

# Large Ovarian Hemorrhagic Cyst and Immune Thrombocytopenia as Early Manifestations of Systemic Lupus Erythematosus (SLE)

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Submit: 30-03-2011, Accept: 20-05-2011

## Abstract

Hematologic abnormalities are generally present among systemic lupus erythematosus patients. Idiopathic thrombocytopenic purpura can be the first manifestation of SLE, followed by other symptoms and signs of disease appearing several years later. Although bleeding due to immune thrombocytopenic purpura is usually mild and occurs in mucocutaneous surfaces, but it may be severe and represent in unusual sites such as an ovarian cyst. We report a case of SLE presented with immune thrombocytopenia and a large hemorrhagic ovarian cyst.

**Key Words:** Hemorrhagic, ovarian cysts, lupus erythematosus, systemic, thrombocytopenia

## Introduction

Systemic lupus erythematosus (SLE) is a chronic inflammatory and multisystem disease of unknown cause. Hematologic abnormalities are commonly present in patients with SLE. The major hematologic findings are anemia, lymphopenia, thrombocytopenia and the antiphospholipid syndrome.<sup>1,2</sup>

The mechanism of immune thrombocytopenia in SLE is binding of antibodies to platelets followed by phagocytosis in the spleen.<sup>3</sup> Idiopathic thrombocytopenic purpura (ITP) may be the first manifestation of adults with SLE<sup>4</sup>, followed by other symptoms and signs several years later. It has been estimated that 3 to 15 percent of patients with ITP progress to develop SLE.<sup>5</sup> Severe bleeding due to immune-mediated thrombocytopenia is rare. SLE patients with thrombocytopenia often have significant associated organ involvement, such as lungs, kidneys and the CNS.<sup>6</sup> The most common clinical manifestations of immune thrombocytopenia are petechiae, purpura, and ecchymoses, especially on the limbs. Idiopathic thrombocytopenic purpura may be the first presentation of SLE, preceding this diagnosis by many years.<sup>7-8</sup> Corticosteroids are the mainstay of treatment for immune thrombocytopenia in

patients with SLE. Rituximab is also used in SLE patients with refractory immune thrombocytopenia. In 2006 Ylmaz et al. reported a 17 y/o female; with massive intra-abdominal hemorrhage secondary to distended ovarian follicle rupture.<sup>9</sup> Mori et al. have also reported a girl with chronic ITP who developed intraperitoneal hemorrhage caused by hemorrhagic ovarian cyst at the luteal phase.<sup>10</sup> Kaplan et al. reported an 18 y/o female with EBV-associated ITP, who developed a severe intra abdominal bleeding secondary to a hemorrhagic ovarian cyst.<sup>11</sup> We report a case of SLE presented with skin petechial lesions, immune thrombocytopenia and a large hemorrhagic ovarian cyst.

## Case Report

A 28 y/o female was admitted with skin petechiae, ecchymoses and thrombocytopenia.

Laboratory findings included: WBC=10,000/ $\mu$ l (N=75%, Lymph=25%), Hb=9.5 gr/dl, MCV=81 fl, PLT=7,000/ $\mu$ l and reticulocyte count=2.1%.

BUN, Cr, liver function tests, PT, PTT, Iron and TIBC were all in normal ranges. Serum ferritin was low. Coomb's test (direct and in direct) was negative. Study of peripheral blood smear showed large and giant Platelets and mild hypochromia in

RBCs series. Bone marrow aspiration and biopsy revealed polymorphic, normocellular marrow with increased number of megakaryocytes.

Abdomino-pelvic ultrasound revealed a 6×7 Cm heterogeneous mass in right ovary. AFP and β-HCG were negative. Trans-vaginal ultrasonography revealed a right ovarian hemorrhagic cyst (52×58mm). Serologic tests for ANA and Anti-ds DNA were Positive. LDH and CA-125 were also normal. Ferrous sulfate and oral prednisolone 1 mg/kg/day were prescribed. Over a 4 days period platelet counts increased to 140,000/μl. Diagnosis of SLE was made based on immune thrombocytopenia and positive ANA and Anti-ds DNA.

## Discussion:

Immune thrombocytopenia is a common manifestation of SLE and may be the first manifestation of it. Although bleeding due to immune thrombocytopenic purpura is usually mild and occurs in mucocutaneous tissues, but it may be severe and appear in unusual sites. A review of literature shows that hemorrhagic ovarian cyst may be secondary to immune thrombocytopenia. Mori et al. reported a girl with chronic ITP who developed intraperitoneal hemorrhage caused by hemorrhagic ovarian cyst at the luteal phase.<sup>10</sup> Kaplan et al. presented an 18-year-old female with EBV-associated ITP, who developed a severe intra-abdominal bleeding secondary to a hemorrhagic ovarian cyst.<sup>11</sup> Takai presented a 24-year-old female with acute idiopathic thrombocytopenic purpura presenting as ovarian and peritoneal hemorrhage.<sup>12</sup> We presented in this report a case of SLE with immune thrombocytopenia and a large asymptomatic ovarian hemorrhagic cyst.

Conclusion: SLE may be preceded by immune thrombocytopenia in the presence of positive ANA for prolonged periods of time. It is recommended that patients with a positive ANA and ITP serially be looked for clinical and laboratory manifestations of SLE in other organs. Abdomino-pelvic sonography should be recommended for all patients with immune thrombocytopenia and/or SLE with abdominal pain.

**Conflict of interest: none declared.**

## References:

1. Keeling DM, Isenberg DA. Haematological manifestations of SLE. *Blood Rev* 1993; 7:199.

2. Chen JL, Huang XM, Zeng XJ, et al. Hematological abnormalities in systemic lupus erythematosus and clinical significance thereof: comparative analysis of 236 cases. *Zhonghua Yi Xue Za Zhi*, 2007;87( 19): 1330-3
3. Michel M, Lee K, Piette JC, et al. Platelet autoantibodies and lupus-associated thrombocytopenia. *Br J Haematol* 2002; 119:354.
4. Unal I, Ceylan C, Ozdemir F, et al. ITP as an initial manifestation of subacute cutaneous lupus erythematosus. *J Dermatol* 2005; 32(9):727-30
5. Karpatkin, S. Autoimmune thrombocytopenic purpura. *Blood* 1980; 56:329.
6. Ziakas PD, Giannouli S, Zintzaras E, et al. Lupus thrombocytopenia: clinical implications and prognostic significance. *Ann Rheum Dis* 2005; 64:1366.
7. Hepburn MJ, English JC, 3rd, Keeling JH, 3rd. Autoimmune idiopathic thrombocytopenic purpura with the subsequent occurrence of systemic lupus erythematosus. *Cutis* 1997; 60:185.
8. Mestanza-Peralta M, Ariza-Ariza R, Cardiel MH, Alcocer-Varela J. Thrombocytopenic purpura as initial manifestation of systemic lupus erythematosus. *J Rheumatol* 1997; 24:867.
9. Yilmaz S, Demircioğlu F, Türker M, et al. An Extremely Uncommon Complication of ITP: Spontaneous Rupture of an Ovarian Follicle Cyst and Massive Intra-abdominal Bleeding. *Journal of Pediatric Hematology/Oncology*.2006; 28(11):755-6
10. Mori J, Morimoto A, Kinoshita Y, et al. Massive Intraperitoneal Hemorrhage Caused by Hemorrhagic Ovarian Cyst in a Patient with Chronic Idiopathic Thrombocytopenic Purpura. *Japanese Journal of Pediatric Hematology*,2003 :399-401
11. Kaplan J, Bannon CC, Hulse M, Freiberg A.. Peritoneal hemorrhage due to a ruptured ovarian cyst in ITP. *J Pediatr Hematol Oncol*. 2007 ; 29(2):117-20.
12. Takai Y. Acute idiopathic thrombocytopenic purpura presenting as ovarian and peritoneal hemorrhage. *Med Postgrad*.2006;44( 1):77-9