ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 04 | April - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

LIFE-THREATENING HAEMOPERITONEUM SECONDARY TO RUPTURE OF SIMPLE OVARIAN CYST.

Obstetrics & Gynaecology

Dr. Mansi Jain* Junior Resident, Government Medical College, Nagpur *Corresponding Author
Dr. Kirti Rachwani Assistant Professor, Government Medical College, Nagpur

ABSTRACT

Bleeding into the corpus luteum following ovulation rarely has clinical significance in healthy women, but may lead to life-threatening hemorrhage in women with congenital or acquired bleeding disorders.We present to you a case of 17 yrs old female who came to casualty with acute abdomen with massive hemoperitoneum with b/l haemorrhagic ovarian cysts managed by surgical management.Congenital disorders of coagulation are uncommon but present significant challenges in the perioperative period. Hence should always be kept in mind.

KEYWORDS

haemorrhagic ovarian cyst, hemoperitoneum, bleeding disorder

INTRODUCTION

Von Willebrand's Disease is most common of all inherited bleeding disorders. The prevalence of VWD is one in 100 but is asymptomatic in the majority of patients and is clinically significant in only one in 10000 patients.

VWD is classified by the International Society of Thrombosis and Haemostasis (ISTH) based on quantitative or qualitative defect of VWF into three types. In type 1 VWD, there is a quantitative defect, whereas in type 2 VWD, there is a qualitative defect. In type 3 VWD, there is a complete absence of VWF. Type 1 VWD is a mild-moderate bleeding disorder, whereas type 3 VWD is a severe bleeding disorder. VWD shows autosomal inheritance (dominant or recessive) depending on the subtype. Often the only abnormality is a prolonged bleeding time as normal screening tests do not rule out VWD. Deficiency of VWF leads to easy bruising from trivial trauma;bleeding from mucosal surfaces, epistaxis, gums, and bowel. Depending upon the type of VWD, patients can have prolonged bleeding after surgery.[1]

Case Study:

A 17 years old female, a known case of Von Willebrand's disease type 3 diagnosed in childhood presented to casualty with complaints of nausea, vomiting, pain abdomen since 2 days and distention of abdomen since 1 day. Patient had past history of episodes of prolonged menstrual bleeding that needed treatment. No other known comorbidities. No history of past surgical interventions. On examination she was conscious, oriented, afebrile, pulse 120/min, BP 90/60 mm of Hg, spO2 99% on air. Initial investigations revealed Hb 5.4gm%, TLC 8900/cu mm, platelets 147000/cu mm, INR 1.45, KFT was within normal limits.Ultrasound of abdomen-pelvis revealed masssive Hemoperitoneum with ?ruptured right ovarian cyst. Patient received 1U PRC , 1U cryoprecipitate, 1 U FFP and INJ tranexamic acid 1 gm iv preoperatively and was taken for emergency exploratory laparotomy due to deteriorating hemodynamics.. Surgery conducted under general anaesthesia. Hemoperitonium collection of approx. 1.2 litres and blood clots approx. 1.1 litres were removed intraop.

Additional bleed from surgery was 400ml due to continuous oozing from surgical site. A ruptured simple right ovarian cyst was identified and the haemorrhage was successfully stopped by diathermy. Cystectomy done.B/l tubes were edematous. Another tubo-ovarian mass of 7 x 7 cm was noted on the left side which drained cheesy material.Cyst wall separated and sent for HPE. Adhesions present in the pouch of Douglas.Evidence of pelvic inflammatory disease noted.VWF/FVIII concentrates for transfusion were unavailable and there was limited availability of cryoprecipitate in the blood bank. Patient was transfused 2U PRC, 2U cryoprecipitate, 4U platelets, and 2U FFP intra- operatively in addition to the crystalloids to compensate for ongoing losses and to maintain adequate perfusion and stable vitals throughout the procedure. Hemostasis achieved and abdomen was closed with bilateral drains in situ. Surgery was uneventful. Post operatively patient was monitored in ICU.Patient was transfused 2U cryoprecipitate additionally once they were available.Post op period was uneventful with No thromboembolic events. Patient discharged on day 10.

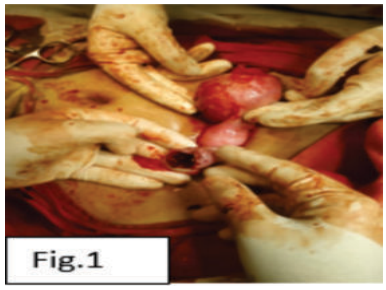

Fig.1 Showing the intraoperative image of right sided ruptured ovarian cyst and left sided tubo-ovarian mass.

DISCUSSION

VON WILLEBRAND'S DISEASE (vWD) is an autosomal inherited disorder resulting from a quantitative or a qualitative defect of von Willebrand factor (vWF) which is involved in primary hemostasis (adhesion of platelets to sub-endothelium and platelet aggregation) and acts as the carrier of coagulation factor VIII [2,3]. Expected plasma levels of vWF range from 50 to 200 IU/dL. The ABO blood type is the genetic factor that most significantly influences vWF levels. THE ABH antigens expressed on the N-glycosylated chains of the vWF influence its clearance and proteolysis [4], thus group O subjects have lower vWF levels of about 25 IU/dL due to accelerated clearance and increased susceptibility to proteolysis by ADAMTS13 with an estimated lower limit value of 40 IU/dL. This rate also fluctuates during the menstrual cycle with a lower rate observed during menstruation[5]. During pregnancy, it gradually increases from the 11th week of amenorrhea until it reaches 3 times the baseline rate by the end of the 3rd trimester.

Taking an estrogen pill raises this rate more moderately.These variations explain the fluctuations in the rate of vWF in a subject and the possible difficulty of making a diagnosis of vWD, so repeat testing and consultation with a hematologist regarding additional tests may be necessary for patients with strong personal and family histories of menorrhagia.[6]

Von Willebrand disease is responsible for symptoms such as mucocutaneous bleeding,excessive bleeding after trauma or invasive procedures, gastro-intestinal bleeding or hemarthrosis in the most severe forms of the disease.

In these cases of difficult diagnosis, the clinical history of a hereditary bleeding disorder which affects both sexes and is transmitted as a Mendelian dominant immediately suggests von Willebrand's disease. It is important, however, to remember that a negative family history, as in haemophilia, does not exclude the diagnosis [7].

Diagnosis relies primarily on clinical symptoms but requires the use of several laboratory analyses: von Willebrand factor activity and antigen testing and factor VIII activity. The ristocetin cofactor assay of von Willebrand's factor (vWF) function may be the best single screening test for von Willebrand's disease [8]. More specialized assays allow classification of the disease in various types and subtypes which imply

different management strategies (Table 1) . It should be determined which type of von Willebrand's disease a particular patient has because treatment depends on type.

Table 1 Different types of vWD disease

Type	Description
1	Partial quantitative deficiency of vWF
2	Qualitative defects of vWF
2A	Decreased vWF-dependent platelet adhesion and absence of vWF polymers
2B	Increased binding affinity of vWF for platelets
2M	Decreased vWF-dependent platelet adhesion; presence of vWF polymers
2N	Markedly decreased binding affinity of vWF for factor VIII
3	Virtually complete deficiency of vWF

The most important clinical manifestations mainly affect women. Although autosomal transmission predicts a similar prevalence of Willebrand disease in both sexes, menstruation, pregnancy and childbirth contribute to a higher prevalence of clinical manifestations of the disease in women, in the order of 60%.

Menorrhagia is common . Women with vWD's disease often complain about bleeding disorders with significant negative impact on the quality of life. There is an increased likelihood of developing endometriosis, bleeding and pain with ovulation, and sometimes haemoperitoneum [9]. Severe menorrhagia and life-threatening postpartum haemorrhage are frequent in the families of sufferers from von Willebrand's disease.

The menstrual cycle is characterized by midcycle ovarian follicular rupture. Ovulation is not normally accompanied by any significant bleeding, but women who have a congenital bleeding disorder such as Willebrand disease, a bleeding potential exists into the peritoneal cavity or into the residual follicle, resulting in a hemorrhagic ovarian cyst or retroperitoneal hematoma.

Acute treatment of hemorrhagic ovarian cysts can be based on surgical therapy, tranexamic acid and substitution with coagulation factor VIII or recombinant vWF concentrate. Treatment options include also oral contraceptive drugs, desmopressin acetate, antifibrinolytic agents, and plasma-derived concentrates rich in the high- molecular-weight multimers of vWF. Consultation with or referral to a haematologists is frequently helpful to assist in the management of patients with severe disease. Oral contraceptive therapy has been successfully used by gynecologists as first-line therapy for the management of von Willebrand's disease for many years. Oral contraceptives have been reported to be successful in the management of menorrhagia associated with von Willebrand's disease in 88% of patients [10]. Hormonally induced therapeutic amenorrhea may be appropriate for patients with severe disease.

Desmopressin acetate, which is available in parenteral form for intravenous use and in a highly concentrated intranasal spray formulation, is the treatment of choice for classic type I disease. It can be used as home therapy before the onset of menses or only during menses [8].

Allo-antibody formation occurs in 10-15% of type III VWD. In patients with inhibitors, there is very limited experience reported in the literature but recombinant VIIa (rVlla) and continuous infusion of high doses of rFVIll have been successfully used. Platelet infusions should be considered if bleeding persists, despite replacement VWF concentrates. Cryoprecipitate which was used in this case due to unavailability of VWF/factor VIII concentrates has unpredictable effect and risk of transmissible infections; should be used for management of VWD only when other treatment modalities have failed or unavailable.[9]

The major interest of a conservative treatment based on medical treatment and sparing surgery is to allow subsequent fertility and avoid repetitive surgical interventions, given that the symptomatology is recurrent.

CONCLUSION

Perioperative management of bleeding disorders require multidisciplinary approach comprising haematologist, surgeon, anaesthetist and laboratory services to ensure that appropriate factor concentrates are available and in sufficient quantity which is not always possible in resource constrained setups. Due to multiple limiting factors like unavailable preop investigations, inadequate transfusion resources and emergent nature of the case there was watchful digression from the standard first line of management. Surgery must always be preceded by a conditioning and drug preparation beforehand because hemostasis cannot be only surgical and the medical component proves to be primordial and decisive.Patient was managed successfully and uneventfully using combination of second and
third line of treatment protocol.

REFERENCES

1. Zimmerman, T.S. (1982) Von Willebrand's Disease. Progress in Hemostasis and Thrombosis, 6, 203.
2. Rodeghiero, F., Castaman, G. and Dini, E. (1987) Epidemiological Investigation of the Prevalence of von Willebrand's Disease. Blood, 69, 454-459.
3. Calmette, L. and Clauser, S. (2018) Von Willebrand Disease. La Revue de Médecine Interne, 39, 918-924.
4. Casari, C., Lenting, P.J., Wohner, N., Christophe, O.D. and Denis, C.V. (2013) Clearance of von Willebrand Factor. Journal of Thrombosis and Haemostasis, 11, 202-211.
5. Miller, C.H., Dilley, A.B., Drews, C., Richardson, L. and Evatt, B. (2002) Changes in von Willebrand Factor and Factor VIII Levels during the Menstrual Cycle. Throm- bosis and Haemostasis, 87, 1082-1083.
6. David, J.L., Gaspard, U.J., Gillain, D., Raskinet, R. and Lepot, M.R. (1990) Hemostasis Profile in Women Taking Low-Dose Oral Contraceptives. American Journal of Obstetrics & Gynecology, 163, 420-423.
7. Biggs, R. and Matthews, J.M. (1963) The Treatment of Haemorrhage in von Willebrand's Disease and the Blood Level of Factor VIII (AHG). British Journal of Haematology, 9, 203-214.
8. Phillips, M.D. and Santhouse, A. (1998) von Willebrand Disease: Recent Advances in Pathophysiology and Treatment. The American Journal of the Medical Sciences, 316, 77-86.
9. Dkeidek, A., Mattinson, R. and Grover, S.R. (2021) Bleeding Disorders: Are Gynaecologists Testing Women at Risk? Haemophilia, 27, e534-e536.
10. Nafidi, M., Khald, A., Aboulfalah, A. and Soummani, A. (2022) Recurrent Cyst within Willebrand Disease: Place of Surgery? Open Access Library Journal, 9: e8731.