

## **Von Willebrand's disease diagnosed after hemorrhage following hysteroscopic myoma resection and endometrial band excision**

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### **Abstract**

*Von Willebrand factor, the largest human plasma protein, is an adhesive multimeric glycoprotein that mediates platelet adhesion to both the subendothelial matrix and endothelial surfaces and acts as a carrier for coagulation factor VIII in the circulation. Von Willebrand disease (vWD) is the most common inherited bleeding condition that involves extended or excessive bleeding and is caused by the deficiency or defect of vWF. In this case report, we present vWD diagnosed after hemorrhage following hysteroscopic myoma resection and endometrial band excision. The importance of a detailed medical history is emphasized as even health care workers cannot spontaneously give this information. Further tests are recommended in patients who have a history of prolonged bleeding as bleeding time and other routine coagulation tests done preoperatively cannot always make a definite diagnosis.*

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### **Introduction**

The success of a surgical intervention depends on the control of the infection,

hemorrhage and pain. The diagnosis and treatment of an abnormal hemostatic condition and the prevention and control of hemorrhaging are essential for the success of an operation. Preoperative, intraoperative and postoperative hemorrhages are potential complications for each patient undergoing surgery.<sup>1</sup>

Herein, we report a case whose medical history, physical examination and hemostasis tests were normal, yet developed postoperative hemorrhage and was diagnosed with von Willebrand's disease (vWD).

### **Case Report**

The 26 year old patient, who is a nurse, was admitted to the Gynecology and Obstetrics Clinic of Universal Malatya Hospital with intermenstrual bleeding, menorrhagia and secondary infertility on November 17, 2012. Vaginal examination revealed a normal vulva and vagina, and a cervical

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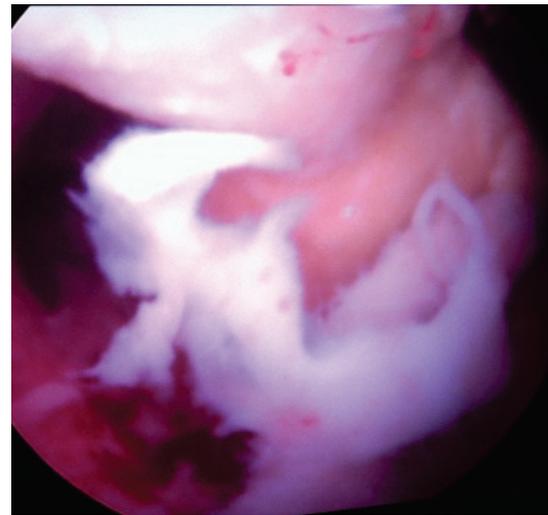
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erosion measuring 3x2 cm. Endometrial thickness was 18 mm in the middle parts of the endometrium and in the area adjacent to the cervix. An intramural subserous myoma measuring 28x26 mm was detected near the fundus. The patient underwent operative hysteroscopy 3 days later. Hysteroscopy revealed a grayish-white avascular endometrial band connecting both lateral walls of the uterus (Figure 1).

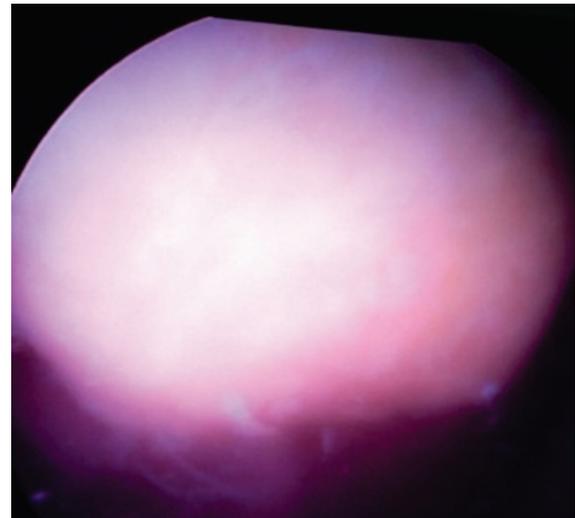


**Figure 1: Hysteroscopy revealed a grayish-white avascular endometrial band connecting both lateral walls of the uterus.**

The band was excised through a resectoscope and it proceeded into the cavity. A subserous myoma originating from the right wall of the uterus close to the fundus and measuring 3x3 cm was detected (Figure 2). Hysteroscopic myoma resection was completed and the patient was discharged 4 hours after the operation. The patient reported vaginal hemorrhaging and she was re-hospitalized. The erythrocyte count



**2a**



**2b**

**Figure 2 (focused submucous myoma): Submucous myoma originating from the right wall of the uterus close to the fundus and measuring 3x3 cm was detected.**

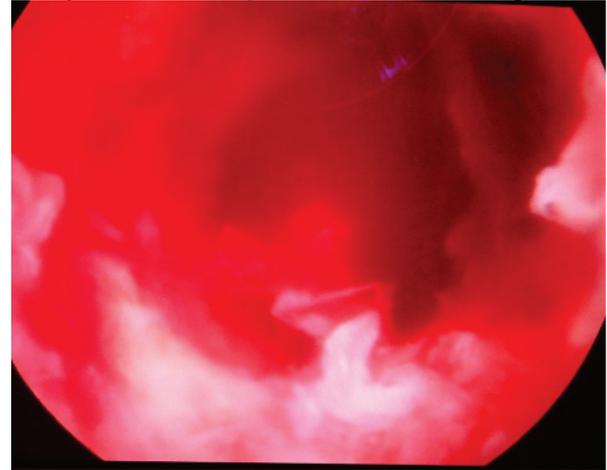
was  $5.02 \times 10^6$  uL, hemoglobin was 15.5 g/dl and hematocrit was 44.1% in the preoperative period. Platelet count and coagulation tests were normal. The erythrocyte count was found to be  $3.94 \times 10^6$ , hemoglobin was 11.9 g/dl and hematocrit was 34.6% following

the vaginal hemorrhage. A vaginal leakage originating from the uterus was detected on vaginal examination and diagnostic hysteroscopy (Figure 3). The patient was found to have prolonged menstrual bleeding and wound healing. She stated that she had prolonged bleeding during and after a caesarean section and that she had received a blood transfusion in the postoperative period. vWD was suspected based on these findings, and desmopressin and antifibrinolytic therapy was initiated. Bleeding was reduced but did not cease. A diagnosis of vWD was made as von Willebrand's factor (vWF) antigen level and vWFRcoF (ristocetin cofactor) activity was found to be 28. The patient was administered factor VIII (FVIII) concentrate containing 30 U/kg vWF via intravenous route and bleeding was then under control.

### **Discussion**

A patient who is being prepared for an operation must be questioned in terms of family history of hemorrhagic diathesis, previous operations and tooth extractions, history of hematuria, gastrointestinal hemorrhage, easily developing ecchymosis with simple traumas, hemarthrosis, menometrorrhagia or epistaxis. Even health care workers cannot give this medical history despite previous histories, as in our case. A detailed history of previous operations and menstrual bleeding patterns could have led us to suspect this disease. Postoperative hemorrhage may be related to both a reduced number of platelets and also platelet dysfunctions. Congenital diseases associated with poor platelet functions are divided into four major groups according to the type

of dysfunction; a) adhesion to collagen,



**Figure 3: A vaginal leakage originating from the uterus was detected on vaginal examination and diagnostic hysteroscopy.**

b) adhesion to subendothelium, c) release defects, d) ADP aggregation defects. All congenital defects except vWD in which there is a defect of adhesion to the subendothelial region are rare. vWD is an autosomal dominantly inherited disease characterized by absence, reduced production or abnormal functioning of a large multimetric protein, vWF, synthesized by the vascular endothelium and megakaryocytes. vWF is responsible for the regular binding of platelets to the collagen surface that emerges with a vascular trauma. Its absence leads to insufficient binding of platelets to the tear in the vascular bed by hindering platelet plug formation, which is essential for normal hemostasis. This condition cannot be detected in many patients until a vascular trauma develops or a surgical intervention is applied. In addition, those patients are partially sensitive to aspirin or other antiplatelet therapies, and a surgical intervention leads to severe bleeding in

those patients. vWD is the most common congenital coagulation disorder and the likelihood of being undetected until surgery is the highest. This disease is particularly dangerous because there is no history of surgical bleeding and preoperative coagulation tests are normal in its mild forms.<sup>1</sup>

Prevalence of vWD is 1% and excessive uterine bleeding is the most common problem for female patients, usually in the form of menorrhagia.<sup>2</sup> vWD is detected in 10-20% of the patients who are admitted to gynecology clinics with complaints of menorrhagia.<sup>3,4</sup> The history of menorrhagia could be alarming for the diagnosis of the disease and vWD could not be detected preoperatively with screening tests like bleeding time, coagulation time and prothrombin time, in our case. Classical laboratory findings of vWD include reduced vWF level, vWF ristocetin cofactor activity (RcoF), FVIII coagulation activity and reduced or increased platelet aggregation with ristocetin (RIPA).<sup>5</sup> Diagnosis was not suspected initially in our case; however, a diagnosis of vWD was suspected thereafter due to postoperative hemorrhage, prolonged bleeding and partial response to desmopressin and antifibrinolytic treatment. The disease was detected with further tests. Prothrombin time is normal in a great proportion of patients with type I vWD and the vast majority of patients with type 2A, 2B, 2M as it is prolonged only when FVIII-C level decreases below 40% of normal values. Therefore, tests of bleeding time, FVIII-C procoagulant activity, vWF antigen level, RcoF activity and ristocetin-induced platelet aggregation tests must be performed together in patients who are suspected to have

vWD. The bleeding time test has previously been accepted as essential; however, its reliability is poor as bleeding time is found to be normal in a great proportion of type I vWD patients and the reproducibility of the test is low. The PFA-100 system which has been developed in recent years is an in vitro primary hemostasis test which has taken the place of the bleeding time test and it is emphasized that it may exclude diagnosis of vWD. Further studies are needed to take the place of a more specific vWF assay test.<sup>6</sup>

### **Conclusion**

In conclusion, a good medical history and family history must be obtained from the patient in order to make an early diagnosis and prevent complications in subclinical cases of vWD, which is the most common hemorrhagic diathesis. The importance of a detailed medical history is emphasized. Further tests are recommended in patients who have a history of prolonged bleeding as bleeding time and other routine coagulation tests done preoperatively cannot always make a definite diagnosis.

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