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Haemorrhagic Ovarian Cyst with Hemoperitoneum in a Young Woman with Type 3 Von Willebrand Disease: A Case Report

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Abstract: ***Background**: Von Willebrand disease (VWD) is the most common inherited bleeding disorder. Type 3 VWD, characterized by a near-complete deficiency of von Willebrand factor, is the most severe form and is associated with spontaneous and potentially life-threatening bleeding episodes. Haemorrhagic ovarian cyst leading to hemoperitoneum is an uncommon but important gynaecological manifestation. **Case Presentation**: A 23-year-old nulligravida woman with known Type 3 VWD presented with severe lower abdominal pain, dizziness, and symptomatic anaemia. She had a previous history of hemoperitoneum requiring laparoscopic intervention. On evaluation, a significant decline in haemoglobin level from 11.5 g/dL to 5.7 g/dL was noted over a two-week period. Ultrasonography revealed moderate hemoperitoneum, a right adnexal organized hematoma measuring approximately 100–120 cc, and a left haemorrhagic ovarian cyst. Despite hemodynamic compromise at presentation, she was successfully managed conservatively with blood component therapy, von Willebrand factor concentrate, tranexamic acid, and close monitoring. Ovulation suppression with combined oral contraceptive pills was initiated to prevent recurrence. **Conclusion**: This case highlights haemorrhagic ovarian cyst with hemoperitoneum as a rare but potentially life-threatening manifestation of severe VWD. Early diagnosis, multidisciplinary management, and preventive ovulation suppression can reduce morbidity and avoid unnecessary surgical intervention.*

Keywords: Von Willebrand disease Type 3, hemoperitoneum, haemorrhagic ovarian cyst, inherited bleeding disorder, ovulation suppression

1. Introduction

Von Willebrand disease (VWD) is an inherited bleeding disorder resulting from quantitative or qualitative abnormalities of von Willebrand factor (vWF), a glycoprotein essential for platelet adhesion and stabilization of coagulation factor VIII. Type 3 VWD is the rarest and most severe form, characterized by an almost complete absence of circulating vWF and markedly reduced factor VIII levels.

Women with VWD commonly present with heavy menstrual bleeding; however, ovulation-related bleeding manifestations are less frequently recognized. Haemorrhagic ovarian cysts and corpus luteum rupture may result in significant intraperitoneal bleeding, particularly in patients with severe coagulation defects. Prompt recognition is crucial, as delayed diagnosis may lead to life-threatening haemorrhage and unnecessary surgical intervention.

We report a case of recurrent hemoperitoneum secondary to haemorrhagic ovarian cyst in a young woman with Type 3 VWD that was successfully managed conservatively.

2. Case Presentation

A 23-year-old unmarried nulligravida woman with a known diagnosis of Type 3 Von Willebrand disease presented with severe lower abdominal pain for three days, associated with nausea, dizziness, and generalized weakness. There was no history of trauma, fever, bowel disturbances, or urinary complaints.

She reported regular menstrual cycles accompanied by heavy menstrual bleeding and dysmenorrhea. Her last menstrual period was on 15 January 2026.

Her medical history was significant for a previous episode of hemoperitoneum in November 2024 that required laparoscopic intervention. Prior to referral, her haemoglobin level had declined from 11.5 g/dL to 5.7 g/dL over approximately two weeks. She had received transfusion support consisting of four units of fresh frozen plasma, six units of cryoprecipitate, and two units of packed red blood cells.

On admission, she appeared pale and clinically unstable, with tachycardia (110 beats/min) and hypotension (90/60 mmHg). Abdominal examination revealed lower abdominal tenderness without signs of peritonitis.

Laboratory investigations demonstrated:

- Haemoglobin: 7.8 g/dL
- Platelet count: $2.26 \times 10^5/mm^3$
- PT/INR: 15.17 seconds / 1.12
- Fibrinogen: 267 mg/dL
- Von Willebrand factor activity: 1 IU/dl

Pelvic ultrasonography demonstrated moderate hemoperitoneum with internal echoes suggestive of blood products. A right adnexal organized hematoma measuring approximately 100–120 cc was noted, likely representing residual blood from a previous bleeding episode. A left-sided haemorrhagic ovarian cyst measuring 3 × 1.6 cm was also identified. In the context of spontaneous hemoperitoneum and severe Type 3 von Willebrand disease, the haemorrhagic ovarian cyst was considered the most likely source of bleeding.

Given the patient's known bleeding disorder and absence of active ongoing hemodynamic deterioration, conservative management was done. She received von Willebrand factor concentrate (1000 IU daily for two days), tranexamic acid 1 g three times daily, blood component support as required, and close clinical monitoring.

The patient's symptoms improved progressively over a period of 7 days, without further decline in haemoglobin levels or evidence of ongoing intra-abdominal bleeding. Surgical intervention was avoided. Final haemoglobin on discharge was 10.4g/dl and improvement was seen in vwf factor activity was 120IU/dl

Repeat ultrasonography after one week revealed significant resolution of the hemoperitoneum, with no appreciable change in the size of the hemorrhagic ovarian cyst.

Prior to discharge, combined oral contraceptive therapy (Mala-N) was initiated for ovulation suppression and prevention of recurrent haemorrhagic ovarian cyst formation. She was advised for weekly follow up for VWF assessment and USG. The patient received counselling regarding the lifelong implications of VWD, recurrence risk, and future reproductive considerations including pregnancy-related bleeding risks.

3. Discussion

Hemoperitoneum secondary to haemorrhagic ovarian cyst is an important gynaecological emergency that may mimic other acute abdominal conditions such as ectopic pregnancy, adnexal torsion, appendicitis, or ruptured endometrioma. In healthy women, ovulation-related bleeding is usually self-limiting; however, in patients with underlying coagulation disorders, physiological follicular rupture may result in significant intra-abdominal haemorrhage requiring urgent medical intervention. Although haemorrhagic ovarian cysts are relatively common during the reproductive years, massive hemoperitoneum associated with inherited bleeding disorders remains a rare clinical entity.

Von Willebrand disease (VWD) is the most common inherited bleeding disorder and results from quantitative or qualitative defects of von Willebrand factor (VWF), a multimeric glycoprotein that plays a crucial role in platelet adhesion and stabilization of factor VIII. Type 3 VWD is the rarest and most severe form, characterized by near-complete absence of VWF and markedly reduced factor VIII levels. Consequently, affected individuals experience severe mucocutaneous bleeding, menorrhagia, postoperative haemorrhage, and deep tissue bleeding. Gynaecological manifestations are particularly common and significantly impact quality of life and reproductive health.

Heavy menstrual bleeding is the most frequently reported gynaecological symptom in women with VWD and may represent the first clinical manifestation of the disease. Other important manifestations include ovulation-related bleeding, haemorrhagic ovarian cysts, recurrent corpus luteum haemorrhage, endometriosis-associated bleeding, and postpartum haemorrhage. The prevalence of haemorrhagic ovarian cysts in women with inherited bleeding disorders has been reported to be considerably higher than in the general population, emphasizing the role of defective haemostasis in the pathogenesis of these complications.

Ovulation-associated hemoperitoneum is an underrecognized consequence of severe inherited bleeding disorders. During normal ovulation, rupture of the dominant follicle is accompanied by a small degree of physiological bleeding. In women with severe coagulation defects, however, this bleeding may continue unchecked, resulting in formation of a haemorrhagic ovarian cyst or massive intraperitoneal bleeding. Several case reports have described recurrent corpus luteum haemorrhage and spontaneous hemoperitoneum in women with Type 3 VWD, highlighting the need for a high index of suspicion when a reproductive-age woman presents with acute abdominal pain and unexplained hemoperitoneum.

The diagnosis of haemorrhagic ovarian cyst with hemoperitoneum relies on a combination of clinical, laboratory, and imaging findings. Patients typically present with sudden-onset lower abdominal pain, abdominal distension, nausea, vomiting, and varying degrees of hemodynamic compromise depending on the extent of blood loss. Ultrasonography remains the first-line imaging modality and commonly demonstrates a complex ovarian cyst with free intraperitoneal fluid. In our patient, imaging findings together with the known diagnosis of Type 3 VWD supported the diagnosis of a ruptured haemorrhagic ovarian cyst with hemoperitoneum.

Management of hemoperitoneum in patients with VWD differs significantly from that in women without bleeding disorders. The primary goal is stabilization of the patient while correcting the underlying hemostatic defect. Current evidence supports conservative management whenever clinically feasible, particularly in hemodynamically stable patients. Treatment includes close monitoring of vital signs, serial haemoglobin assessment, analgesia, blood transfusion when required, antifibrinolytic therapy, and administration of VWF-containing factor concentrates.

Surgical intervention should be reserved for selected situations, including persistent hemodynamic instability despite adequate replacement therapy, ongoing active bleeding, diagnostic uncertainty, or suspicion of an alternative surgical pathology

An important aspect of long-term care is prevention of recurrent ovulation-related bleeding. Hormonal suppression remains the mainstay of prophylaxis and may be achieved using combined oral contraceptive pills, progestin-based therapies, or gonadotropin-releasing hormone analogues in selected patients. Regular hematology follow-up and individualized replacement protocols may further reduce the risk of recurrent haemorrhagic ovarian cysts and hemoperitoneum. Patient education regarding recognition of symptoms and prompt medical consultation is equally important.

In conclusion, haemorrhagic ovarian cyst with hemoperitoneum is a rare but potentially life-threatening complication of Type 3 von Willebrand disease. This case highlights the importance of considering inherited bleeding disorders in reproductive-age women presenting with spontaneous hemoperitoneum. Early diagnosis, prompt hemostatic replacement therapy, and multidisciplinary management are essential for achieving favourable outcomes.

4. Conclusion

Haemorrhagic ovarian cyst rupture with hemoperitoneum is a rare but potentially life-threatening complication of Type 3 Von Willebrand disease. Early recognition and management can result in favourable outcomes while avoiding unnecessary surgery. Long-term preventive strategies, particularly ovulation suppression, are essential to reduce recurrence and improve quality of life.

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