Zumberg MS, Widyn JG, et al. (2014) Hemophilia C in a patient undergoing cardiac surgery: Perioperative considerations. Semin Cardiothorac Vasc Anesth 18: 297-301.