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A case of disseminated intravascular coagulation developed after surgical management of corpus luteal hemorrhage in a patient with Klippel-Trenaunay syndrome

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Summary

Klippel-Trenaunay syndrome (KTS) is a complex congenital disorder characterized by a triad of varicose veins, cutaneous capillary malformation, and hypertrophy of bone and/or soft tissue. KTS may be associated with massive hemorrhage or coagulopathy that be a life-threatening situation. Although women in reproductive age are at risk of ruptured corpus luteum with active arterial bleeding, if it managed properly, the development of serious complications, such as disseminated intravascular coagulation (DIC) rarely develops. However, in case of patient with vascular malformation, there is possibility of unexpected complication occurrence such as DIC. The authors report a case of a 29-year-old female with KTS who presented with corpus luteal hemorrhage and which lead to DIC, despite adequate surgical and medical treatment.

Key words: Klippel-Trenaunay syndrome; Congenital vascular anomaly; Corpus luteum; Hemoperitoneum.

Introduction

Klippel-Trenaunay syndrome (KTS) is a rare congenital vascular malformation characterized by the clinical triad of bony or soft tissue hypertrophy, usually affecting one extremity; hemangiomas and/or lymphangiomas, and varicosities or venous malformations [1]. Vascular malformation of KTS can involve several organs and be a source of significant morbidity and even mortality [2]. Clinical manifestations of KTS range from occult to massive, life-threatening hemorrhage [3]. Several cases of life-threatening gastrointestinal bleeding in KTS patients have been reported [3-6]. In contrast, large venous malformations can be associated with low-grade consumptive coagulopathy [7, 8].

Spontaneous hemoperitoneum may occur in various gynecological conditions. The most common gynecological causes of spontaneous hemoperitoneum in women of childbearing age are ectopic pregnancy and ruptured corpus luteal cyst [9]. The treatment of spontaneous hemoperitoneum of gynecological causes is well established. However in patients with coagulopathy, the management has to be planned differently.

The authors present a case of a 29-year-old female with KTS who presented with a spontaneous hemoperitoneum by corpus luteal hemorrhage and subsequent life-threatening disseminated intravascular coagulation (DIC) after ovarian cystectomy.

Case Report

A 29-year-old female presented to the emergency room with sudden onset abdominal pain. She had a history of multiple admissions for cutaneous large hemangioma, presenting mainly as hemangioma bleeding that started at the age of 13 and diagnosed KTS. She had regular menstruations since the age of 13, lasting three to five days, occurring every 28 to 30 days. She presented with tachycardia (pulse rate 112 beats per minute), abdominal distension, and moderate rigidity on the lower quadrants on palpation on physical examinations. In addition, blood pressure (BP) was 90/60 mmHg and general appearance was acute ill-looking but alert and oriented. Laboratory results showed anemia (hemoglobin 7.6 g/dl) and slight elevation of PT (INR 1.40) and aPTT time (44.6 sec). Transabdominal ultrasound revealed a moderate amount of fluid in the abdomen and in the pouch of Douglas and a seven-cm sized right adnexal mass with signs of peripheral vasculatization. An abdomen CT scan showed hemoperitoneum due to rupture of hemorrhagic cyst in right adnexa and diffuse hemangioma with venous malformation in left side of body (Figure 1).

The patient was taken to the operating room with a presumptive diagnosis of a ruptured hemorrhagic cyst. She underwent exploratory laparotomy via Pfannenstiel's skin incision on right side of lower abdomen. There was a massive hemorrhage due to corpus luteum cyst rupture on the surface of the right ovary. Both fallopian tubes, left ovary, and uterus appeared to be normal. During the surgery, an amount of hemorrhagic fluid was evacuated from the abdominal cavity. Hemostatic electric coagulation was applied to the bleeding site and right ovarian wedge resection was performed. In addition, five units of packed red blood cell and five units of fresh frozen plasma were transfused during the surgery.

Postoperative course was complicated by hemoperitoneum, worsening coagulopathy, sepsis, and DIC. The enhanced CT

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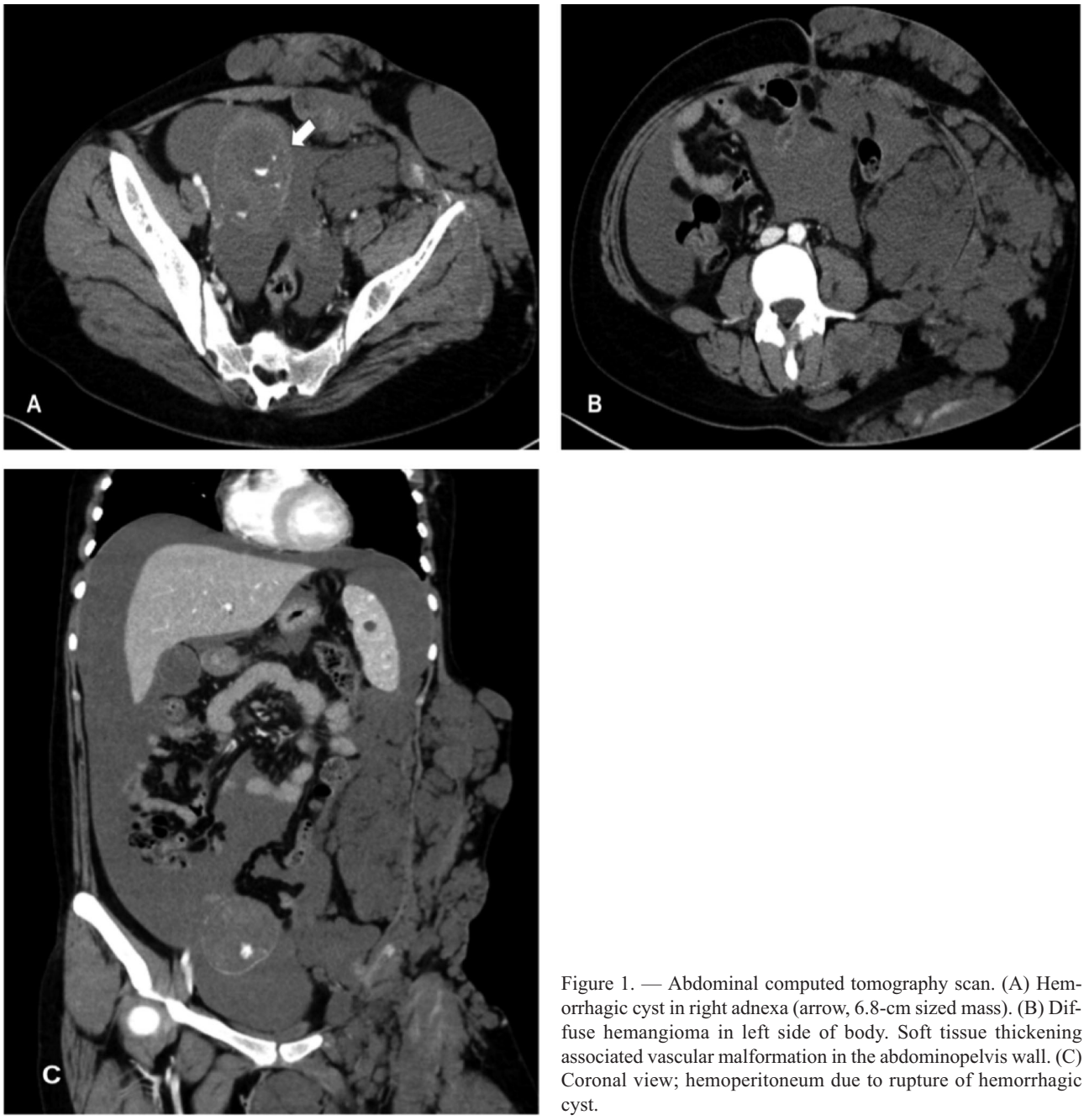

Figure 1. — Abdominal computed tomography scan. (A) Hemorrhagic cyst in right adnexa (arrow, 6.8-cm sized mass). (B) Diffuse hemangioma in left side of body. Soft tissue thickening associated vascular malformation in the abdominopelvis wall. (C) Coronal view; hemoperitoneum due to rupture of hemorrhagic cyst.

scan showed active contrast leakage in pelvic cavity and hemoperitoneum around uterus (Figure 2). The authors attempted to manage hemorrhage in regards to the treatment of DIC. However, active arterial bleeding in cystectomy site was observed on CT follow-up. The follow-up secondary laparotomy was decided on the next day. A right oophorectomy was performed and the bleeding site was sutured to manage hemorrhage. Immediate postoperative hemoglobin was 7.9 g/dl and platelet count was 60,000/ul and the patient received massive transfusion after surgery due to chronic DIC with multiple hemangioma in KTS syndrome. Although active bleeding was controlled and stable vital sign was maintained after the re-operation, the patient was transferred to the department of internal medicine because of continuous bleeding in the drain site due to DIC, and was eventually discharged after three months.

Discussion

KTS is a rare congenital disorder with variable clinical presentation related to malformations of blood and lymph vessels, and disturbed growth of bone and soft tissue [1]. KTS is a complex congenital syndrome which is associated with life threatening complications like bleeding in geni-

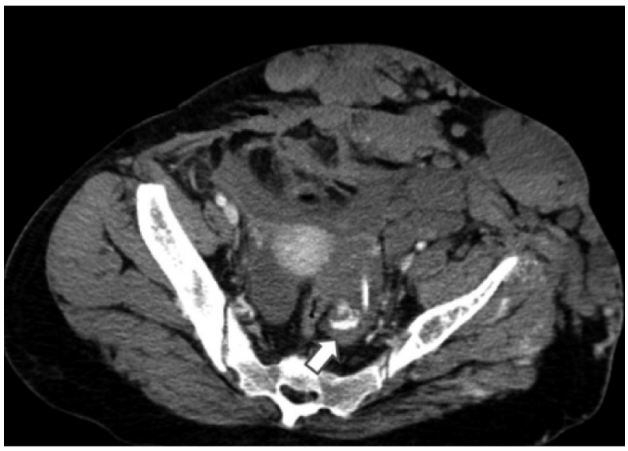

Figure 2. — Abdominal computed tomography scan (arrow indicates active contrast leakage in the right ovary and in the left pelvic cavity).

tourinary system, spleen, liver, gastrointestinal tract, or central nervous system [10]. Several cases of life-threatening situations due to bleeding in KTS patients have been reported. There are several treatments such as sclerotherapy, excision of vasricose vessel, and conservative therapy that are attempted in the patients, yet, no appropriate treatment has been determined. In the previous reports, organ involvements of hemangioma were reported, not corpus luteal hemorrhage in KTS [3, 6]. Corpus luteum hemorrhage may occur spontaneously or often triggered by coitus, trauma, exercise, or vaginal examination. The risk of hemorrhagic complications of ovulation begins on the ovulation day and extends throughout corpus luteal life span, which is 14 days without pregnancy.

The treatment of corpus luteal hemorrhage includes both conservative and surgical management. Conservative management is successful in the majority of patients with ruptured ovarian cysts with hemoperitoneum. However, surgical intervention is absolutely indicated in presence of low blood pressure and a large amount of hemoperitoneum [11]. Surgical management includes the following: electrocauterization of the ovarian surface, cystectomy, wedge resection, and ovarian reconstruction.

The present authors performed ovarian cystectomy and confirmed the absence of active bleeding before leaving the operation room. After the first surgery, hemorrhage was observed and it was considered due to the active bleeding caused by DIC. After taking the volume of bleeding and transfusion into consideration, coagulopathy in KTS might be the main cause of bleeding.

In ovarian cystectomy, the suture in ovarian stroma is required and this may increase the risk of bleeding in the presence of DIC compared to when oophorectomy is performed.

In the cases of women with coagulopathy, we have to consider more bleeding preventive operative technique such as oophorectomy, not cystectomy. Preserving ovarian function must be considered in the surgery of young women who have coagulopathy. However we have to bear in mind that DIC can develop regardless of volume of bleeding in the operation of the patient with coagulopathy.

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