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CASE REPORT

Recurrent Ruptured Hemorrhagic Corpus Luteal Cyst in a Known Case of Combined Deficiency of Factor V and VIII

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Introduction

Combined deficiency of factor V (FV) and FVIII (F5F8D) is a rare bleeding disorder. As per other rare autosomal recessive disorders, F5F8D is often associated with consanguinity [1]. F5F8D is suspected in bleeding patients with prolonged prothrombin time (PT) and activated partial thromboplastin time (APTT). Diagnosis is made on laboratory findings of decreased FV and FVIII levels in plasma, usually in the range of 5–30% of normal (typically 10–20%) [1]. Ultimate confirmation of F5F8D comes from the identification of mutations in either *LMAN1* or *MCFD2* which is done on a research basis in few medical centers [1]. Fortunately, clinical management of F5F8D does not rely on molecular diagnosis [1].

Symptoms of F5F8D are generally mild. Common spontaneous bleeding symptoms include epistaxis, gum bleeding, easy bruising, and menorrhagia. Less frequently reported are hemarthroses, gastrointestinal bleeding, hematuria, and intracranial bleeding. In males, bleeding due to circumcision is frequently reported in regions where it is commonly practiced. In females, menorrhagia is found in a majority of patients in all reported studies.

Case Report

We report a case of F5F8D with recurrent hemorrhagic corpus luteal cyst rupture without any history of menorrhagia. An eighteen-year-old unmarried girl who is the second daughter of non-consanguineous couple was referred to our hospital in view of severe pain in abdomen for

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4 days and was diagnosed as right-sided twisted ovarian cyst of 5 × 5 cm size and free fluid in the peritoneum. She had a history of irregular periods since menarche. She attained menarche at the age of 12 years and had irregular periods once in 2–3 months with no history of menorrhagia. She is a hypothyroid patient and is on treatment for last 2 years. She was diagnosed as F5F8D deficiency at the age of 6 years when investigated for easy bruising and excessive bleeding from gums. But she lost follow-up with hematologist afterward.

On presentation at our hospital, she had severe pain in abdomen which was not relieved with parenteral analgesics. There was no pallor and her vitals were stable. She had diffuse abdominal tenderness with guarding which was more on the right side. Factors V and VIII were only 24.8 and 15%, respectively. PT was 18.8 s (mean control—12 s), APTT was 70 s, and INR was 1.6. Thrombin time was 6 s, and plasma fibrinogen was 400 mg/dl. Bleeding time was 2 min and VWF was 127% (50–160%) and platelet count was 264,000 with Hb of 12.3 g% and TLC 8000/μl with normocytic normochromic peripheral smear. USG showed 6 × 5.7 cm heterogeneously mixed echogenic focal lesion at right adnexal region, localized posteriorly to right ovary with intrinsic vascularity and moderate fluid in POD. Left ovary was polycystic. MRI scan done outside showed a heterogenous T2 hypo- and T1 hyperintense nodular lesion in right ovarian tissue with peripheral follicles and mild fluid collection in POD, possibilities? Torsion? Endometrioma.

She was given 10 units of cryoprecipitate and 4 units of FFP in consultation with hematologist and anesthesia team. PT/APTT and INR were repeated subsequently which were 16 s, 35 s, and 1.1, respectively. She was taken up for laparoscopy in view of intractable pain without any relief with opioid analgesics.

On laparoscopy, uterus and left ovary and bilateral tubes were normal. Right-sided ruptured hemorrhagic ovarian cyst of 5 × 5 cm with hemorrhagic fluid of 100–200 ml was seen in the peritoneum. Cyst wall was excised and sent for HPE, which showed hemorrhagic corpus luteal cyst. Postoperatively, she was closely monitored in the ICU. On first postoperative day, repeat APTT was 55 s for which she was given 2 units of FFP. She was stable and discharged on day two. At discharge, she was advised to follow-up with hematologist and was advised to take oral contraceptive pills for regularizing periods and to suppress ovulation which helps in decreasing the risk of recurrent hemorrhagic corpus luteal cyst.

After 6 months, she came with same complaints and similar USG abdominal findings. She stopped OCPs after using for 3 months as the parents had a concern about using OCPs before marriage. Coagulation parameters were corrected, and laparoscopy was done for severe abdominal pain. Laparoscopy revealed similar findings as seen in the first episode. Biopsy revealed hemorrhagic corpus luteum cyst.

At discharge, she was given Inj Depot Provera 150 mg IM and advised to take oral contraceptive pills after 3 months regularly to avoid recurrent hemorrhagic corpus luteal cyst. She was advised to have regular follow-up with hematologist and was advised to come for preconceptional counseling. Since then, she is on OCPs for the last 2 years and is asymptomatic.

Discussion

Ovulation may rarely be complicated by rupture of the corpus luteum. In women with normal hemostatic function, this may be of little clinical consequence. More serious and even life-threatening bleeding episodes have been described in women with clotting disorders [2]. At the time of ovulation, when the ovum is extruded from the Graafian follicle, bleeding may occur into the peritoneal cavity. Following ovulation, the follicle develops into the corpus luteum and spontaneous bleeding may occur into the central cavity to form the corpus hemorrhagicum. If rupture of the latter occurs, then hemoperitoneum results. As blood irritates the peritoneum, this may lead to clinical symptoms. Bleeding is often minor but may be more serious in women with a bleeding disorder. Rupture is more likely, if abnormal follicle maturation occurs, such as if the follicle becomes cystic as the corpus luteum involutes. Bleeding occurs most frequently from the right ovary. The presence of the rectosigmoid colon is thought to help protect the left ovary from trauma [3].

F5F8D is a rare autosomal recessive bleeding disorder with mild clinical symptoms. Bleeding from trauma/surgery is the most frequently reported clinical manifestation. In females, menorrhagia is found in a majority of patients in all reported studies. In congenital coagulation factor deficiencies, the mainstay of treatment is to supplement factors and laparoscopy is reserved only in case of hemorrhagic shock [4]. In our case, in spite of correcting the clotting factors, patient had to go for laparoscopy in view of severe pain in abdomen rather than for shock. As the patient's parents were worried about the usage of hormonal pills in an unmarried girl, it had lead to the recurrence of ruptured hemorrhagic cyst problem. The prevention of recurrence is desirable to avoid life-threatening bleeds, to preserve fertility and to avoid the potential morbidity caused by loss of ovarian function with repeated surgeries. Hormonal suppression of ovulation with the use of the combined oral contraceptive pill has successfully prevented recurrence in a number of cases reported. The patient and attendants have to be counseled regarding the

need and safety of prolonged usage of contraceptive pills to suppress the ovulation and prevent recurrent problem [3].

Compliance with Ethical Standards

Conflict of interest None.

Informed Consent Informed consent was taken from the patient.

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