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Case Report

Surgical management of the patient with factor V deficiency: a case report

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ABSTRACT

Factor V (FV) is a rare bleeding disorder that its incidence is one in a million. Patients with FV deficiency have typically mucosal bleeding and prolonged bleeding after an invasive procedure. There are no certain treatment modalities and there is no product of FV concentrates in the markets. The bleeding diathesis and protect the ovarian viability are a challenge as an opposite situation for treatment options and ovarian preservation after detorsion is controversial. We aimed to provide ovarian blood-stream, control bleeding and protect thromboembolism from the patient with all of our treatment in the light of literature and a few case reports.

Keywords: Factor V deficiency, Rare bleeding disorders, Gynaecological surgical management, Ovarian torsion

INTRODUCTION

Factor V (FV) is a glycoprotein and has an important role in the formation of the thrombinase complex that converts prothrombin into thrombin. FV deficiency is a rare bleeding disorder and its incidence is one in a million.[1] Clinical findings are especially mucosal bleeding and prolonged bleeding after invasive procedures typically.[2] It is diagnosed by prolonged coagulation parameters and the measurement of plasma FV activity. Although FV concentrates have started to be produced in recent years, fresh frozen plasma (FFP) is currently being used as experimental in treatment.[3]

Aim

We aimed to investigate the patient applied to our gynecologic emergency service with FV deficiency and ischemic torsioned ovarian cyst in the light of literature.

CASE REPORT

22 years old woman applied to our emergency service with rapidly onset groin pain. She was diagnosed as a genetical factor V deficiency disease before and had a history of 2 times prolonged bleeding after dilatation curettage for abortion. We performed transvaginally ultrasound (TV USG) and found; 8×6 centimeter (cm) mass in the left ovary. Its appearance resembled a hemorrhagic ovarian cyst and contained some solid areas within (Figure 1). We did not see blood-stream in the center and around in the mass by the tv doppler ultrasound. Free fluid was in the douglas pouch. She has tenderness and defense with the abdominal examination. She was hospitalized with a diagnose of hemorrhagic torsioned ovarian cyst and we decided to emergent operation. We performed the coagulation test and consulted to the hematology department before the operation. 2 units FFP and 3 ampules tranexamic acid in 500 cc saline solution were given to the patient according to the recommendation of the hematology department.

They advised continuing the replacement with FFP until International normalized ratio (INR) become under 1.5.

Figure 1: (A) preoperative USG image of the left ovary (B, C, D, E) images of the left ovary during operation (E) postoperative Doppler USG image of the left ovary.

Preoperative laboratory findings were hemoglobin (HB): 7.2 g/dL, prothrombin time (PT): 22.7 activated partial thromboplastin time (APTT): 57.6, INR: 1.9. The patient with stable hemodynamics was taken to the operating room after FFP and tranexamic acid replacement for bleeding prophylaxis. The anesthetist was informed about patient diseases and general anesthesia was applied to the patient. We performed incision of mini-laparotomy. The abdominal layers were passed respectively with bleeding control. 250 cc free hemorrhagic fluid in abdomen and 8 cm hemorrhagic cyst in the left ovary and 2 times twisted ovary were seen (Figure 1B). The left ovary was detorsioned. The cyst capsule was extracted and a cystectomy was performed. After 30 minutes of making the ovary detorsion, the ovary was reassessed (Figure 1C). Although 80 percent of the ovary was seen to bluish-black we preserved ovary for the reproduction status of the patient. Suturation was made to the cyst for control the bleeding on the cyst wall (Figure 1D/E). The abdomen was washed with 1000 cc of hot fluid. We put the silicone drain to the douglas pouch. Then, the abdominal layers were closed according to the anatomy, respectively, by making effective bleeding control. Transfusion of 2 units of erythrocyte was made to the patient during the operation. After 2 hours from the operation, coagulation tests were PT: 18,9 PTT: 44,3 INR: 1,58 and hemoglobin was 10,6 g/dL. 2 units FFP replacement was made the first day of the operation. Drainage was 100 cc serohemorrhagic from the drain. 3rd hours after the operation, 4000 mill units low molecular weight heparin (LMWH) was injected intramuscular (IM) to the patient for high risk of thrombosis after detorsion and maintain the ovarian viability and we continued the LMWH injection IM in the following days. In the postoperative 4th days, 4 units FFP replacement was made. Coagulation parameters were PT: 18.8, A PTT: 37.9, INR: 1.57 in the 3rd day of operation. We performed tv doppler USG in the 4th day of the operation and the bloodstream was seen in the left ovary (Figure-1F). There was no free fluid in the douglas pouch. The patient with a good general condition, stable vitality, and normal abdominal examination was discharged.

DISCUSSION

There are not enough data about the management of the patient with FV deficiency in gynecological operations except case reports. It is uncertain how long TDP replacement should continue after the operation. From a case report published in 2016, the patient was replaced by 5 units FFP before and 2-unit FFP after laparoscopy-assisted vaginal hysterectomy operation. The patient was followed up with 2 units FFP replacement 2 times in a day for 72 hours after the operation.[4]

FV deficiency can lead to life-threatening bleeding.[10] In our patient, the bleeding did not stop from the hemorrhagic cyst and torsioned has occurred in the left ovary. Therefore the need for operation arose.FFP and thrombocyte replacement was recommended in FV deficiency.[5,6] Peyvandi et al emphasized that there is no FV product in the markets and cryoprecipitate is ineffective, and the only effective replacement product is FFP in the Journal of the American Society of Hematology.[10] As stated before, since there is not enough research and data on this subject, FFP replacement and the tranexamic acid replacement was done before the operation; In the postoperative period, FFP was given for achieving bleeding control in the light of a few case reports and data in the literature and we closely followed the coagulation parameters. Although we did platelet preparation, we did not need it.

Studies are showing that preserving the ovary ensures that the blood supply returns even if no blood is seen after detortion in the torsional ovary.[8,9] In the systematic review of ovarian torsion in pediatric and adolescents, Dasgrupta et al suggested that gross ovarian appearance and the prediction about necrosis after detortion do not give information about ovarian function after the operation and should not be used as the sole determining factor for ovarian resection.[9] In a study by Oelsner et al when they made detorsion the ovary and followed the blue-black torsioned ovaries, they saw that ovarian functions returned and follicle development occurred.[8] Besides, in a study conducted by Kaya et al on rats, they detorted after making torsion in ovaries, followed one group without LMWH, and applied LMWH to the other group. As a result, the number of follicles was found

higher in the group where LMWH was applied and the antimullerian hormone was found less in the group without LMWH.[11] We also preserved the ovary in our patient and we saw the bloodstream in the ovary by Doppler USG in 3rd day of operation. It has been reported that low molecular weight heparin after detorsion supports ovarian blood supply.[7] Although both bleeding diathesis and ovarian viability for fertility are conditions that complicate the treatment options, we aimed to protect ovary and maximize the ovarian viability with the replacement of FFP and LMWH injection.

CONCLUSION

Consequently, bleeding disorders such as factor V deficiency are difficult for surgeons to manage. In this case, we tried to keep the amount of bleeding under control, while we wanted to protect ovarian blood supply due to young age and fertility desire. We used tranexamic acid and FFP replacement before the operation and FFP with LMWH after the operation. When we look at the current literature, there is a need for more publications on this subject since there are no definite recommendations on this issue. Our case can also be a resource in this regard.

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