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Thrombocytopenia and Deep Vein Thrombosis on the Top of an Iceberg

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ABSTRACT

Systemic Lupus Erythematous (SLE) is a chronic autoimmune disorder with a relapsing-remitting course. Besides, SLE most commonly occurs in child-bearing-age women. Due to protean manifestations, the diagnosis may be challenging; however, a high index of suspicion, i.e. achieved by experience and perceptivity is the key to a correct decision. Here, we present an SLE patient; her initial symptoms resembled a malignant process, but important elements guided us to the underlying autoimmune process. A 34-year-old woman presented with a uterine mass, deep vein thrombosis, and significant thrombocytopenia. Our first impression was malignant processes; however, considering her age with thrombosis and thrombocytopenia as the key components of her presentation, we suspected antiphospholipid syndrome and SLE. Her abdominal mass was a benign leiomyoma. We treated her with glucocorticoids and anticoagulants. Accordingly, after improving cytopenia, we discharged her and the follow-up examination result was satisfactory.

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disorder with relapsing-remitting courses; SLE most commonly occurs in child-bearing age woman which may affect almost every organ. It could present with various manifestations, with constitutional, hematologic, and musculoskeletal symptoms as the most prevalent ones [1, 2].

The protean manifestations of SLE make the diagnosis a challenging issue in some situations; however, a high index of suspicion, i.e. achieved by experience and perceptivity, is the key to making the correct diagnosis. Here, we presented a case of SLE; her initial symptoms

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resembled a malignant process, but important elements guided us to the correct diagnosis.

Case Presentation

A 34-year-old woman presented to the emergency department of the hospital with a one-week history of right leg cramps and swelling. On examination, the pitting edema was evident all over her leg, extended to the upper calf. Colour Doppler ultrasonography confirmed deep vein thrombosis in the affected leg.

Her medical history was notable for a few months' experience of abdominal pain and heaviness. A further workup with ultrasonography revealed an abdominal mass. An abdominopelvic Computed Tomography (CT) scan indicated a large mass with central necrosis arising from the posterior wall of the uterine body and fundus extending to the right side of the abdominal cavity. Furthermore, the mass was compressing adjacent structures without any invasion or lymphadenopathy (Figure 1). As a result, she was a candidate for surgical resection. Additionally, she reported a history of hypothyroidism, i.e. under treatment by levothyroxine.

Initial laboratory studies data revealed isolated significant thrombocytopenia of 22000/µL. Reviewing her peripheral blood smear confirmed thrombocytopenia but no platelet aggregation or other abnormalities. We considered initiating platelet transfusion concomitant with anticoagulant treatment by unfractionated heparin. This is because her platelet count was <30000/µL and she was at risk for bleeding with anticoagulation.

Abdominal mass CT-guided biopsy demonstrated benign spindle-shaped cells, suggestive of leiomyoma. Bone marrow aspiration revealed normal marrow cells with megakaryocytosis.

The autoimmune panel revealed positive Antinuclear Antibody (ANA) >100 U/mL (>×10 times normal level), double-stranded DNA 610 IU/mL (>×6times normal level), and positive antiphospholipid antibodies [anti cardiolipin IgM 17.9 U/mL (>7: positive) IgG 10 U/mL (>10: positive), lupus anticoagulant 42 s(30-38s) beta-2 glycoprotein IgM & IgG were negative]; however, renal and thyroid function tests, complements level, and activity was normal. The alphabetic hepatitis virus markers of A, B, and C were also negative. ESR was measured as 12 mm/h and C-reactive Protein (CRP) was negative. Anti SSO and Anti La Ab were calculated as <3 and negative.

Careful history revealed that she had a cousin who expired from severe SLE. The patient presented no signs of malar rash, photosensitivity, or oropharyngeal ulcers. We considered treating her with intravenous immunoglobulin, glucocorticoid (prednisolone 60 mg/day), hydroxychloroquine, and a therapeutic dose of heparin. Her platelet count started to rise, reached 179000/µL, then she was referred for the surgical removal of a uterine mass. Her abdominal mass was 28×20×13 cm in size, arising from the uterine. Pathologic examination finding was consistent with leiomyoma.

Her follow-up visit examination result was satisfactory. Moreover, her platelet counts stabilized around 280000/µL, and prednisolone (60 mg/d) and hydroxychloroquine (300 mg/d), and warfarin (5 mg) were continued for her. During two months, her prednisolone was tapered to 5 mg, without any decrease in platelet level or any other problems. After three months, her ANA and Anti ds DNA became negative. Anti-Cardiolipin Ab IgM was equal to 9.6 U/mL (>7 positive) and IgG<3 GPLU/ml (negative) lupus anticoagulant 48s (30-38s), APTT 49s (31-44), Beta-2 glycoprotein IgM and IgG were again negative. Anti SM Ab was measured as 0.8 U/mL and negative.

Discussion

SLE is a well-known autoimmune syndrome with a particular tendency for young women which can involve any organ system [1]. Among hematologic manifestations, thrombocytopenia is relatively common in SLE (occurs in 7%-40% of cases); however, severe thrombocytopenia is rare [3, 4]. Three mechanisms are proposed to be involved in the pathogenesis of thrombocytopenia of SLE, as follows: myelosuppression, redistribution (in spleen), and peripheral destruction -which later consti-

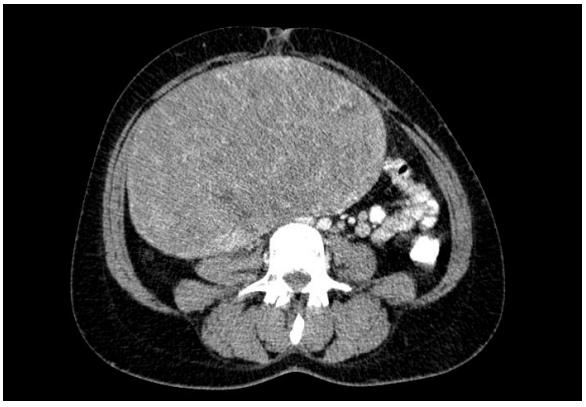

Figure 1. Abdominopelvic CT scan

A large mass with central necrosis arising from the posterior wall of the uterine body without any invasion or lymphadenopathy.

tute the most important cause- induced by anti-platelet and antiphospholipid antibodies [4-6]. As thrombocytopenia in SLE is immune-mediated, glucocorticoids are useful for treatment [3].

There is an association between SLE and APS, i.e. often presented with recurrent vascular thrombosis and fetal losses (lower limb deep vein thrombosis is the most frequent presentation). Approximately 40% of SLE patients present Antiphospholipid (aPL) antibodies in their sera; however, they become less symptomatic (i.e. APS). Besides the vascular and pregnancy complications of APS, patients with SLE who encounter an aPL or APS are at higher risks of developing neuropsychiatric and renal involvements and lower survival, compared to those without an aPL or APS [7].

Our patient was a young woman presenting with a large abdominal mass, Deep Venous Thrombosis (DVT), and thrombocytopenia. When we confronted abdominal mass with thrombosis, our first impression was malignant causes; however, she was a young woman who was presented with throm- bosis and thrombocytopenia. Thus, we were strongly suspicious of autoimmune disorders of SLE and APS. Ac- cordingly, concurrent with a uterine mass tissue biopsy, we conducted other workups for thrombocytopenia.

She presented no history of dyspnoea, chest pain, or bleeding. The PT time was normal; subsequently, we did not work her up for pulmonary emboli or disseminated intravascular coagulation.

She fulfilled the hematologic and immunologic components of lupus criteria [8]. Furthermore, according to the 2012 SLICC SLE criteria, she had 1 clinical and 3 immunologic criteria [8]. The SLEDAI score was equal to three. A family history of SLE is a valuable guiding clue [1], she also reported a personal history of autoimmune disease (hypothyroidism).

Rare leiomyoma could release anti-platelet antibodies and induce immune thrombocytopenia [9]. Sever hypothyroidism could also cause thrombocytopenia [10]; however, she was euthyroid with pharmacologic treatment. Abdominal mass's compressive effect and APS were the main predisposing factors for lower limb DVT in this patient.

We considered treating her with intravenous immunoglobulin, glucocorticoid (prednisolone 60 mg/day), hydroxychloroquine, and therapeutic doses of heparin. Our diagnosis was SLE/APS and the immunologic cause is suspicious in these diseases. IVIG inhibits Fc-receptor-mediated platelet phagocytosis and suppresses anti-platelet antibody production [11].

The key point, in this case, is that, in young women with thrombosis and thrombocytopenia (besides another important diagnosis), we should also consider SLE and APS. Accordingly, it is recommended to conduct meticulous personal and family history taking and physical examinations, and obtain proper laboratory data.

This case was complicated because she had severe thrombocytopenia with DVT and tumor, so anticoagulation was complicated. Additionally, without another symptom, the immunologic criteria for APS/SLE were positive in the reported patient.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles are considered in this article.

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Conflict of interest

The authors declered no conflict of interest.

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