Spontaneous massive hemoperitoneum from hemorrhagic corpus luteum cyst as initial presentation of aplastic anemia

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ARTICLE INFO

Article history:
Received 2 May 2014
Received in revised form
17 June 2014
Accepted 23 June 2014

Key words:
Spontaneous hemoperitoneum
Hemorrhagic ovarian cyst
Aplastic anemia

A B S T R A C T

While hemorrhagic ovarian cysts are quite common, rupture leading to spontaneous severe hemoperitoneum is much more rare, and in the setting of an acquired bone marrow failure syndrome, it can be a life-threatening event. We describe a 17-year-old female presenting with syncope, abdominal pain, and large-volume hemoperitoneum, associated with diffuse petechiae and oral purpura, who was found to have a hemorrhagic corpus luteum cyst at laparoscopic exploration. After discharge, the patient represented with severe anemia and thrombocytopenia, and was diagnosed with acquired severe aplastic anemia of unknown etiology.

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Spontaneous massive hemoperitoneum secondary to a hemorrhagic corpus luteum cyst is an exceedingly rare, but potentially life-threatening presentation, with few cases reported in the literature. Severe hemorrhage from a corpus luteum cyst as initial presentation of an underlying bone marrow failure syndrome has not previously been described. We present a case of a 17-year-old female presenting in extremis with massive hemoperitoneum from an ovarian cyst.

1. Case report

A 17-year-old female presented to an urgent care center with syncope, emesis, and severe abdominal pain for the previous 4–6 h. Her initial heart rate was 103 beats/minute, with a measured blood pressure of 62/34 mm Hg. On exam, she was pale, diaphoretic, and hypotensive, and was noted to have scattered, purpuric lesions on her left lower extremity, with rare petechiae on all four extremities. On initial laboratory work-up, she was noted to have a hemoglobin (Hgb) of 8 g/dL, hematocrit of 23.6%, and platelet count of 8 × 10^9/L. She was transferred to Le Bonheur Children’s Hospital (LBCH) after receiving 1.5 L of intravenous fluid resuscitation. On arrival to LBCH, she continued to be tachycardic and hypotensive, with a measured blood pressure of 56/36 mm Hg. Repeat blood work showed a Hgb of 5.7 g/dL, white blood cell (WBC) count 7.2 × 10^9/L, reticulocyte count 28.4 × 10^9/L, mean corpuscular volume 97.0 fL, absolute neutrophil count (ANC) 6696 × 10^6/L, and a platelet count of 7 × 10^9/L, with normal prothrombin time and activated partial thromboplastin time. The patient did not have any previous peripheral blood counts with which to compare these results. A computed tomography scan of the abdomen and pelvis showed large hemoperitoneum, associated with diffuse petechiae and oral purpura, who was found to have a hemorrhagic corpus luteum cyst at laparoscopic exploration. After discharge, the patient represented with severe anemia and thrombocytopenia, and was diagnosed with acquired severe aplastic anemia of unknown etiology.

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normal. Gynecology was also consulted, and on further history, she admitted to having heavy and prolonged menstrual cycles. The patient was admitted to the pediatric intensive care unit and resuscitation completed.

After the patient was resuscitated and hemodynamically stable, she was taken to the operating room by pediatric surgery and gynecology services for a diagnostic laparoscopy. On exploration, the patient was found to have approximately 2 L of hemoperitoneum and a right ovarian cyst with an area of ulceration (Fig. 2). This cyst was removed while preserving the ipsilateral ovary, sent for frozen section as well as permanent pathology and shown to be a corpus luteum cyst. Post-operatively, the patient recovered well with stabilization of her hematocrit and platelet count. She had appropriate return of bowel function and was discharged on post-operative day two. Peripheral blood counts at discharge showed a Hgb of 8.6 g/dL, WBC 2.8 × 10^9/L, ANC 840 × 10^9/L, and platelets 54 × 10^9/L. The patient had a scheduled outpatient follow-up appointment with the hematology service two weeks after discharge.

On post-operative day 11, the patient presented to the hematology clinic at St. Jude Children’s Research Hospital with a one-week history of fatigue, worsening petechiae on the extremities and chest, bruising on legs, a prolonged menstrual cycle, diffuse purpura on lips, and a one-day history of gingival bleeding. On further questioning, she admitted to having had diffuse petechiae and easy bruising for the last several months. On exam, she was again pale with orthostatic hypotension. She was found to have a Hgb of 6.9 g/dL, WBC count of 2.6 × 10^9/L, ANC of 1100 × 10^9/L, reticulocyte count of 15.7 × 10^9/L, and platelet count of 6 × 10^10/L. At this time, she was transfused 2 units of PRBCs and 2 units of platelets. The patient had negative hepatitis and human immunodeficiency viral studies. She had positive viral serologies for Epstein-Barr virus IgG and cytomegalovirus IgG, but IgM titers were negative. She had a bone marrow biopsy and aspirate performed the next day, which returned as hypocellular marrow with 5–10% cellularity, scant hematopoiesis, no cellular dysplasia, and absence of megakaryocytes. No structural abnormalities were noted on cytogenetics, and no morphologic evidence of leukemia was seen. Further work-up excluded any underlying inherited bone marrow failure syndrome and paroxysmal nocturnal hemoglobinuria; therefore, she was diagnosed with severe acquired aplastic anemia (SAA).

The patient underwent an allogeneic HLA-matched sibling donor stem cell transplant 35 days after admission, which she tolerated well. At 18-month follow up, the patient’s blood counts were stable with Hgb of 13.4 g/dL, WBC 4.8 × 10^9/L, ANC 2300 × 10^9/L, and platelets 265 × 10^9/L, and she had 100% donor chimerism with no evidence of graft-versus-host disease. She had normal renal, hepatic, cardiac, and pulmonary function, and her Lansky score was 100%. She had not suffered any further bleeding episodes or abdominal complaints.

2. Discussion

Spontaneous hemoperitoneum is a rare and potentially life-threatening condition defined as blood within the peritoneal cavity of non-traumatic etiology [1,2]. Kasotakis recently reviewed this condition and outlined the most common sources of hemorrhage as hepatic, splenic, vascular, or gynecological [3]. This condition typically presents with signs of acute intraperitoneal bleeding, abdominal pain, tachycardia, and hypotension. Computed tomography and ultrasound are the most commonly used imaging modalities.

Hallatt and colleagues described the first large series of patients with corpus luteum hemorrhage and hemoperitoneum [4]. They noted that this entity occurs at all stages of a woman’s reproductive life, and that a wide range of volumes of hemoperitoneum can be found at the time of exploration. Corpus luteum hemorrhage may be a cause of spontaneous hemoperitoneum in patients with bone marrow failure and bleeding disorders, and has been described in women with immune thrombocytopenic purpura (ITP), hemophilia or hemophilia carrier status, afibrinogenemia, von Willebrand disease, and factor X, VII, V, II, and XIII deficiencies [5–10].

Aplastic anemia is a heterogeneous group of blood disorders resulting in pancytopenia and hypocellular bone marrow due to bone marrow failure. Patients present with signs and symptoms of pancytopenia such as bleeding, fatigue, exercise intolerance, pallor, and fever or focal signs of infection. More than 70% of cases are considered idiopathic without any identifiable family history of exposure. Severe aplastic anemia is present when patients meet the following criteria: (1) bone marrow cellularity <25%, and (2) at
least two of the following: ANC $<0.5 \times 10^9$/L, platelet count $<20 \times 10^9$/L, or reticulocyte $<60 \times 10^9$/L [11]. This diagnosis requires exclusion of other congenital or acquired marrow failure syndromes, such as Fanconi anemia, dyskeratosis congenita, and paroxysmal nocturnal hemoglobinuria.

Corpus luteum hemorrhage causing massive hemoperitoneum associated with aplastic anemia has been previously described in a patient with known SAA [12]; however, this patient’s presentation was different in that it was the first episode of severe bleeding in an otherwise previously healthy teenage female, which ultimately prompted the work up and diagnosis of her bone marrow failure syndrome. The signs and symptoms of aplastic anemia, but also of some mild bleeding disorders (for example, von Willebrand disease), can be very subtle and difficult to diagnose. Differentiating the mechanisms of thrombocytopenia after a severe bleeding episode from consumptive causes such as immune destruction (for example, ITP) or non-immune destruction (for example, blood loss, disseminated vascular coagulation, or fever-induced) versus disorders of platelet production (for example, bone marrow failure syndromes, toxic or viral myelosuppression) can be challenging. Taking a detailed bleeding history on all surgical patients is critical, and maintaining a wide differential diagnosis can aid in appropriate work-up and optimize peri-surgical management [11–14]. In patients presenting with corpus luteum cyst with significant bleeding, a hematology consult should be considered.

3. Conclusion

Spontaneous massive hemoperitoneum is a rare and life-threatening condition in the setting of blood disorders, particularly disorders that predispose patients to bleeding. This case adds to the list of rare causes of massive bleeding from ruptured corpus luteum cysts. The facts of interest include: first, the patient appeared to have had signs of aplastic anemia for many weeks prior to presentation, which went undiagnosed. Secondly, the patient presented in extremis with massive hemoperitoneum requiring intensive resuscitation secondary to brisk bleeding. Although the differential diagnosis should include acute trauma, in certain patients, an underlying hematologic condition should be suspected.

Conflict of interest statement
The authors have no conflict of interest to disclose.

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