Gazi Tıp Dergisi / Gazi Medical Journal

OLGU SUNUMU - CASE REPORT

2007: Cilt 18: Sayı 3: 127-129

HEMOPERITONEUM CAUSED BY HEMORRHAGIC CORPUS LUTEUM IN A TEENAGER WITH VON WILLEBRAND'S DISEASE: ULTRASONOGRAPHIC FINDINGS

Suna Özhan Oktar, M. D., Devrim Karaosmanoğlu, M. D., Cem Yücel, M. D., Hakan Özdemir, M. D.

Korpus hemorajikuma sekonder hemoperitoneum, von-Willebrand hastalığı bulunan olgularda sık görülmeyen bir bulgudur. Bu olgu yazısında hastaneye akut abdominal ağrı ile başvuran von-Willebrand hastalığı bulunan14 yaşındaki bir kız çocuğu sunulmuştur. Sonografide alt abdominal kadranlarda önemli miktarda sıvı ve sağ adneksde internal septasyonlar ve sıvı-sıvı seviyeleri içeren kompleks kistik lezyon izlenmiştir. Seri sonografik incelemeler, faktör VIII replasman tedavisi sonrası lezyonun hızla gerilediğini göstermiştir. Bu olgu ile, von-Willebrand hastalığı bulunan ergen kızlarda akut abdomenin ayırıcı tanısında düşünülmesi gereken, korpus hemorajikum ve buna sekonder gelişen eden hemoperitoneumun sonografik bulguları sunulmuştur.

Anahtar Kelimeler: von-Willebrand hastalığı--korpus hemorajikum--hemoperitoneum.

VON-WILLEBRAND HASTALIĞI BULUNAN BİR ÇOCUKTA HEMRAJİK KORPUS LUTEUM NEDENİYLE GELİŞMİŞ HEMOPERİTONEUM: ULTRASONOGRAFİK BULGULAR

Hemoperitoneum secondary to corpus hemorrhagicum is an unusual manifestation in patients with von Willebrand's disease. We report a 14-year-old girl with von Willebrand's disease who was admitted to hospital with acute abdominal pain. Sonography demonstrated a large amount of free fluid in the lower abdominal quadrants and a complex cystic lesion in the right adnexa, containing internal septations and fluid-fluid level. Serial sonographic examinations showed rapid resolution of the lesion with factor VIII replacement therapy. This case presents sonographic findings of corpus hemorrhagicum and hemoperitoneum that have been rarely documented in previously published cases and should be considered in the differential diagnosis of acute abdomen in teenage girls with von Willebrand's disease.

Key Words: von Willebrand's disease, corpus hemorrhagicum, hemoperitoneum.

Gazi University, School of Medicine, Department of Radiology, 06510, Beşevler-Ankara

INTRODUCTION

Von Willebrand's disease is a congenital bleeding disorder that is inherited as an autosomal dominant or recessive trait. The pathogenesis of bleeding is due to absence of platelet adhesion to the subendothelium, which is mediated by von Willebrand's factor. The plasma of the patient is also deficient in factor VIII coagulant activity (1). Females with existing hematological disorders are especially prone to develop hemorrhages from corpus luteum cysts. Therefore, corpus hemorrhagicum must be considered in the initial diagnosis of acute abdomen in females affected by congenital coagulation disorders like von Willebrand's disease.

The aim of this case report was to present the sonographic findings of hemoperitoneum caused by hemorrhagic corpus luteum in von Willebrand's disease, which has rarely been documented in previously published cases.

CASE REPORT

A 14-year-old female who had been followed-up with the diagnosis of Type III von Willebrand's disease since the age of two presented with acute pain in the right lower quadrant. Her medical history revealed menarche at 12 years of age with regular cycles, and no history of menorrhagia. Her last menstrual period was three weeks before the admission, and had lasted for four days. Her menstrual bleeding started the day after her hospitalization and it was normal in amount. She had had her last infusion of factor VIII two months before. Her physical examination revealed diffuse abdominal tenderness and low blood pressure (90/50 mmHg). She was negative for human chorionic gonadotropin. Investigation of peripheral blood revealed a hemoglobin level of 4.1 g/dl, hematocrit of 14.2%, and platelet count of 157,000 per mm^3.

The sonographic examination of the pelvis, performed with an ATL HDI 5000 ultrasound system (Philips-Advanced Technology Laboratories, Bothell, Washington, USA) and 3.5 MHz convex transducer, demonstrated a large amount of free fluid in the lower abdominal quadrants and cul-de-sac. There was a complex right ovarian cyst with multiple septations, internal echoes, and thick walls, measuring 77 x 75 x 67 mm. The lesion contained fluid-fluid level and hyperechoic areas suggestive of a blood clot (Figure 1). The left ovary was normal in size and sonographic appearance. A Doppler examination revealed the presence of blood flow to both ovaries. On the basis of her clinical background and ultrasonographic findings, the diagnosis was hemorrhagic corpus luteum cyst and hemoperitoneum. For the management of this complication, factor VIII replacement therapy was instituted together with red blood cell transfusion. No surgical procedure was performed. On serial sonograms a changing sonographic appearance due to clot lysis confirmed the provisional diagnosis of corpus hemorrhagi-

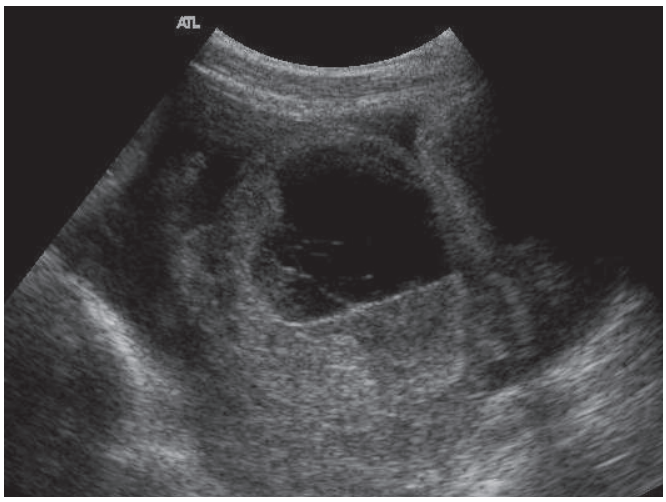

Figure 1: Ultrasound examination reveals a large, complex, thick-walled cystic lesion in the right adnexa, with internal septations and an echogenic area on the dependent site of the lesion suggesting a blood clot. There is also a large amount of free fluid in the pelvic region.

cum (Figure 2). The ultrasound examination performed fourteen days after the initial presentation showed a significant decrease in lesion size and a minimal amount of free fluid in the pelvic region (Figure 3).

With the cessation of menstrual bleeding and stabilization of her hemoglobin and hematocrit levels, the patient was discharged from hospital. A follow-up ultrasound evaluation performed two months after the onset of symptoms showed complete resolution of the lesion.

DISCUSSION

Von Willebrand's disease is the most common congenital bleeding disorder. International baseline population prevalence studies of von Willebrand's disease among children of school age suggested a high prevalence of approximately 1% (0.8-1.3%) (2, 3).

Von Willebrand's disease is due to an abnormality, either quantitative or qualitative, of the von Willebrand factor, which is a plasma protein required for the stabilization of factor VIII and mediates normal adherence of platelets to the site of vascular injury. It has a broad spectrum of clinical and laboratory findings ranging from severe hematological disorder to very mild asymptomatic forms. Females having von Willebrand's disease commonly experience excessive menses and postpartum bleeding (1).

The corpus luteal cyst is the most common etiology of hemorrhagic ovarian cyst, which is a frequent cause of acute pelvic pain in woman of child-bearing age and a common indication of referral for ultrasound evaluation. Intrafollicular bleeding into corpora lutea occurs 2-4 days after ovulation, and it is due to the increased vascularity of the ovary in the luteal phase. This spontaneous limited amount of bleeding fills the cavity of maturing corpus luteum. It has been suggested that if the active hemorrhage continues the intracystic pressure increases and rupture of the corpus luteum is possible, which is uncommon (4). Females who are on chronic warfarin therapy or have preexisting hematological disorders are especially prone to develop hemorrhagic corpus luteum. Von Willebrand's disease is one of the main bleeding disorders in which both primary hemostasis and coagulation are involved. Bleeding during ovulation is a major clinical complication in women with this disease. It can lead to hemoperitoneum when the bleeding is not contained in the ovary, but rather extravasates into the pelvis and abdominal cavity with massive internal blood loss. Such a rupture and hemorrhage are potentially catastrophic for the patient and often are misdiagnosed clinically (5). Therefore, it is important to recognize and diagnose this entity correctly. Acute onset of severe lower abdominal pain is the prominent symptom in those patients (6).

Ultrasonographic appearance of the hemorrhagic corpus

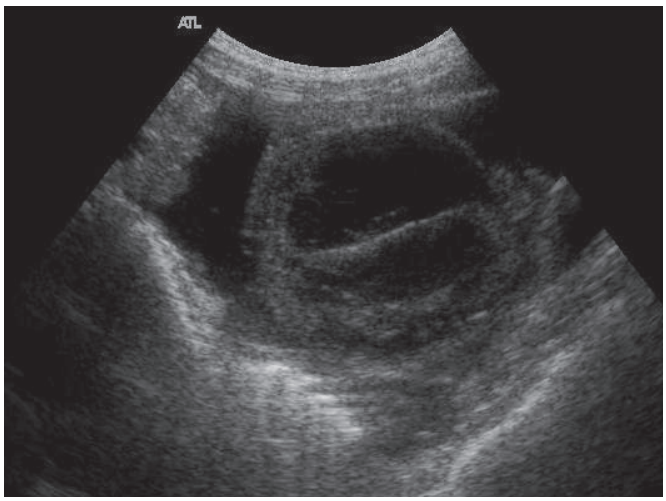

Figure 2: The ultrasound examination performed four days after the onset of symptoms shows evolution of the lesion with a slight decrease in its size.

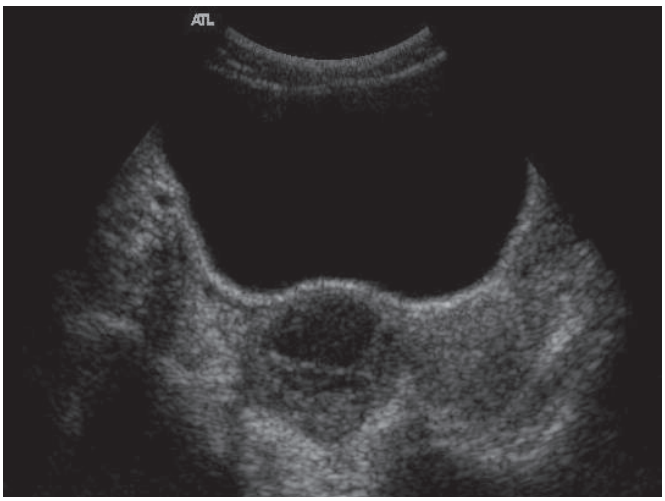

Figure 3: A follow-up ultrasound on the fourteenth day shows a significant decrease in the size of the lesion with minimal residual fluid.

luteum depends on the size of the lesion, and the time interval since the occurrence of hemorrhage (7). Common sonographic patterns include a fluid-containing mass with a thick rim, internal septations, internal echoes, or a homogenously hyperechoic mass (7-9). Ruptured hemorrhagic corpus luteum can present a range of imaging findings. In most cases the dominant imaging feature is hemoperitoneum rather than the cyst itself. Hemoperitoneum from a ruptured hemorrhagic corpus luteum exhibits imaging features similar to those of hemoperitoneum from other causes. This condition, in the setting of von Willebrand's disease, may respond to FVIII replacement therapy without the need for surgical intervention.

This case indicated that a provisional diagnosis of hemorrhagic corpus luteum and hemoperitoneum should be considered when a female patient with von Willebrand's disease develops acute abdomen. Ultrasonography remains the primary diagnostic tool in the evaluation and follow-up of such cases.

Correspondence Address
Suna Özhan Oktar, M.D.
Gazi Üniversitesi Tıp Fakültesi Radyoloji Anabilim Dalı 06510,
Beşevler, Ankara-TURKEY
Phone: 312 212 40 31
Fax: 312 212 19 40
e-mail: sunaoktar@gazi.edu.tr

REFERENCES

1. Kessler CM. Coagulation factor deficiencies. In: Goldmann L, Bennet JC eds. Cecil textbook of medicine. 21st ed.; Saunders, 2000: 1008-1010.
2. Rodeghiero F, Castaman G, Dini E. Epidemiological investigation of the prevalence of von Willebrand's disease. Blood 1987; 69: 454-459.
3. Werner EJ, Broxson EH, Tucker EL, Giroux DS, Shults J, Abshire TC. Prevalence of von Willebrand disease in children: a multiethnic study. J Pediatr. 1993; 123: 893-898.
4. Droegmuller W. Benign gynecological lesions. In: Stenchever MA, Droegmuller W, Herbst AI, Mishell DR, eds. Comprehensive gynecology. 4th ed. St. Louis, MO: Mosby 2001; 507-511.
5. Hertzberg BS, Kliewer E, Paulson EK. Ovarian cyst rupture causing hemoperitoneum: imaging features and the potential for misdiagnosis. Abdominal Imaging 1999; 24: 304-308.
6. Jarvis RR, Olsen ME. Type I von Willebrand's disease presenting as recurrent corpus hemorrhagicum. Obstet Gynecol 2002; 99: 887-888.
7. Yoffe N, Bronsthein M, Brandes J, Blumenfeld Z. Hemorrhagic ovarian cyst detection by transvaginal sonography: the great imitator. Gynecol Endocrinol 1991; 5: 123-129.
8. Baltarowich OH, Kurtz AB, Pasto ME, Rifkin MD, Needleman L, Goldberg BB. The spectrum of sonographic findings in hemorrhagic ovarian cyst. AJR 1987; 148: 901-905.
9. Reynolds T, Hill MC, Glassman LM. Sonography of hemorrhagic ovarian cyst. J Clin Ultrasound 1986; 14: 449-453.