CASE REPORT

Haemoperitoneum due to ruptured ovarian cyst in a 13-year-old girl with factor V deficiency — A case report

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Abstract

Factor V deficiency is a rare autosomal recessive coagulation disorder. We are reporting a case of a 13 year old girl with factor V deficiency presenting as life threatening haemoperitoneum, following bleeding from ruptured ovarian cyst. Prolonged Prothrombin Time, Activated Partial Thromboplastin Time and a normal platelet count pointed towards a disorder of coagulation. Mixing study with factor V deficient plasma and coagulation factor assay revealed markedly reduced plasma factor V clotting activity.

Keywords: Hemoperitoneum, Ovarian cyst, Factor V deficiency.

Introduction

Bleeding disorders are known to treating physicians since the 16th century. Medical literature in the West is full of knowledge regarding different aspects of these disorders. Congenital bleeding disorders are found in all racial groups and have worldwide distribution but very limited information is available in developing countries like Pakistan about their prevalence.[1] Factor V deficiency is a rare bleeding disorder with a prevalence of 1 per million in the general population. It is inherited as an autosomal recessive trait. It was discovered in 1943 by a Norwegian haematologist Paul Owren while studying a woman affected by a haemophilia — like — syndrome. Only 150 cases of congenital factor V deficiency have been reported worldwide since 1943.[2]

We are presenting a case of haemoperitoneum due to ruptured ovarian cyst in a 13-year old girl with factor V deficiency.

Case Report

A 13 year old girl was admitted in Paediatric High Dependency Unit of The Indus Hospital, Karachi in January 2016 with abdominal pain from last 6 months which had increased in severity from the past few days. There was no history of vomiting, dysuria, constipation or loose stools. She had history of prolonged bleeding from minor cuts and wounds and was a known case of some undiagnosed bleeding disorder. She had received multiple blood transfusions every 2-3 months in the form of fresh frozen plasma and packed cells. CT abdomen and pelvis which was done in a private hospital two months back showed a large cystic lesion. She achieved menarche a year back, periods were regular associated with dysmenorrhea and menorrhagia. On clinical examination, she was a conscious, sick looking child with temperature of 36.5°C, pulse rate of 115 beats/minute, respiratory rate of 18 breaths /minute and blood pressure : 90/50 mmHg and oxygen saturation was 96%. She developed marked pallor within a short span of time. Abdominal examination revealed marked generalized tenderness. Examination of other systems was normal.

Her haemoglobin was 9g/Dl which dropped to 7g/Dl within few hours, TLC was 13.6x10^9/L, platelets were 377 x 10^9/mL. Coagulation screen showed both Prothrombin time and Activated Partial Thromboplastin Time of more than 2 minutes. The initial impression was that of ovarian torsion/haemorrhagic ovarian cyst/ruptured appendicitis with hypovolaemic shock. The initial management of the patient included resuscitation with I/V fluids, antibiotics and packed cells transfusion. C.T scan of abdomen and pelvis showed soft tissue density enhancing scrambled mass in pelvis which measured more than 8.2cm in cross section and 4.6cm AP diameter. Another adjoining lesion was seen anteriorly 6.4 x 4.6 cm, most likely originating from the ovary. There was also fluid seen in the cavity. Gynaecologist, haematologist and paediatric surgeon were taken on board. Alpha fetoprotein, CA 125 and B-HCG were sent, all were normal. Gynaecologists proposed the possible need of hysterectomy during laparotomy. There were a lot of conflicts regarding possible hysterectomy. Family was counseled about her situation and a written consent was taken by the Hospital's Ethical Board regarding possible hysterectomy. There was diagnostic dilemma for all the medical personnel indeed because she was to undergo a major surgery with no proper diagnosis of her bleeding disorder. An emergency laparotomy was carried out in which findings were 1000cc of haemoperitoeum, left ovarian capsule was found to be

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ruptured. Bilateral ovarian cysts, right was 5x5 cm and left was 4x4 cm. Histopathology showed no evidence of malignancy.

As both Prothrombin time and Activated Partial Thromboplastin Time were deranged during hospital stay and at multiple times in the previous records, factor levels were sent. Meanwhile, during and post surgery, child was given transfusions of Fresh Frozen Plasma. Three weeks after surgery, with factor V deficient plasma and coagulation factor assay revealed markedly reduced plasma factor V clotting activity. Factor V level was 0.1%. (normal range:50-150%). She did well after surgery. As there are no factor V concentrates available, patient was given Fresh Frozen Plasma post surgery. She was advised to be discharged on oral contraceptive pills by the gynaecologist to prevent her from recurrent bleeding episodes following ovulation. The patient is being followed up four weekly at The Indus Hospital.

Discussion

Haemoperitoneum is a serious and life threatening manifestation. Literature research shows two previously reported cases with similar presentation and age. A case of haemoperitoneum was described in a 13 year old girl with factor V deficiency. The patient presented with several episodes of ovulation — related bleeding which required hospitalization and fresh frozen plasma transfusion. She had to undergo surgery and a left oophorectomy was carried out. After this last episode, she was also placed on oral contraceptives which were very effective in preventing further recurrences. The patient tolerated the medication very well which in addition, were able to control menometrorrhagia with consequent decrease over time in transfusion needs.[3]

OC are the treatment of choice in congenital bleeding disorders to control both the menorrhagia and more importantly, ovulation-related haemoperitoneum. The second case also had a similar find like our case in which a 13-year old girl with factor V deficiency presented with acute haemorrhage. The ultrasound appearance of a large loculated cystic mass was consistent with substantial intraperitoneal bleeding. This patient was managed too by doing stabilisation with blood products followed by GnRH therapy. A study conducted at National Institute of Blood Diseases screened a total of 376 patients. In all 13 (4.0%) patients were with rare bleeding disorders. Factor V deficiency was detected in 1 (0.26%).[4]

The choice of dosages and modalities of treatment of bleeding disorders is based on the type of bleeding and on factor V levels. Replacement therapy with factor V can be administered only through fresh frozen plasma (FFP), because factor V concentrates are unavailable and factor V is not present in cryoprecipitate. Factor V levels should be raised to at least 15 IU/dl by using FFP daily in a loading dose of 15 to 20 ml/kg and then 3-6 ml/kg during a bleeding episode which corrects the deficiency temporarily.[5] The safe level of factor V for adequate surgical haemostasis is 25% of the activity of factor V in the normal control plasma. Postoperatively, FFP should be administered for 3-10 days with careful observation of wound healing.[6]

Conclusion

Rare bleeding disorders present with life threatening conditions as in this case haemoperitoneum with factor V deficiency. Such bleeding disorders are not being properly worked up and reported in our region as they tend to be present in higher rates due to consanguineous marriages.

Consent: Informed consent has been already taken by the parents for this case to be reported.

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