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Case Report

Case report on perioperative management of combined factor V and VIII deficiency in a patient with prolapse uterus

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ABSTRACT

Combined factor V and factor VIII deficiency is a rare autosomal recessive disorder with incidence of one in 10,00,000. We reported a case of seventy-one-year-old patient with third degree uterovaginal prolapse, diagnosed as combined factor V and VIII deficiency and the successful perioperative management during vaginal hysterectomy. With our experience in this, we conclude that for the patient's with factor V and VIII deficiency that the correction should be given for at least 2 weeks post operatively.

Keywords: Factor V and VIII deficiency, Perioperative management, Vaginal hysterectomy

INTRODUCTION

Combined deficiency of factor V and VIII occurs due to the mutation in LMAN 1 or MCFD 2 and was first described in 1954.[1] Patients present with mild to moderate bleeding manifestations. The most common symptoms are gingival bleeding, easy bruising, menorrhagia or prolonged bleeding after tooth extraction or normal vaginal delivery. This disorder is prevalent in Mediterranean areas where the consanguineous marriages are more common and diagnosis is usually made by prolonged PTT levels and deficiency of factor V and VIII levels.

CASE REPORT

We reported a case of successful perioperative management of combined factor V and factor VIII deficiency during vaginal hysterectomy with pelvic floor repair. 71-year-old lady with third degree uterovaginal prolapse and factor V and factor VIII deficiency was admitted at Sri Ramachandra Medical Centre for vaginal hysterectomy with anterior and posterior colpoperineorrhaphy. Patient had no heavy menstrual bleeding and her postpartum periods were uneventful. Patient was a known case of uterovaginal prolapse for past twelve years and was treated conservatively for the past eleven years (as the patient was not willing for surgery) with vaginal pessary which was changed every three months. Pessary was neglected for period of one year during COVID pandemic in 2021 and was removed in December 2021. In February 2022, patient was planned for vaginal hysterectomy. During pre-operative evaluation, patient found to have elevated PT (23.6 seconds) and aPTT (57.4 seconds) levels following which evaluation done and diagnosed as combined factor V and VIII deficiency. On admission: Hb-13 g/dl, platelet-3.08 lakhs, INR-2.8, BT and CT-within normal limits, factor V-8.2%, factor-VIII-19.9% factor VII, IX, XI-within normal range. Preoperatively eight units of fresh frozen plasma and 150 mcg desmopressin nasal spray were given. PT-16, PTT-40.1, INR-1.3, factor V-25.5%, factor VIII-30.5%. As the target levels of factor V and factor VIII were not achieved, patient was given recombinant factor VIII injection 1500 units IV bolus and desmopressin nasal spray 300 ml. Intra operative period was uneventful. Blood loss was 500 ml and patient were given three units of FFPs and one unit of

packed cell. Post operatively, four units of FFPs daily for five days. On day 1, patient was given 100% correction (injection recombinant factor VIII -1500 units twice daily) followed by 70 % correction for 2 days (1000 units twice daily), followed by 50% correction for 3 days (500 units twice daily).

Patient was transfused with thirty-three units of FFPs and was discharged on sixth post-operative day. Patient was readmitted on tenth postoperative day, with complaints of bleeding per vaginum for three days, soaking one pad/day. On admission, factor V-8% and factor 8-23%. Patient was transfused with four units of FFPs per day for seven days along with injection recombinant factor VIII (70% correction for 2 days and 50% correction for 5 days).

Table 1: Laboratory investigation on admission.

Lab investigation	Patient value	Reference value
Hb	**13**	**11.9-15**
Platelets	**3.08**	**1.5-4**
aPTT	**57.8**	**20.4-38**

DISCUSSION

LMAN 1 gene is located on long arm of chromosome 18 and MCFD 2 located on short arm of chromosome 2. LMAN 1 and MCFD 2 forms calcium dependent stable complex which helps in efficient transportation of factor V and factor VIII from endoplasmic reticulum to Golgi apparatus. Most of the mutations are nonsense or frameshift mutations in LMAN1 giving rise to truncated proteins, while in MCFD-2, both missense and null mutations are noted. Patients with homozygous combined factor deficiency, patient may develop spontaneous or post-traumatic bleeding. Factor V is synthesized primarily in hepatocytes and mega karyocytes and is found in the plasma and α granules of platelets as a single chain polypeptide.[3] Recent study identified liver sinusoidal endothelial cell as a significant source of circulating Factor VIII.[4] For patients with combined factor V and VIII deficiency, fresh frozen plasma were used. Factor V extract was not available and hence factor V levels can be achieved only through FFP transfusion. FFP should be freshly transfused, as the factor V is labile protein in patients with moderate to severe bleeding, cryoprecipitates, factor VIII concentrates, desmopressin can be used. Factor V and VIII had the same domain structure s factor V (AI-A2-B-A3-CI-C2) and shared nearly 40% amino acid sequence homology in its A and C domains, this inhibitor molecule may be one molecule that recognizes and neutralizes both factor V and factor VIII.[6]

CONCLUSION

With our experience in this case, for the patients with factor V and factor VIII deficiency correction should be given for at least for 2 weeks post operatively.

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