

SLE with Haemoregic Cyst**Chandra Shekhar Jaiswal¹, Manoo Aggarwal², Atul Kumar³, Dinesh Kumar Thakur⁴**¹Assistant Professor, Department of Skin VD and Leprosy CIMS, Chhindwara.²Senior Resident, Department of Obs. & Gynae Swami Dayanand General Hospital, Shahdara Delhi³Associate Professor, Department of Surgery Government Medical College, Ratlam.⁴Assistant Professor, Department of Medicine CIMS, Chhindwara.

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Corresponding author: Dr Dinesh Kumar Thakur

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Abstract

Various cases of anaemia presenting with positive ANA were studied and a case series of such cases made and studied. SLE may be present along with persistent anemia in the presence of positive ANA for prolonged periods of time. It is recommended that patients with a positive ANA and persistent anemia serially be looked for clinical and laboratory manifestations of SLE in other organs.

Keywords: Hemorrhagic, ovarian cysts, lupus erythematosus, systemic, thrombocytopenia.

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Introduction

Systemic lupus erythematosus (SLE) is a chronic inflammatory and multisystem disease of unknown cause. SLE is a chronic autoimmune disease that has multi systemic manifestation as well as skin manifestations which follows a relapsing and remitting form. It is characterized by presence of antibodies to nuclear and cytoplasmic antigens. Skin lesions present as multiple maculopapular rashes all over body. Hematologic abnormalities are commonly present in patients with SLE. The major hematologic findings are anemia, lymphopenia, thrombocytopenia and the antiphospholipid syndrome.

The mechanism of immune thrombocytopenia in SLE is binding of antibodies to platelets followed by phagocytosis in the spleen. Idiopathic thrombocytopenic purpura (ITP) may be the first manifestation of adults with SLE, 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. 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. 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. Corticosteroids are the main stay of treatment for immune thrombocytopenia in patients with SLE.

Case 1: A 22 year old female presented with complains of malar rash and erythematous macular rashes all over the body for 5 months, periorbital oedema over the face for 15 days and erosions in the oral cavity for 8 days. She also complained of pain in lower abdomen for 5 days. She was positive for serum ANA factor along with increased LDH and CPK-MB levels. The patient had persistent low hemoglobin despite hematinic therapy. There was tenderness in lower abdomen left side. USG showed a cystic lesion with multiple internal echoes noted, suggestive of hemorrhagic cyst. The patient was then started on i.v pulse methyl prednisolone therapy along with blood transfusion as per physician advice following which patient started improving. The ovarian cyst was managed conservatively.

Case 2: A case of ruptured hemorrhagic corpus luteum as a presenting symptom of systemic lupus erythematosus. A young female 26 years presented to casualty with ruptured hemorrhagic corpus luteum (RHCL). Her workup revealed a new diagnosis of SLE with nephritis and positive lupus anticoagulant (LAC) test without thrombocytopenia. Nonsurgical treatment over laparotomy was chosen, and the patient was managed successfully with parenteral steroids and blood transfusion. In young women presenting with massive hemoperitoneum, undiagnosed immune thrombocytopenia should be considered as an etiology. In absence of any identifiable source of hemoperitoneum, these patients may not require laparotomy and treatment with intravenous corticosteroid and platelet transfusions may be preferable and lifesaving.

Case 3: Similar case of a 23-year-old subject who presented with a RHCL that was found to be the presenting symptom of SLE; the patient presented with severe anemia (Hb 6.7 g/dl) and thrombocytopenia (10,000/ml). A 24-year-old woman with a 9-year history of SLE, on corticosteroids, presented with severe abdominal pain, fever, vomiting and diarrhea for 1 day. Transabdominal sonography and abdominal computed tomography showed a right-sided well-defined pelvic mass. Laboratory study revealed leukocytosis. An emergency laparotomy revealed a right ovarian mass with purulent exudate. A right partial oophorectomy was performed. Pathologic examination revealed a hemorrhagic ovarian cyst with focal abscess.

Discussion

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. Valizadeh N et al reported large ovarian hemorrhagic cyst and immune thrombocytopenia as early manifestations of systemic lupus erythematosus (SLE). 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.

Feldman I et al studied a case of ruptured hemorrhagic corpus luteum as a presenting symptom of systemic lupus erythematosus. A young female presented to the emergency room with ruptured hemorrhagic corpus luteum (RHCL). Her workup revealed a new diagnosis of SLE with nephritis and positive lupus anticoagulant (LAC) test without thrombocytopenia. Authors reviewed the literature and found one similar case of a 23-year-old subject who presented with a RHCL that was found to be the presenting symptom of SLE; unlike the current case, the patient presented with severe anemia (Hb 6.7 g/dl) and thrombocytopenia (10,000/ml).

Karadeniz O et al studied Pseudo-pseudo Meigs' syndrome presenting as an acute surgical abdomen. Pseudo-pseudo-Meigs' syndrome is a rare entity of systemic lupus erythematosus, which is defined with the combination of pleural effusion, elevated serum CA-125 levels, and as cited. It has similar clinical aspects with gynecological malignancies which may

lead gynecologists to perform unnecessary surgeries and lab workouts. This review seeks to point out the importance of diagnosing pseudo-pseudo Meigs' syndrome (PPMS) and endeavors to inform gynecologists about the differential diagnoses. PPMS is one of the disregarding entities where gynecologists do not consider it as a differential diagnosis in case of less awareness. When a gynecologist evaluates ascites manifesting as acute abdomen; ectopic pregnancy, ovarian hyperstimulation syndrome, cyst rupture, and malignancy come to mind first. Four-thirds of those patients evaluated by gynecologists were performed hysterectomy leading to unnecessary interventions and economic burden on the health system.

Singh N, Tripathi R et al dealt with a massive spontaneous intraperitoneal hemorrhage in a young female with chronic immune thrombocytopenic purpura masquerading as ruptured ovarian cyst. They chose nonsurgical treatment over laparotomy, and the patient was managed successfully with parenteral steroids and platelet transfusion. In young women presenting with massive hemoperitoneum, undiagnosed immune thrombocytopenia should be considered as an etiology. In absence of any identifiable source of hemoperitoneum, these patients may not require laparotomy and treatment with intravenous corticosteroid and platelet transfusions may be preferable and lifesaving.

Burlui A et al studied multiple risk factors for thromboembolism and pregnancy loss in Systemic Lupus Erythematosus. Systemic Lupus Erythematosus (SLE) is known to pose difficulties when pregnancy is desired. Pregnancy in patients with SLE remains a high-risk situation with an increased frequency of fetal loss, preeclampsia, growth restriction and prematurity. Cases of young female SLE patients positive for the antiphospholipid syndrome remain challenging for clinicians, especially when pregnancy is desired.

Lin YH et al presented a case report of ovarian abscess caused by non-typhoidal Salmonella in a woman with systemic lupus erythematosus. A 24-year-old woman with a 9-year history of SLE, on corticosteroids, presented with severe abdominal pain, fever, vomiting and diarrhea for 1 day. Transabdominal sonography and abdominal computed tomography showed a right-sided well-defined pelvic mass. Laboratory study revealed leukocytosis. An emergency laparotomy revealed a right ovarian mass with purulent exudate. A right partial oophorectomy was performed. Pathologic examination revealed a hemorrhagic ovarian cyst with focal abscess. Patients with SLE are at risk of Salmonella infection. Gynecologists should be aware of the possibility of an ovarian abscess caused by Salmonella in such patients presenting with a pelvic mass and fever.

Al Hammadi N studied a case of new-onset systemic lupus erythematosus presenting with pseudo-pseudo-Meigs' syndrome and after extensive workup to rule out other possibilities like infection and malignancy, she was found to have systemic lupus erythematosus. She responded to prednisone and Immune suppression therapy (including mycophenolate mofetil (MMF) and Plaquenil). Pseudo-pseudo Meigs' syndrome is a diagnosis of exclusion. Ruling out malignancy is crucial. It usually presents with serous effusion and elevated serum CA 125 after ruling out malignancies and infections. Our patient had an extensive workup that included multiple radiographic imaging, fluid analysis, and biopsies to rule out that possibility.

Guballa N et al studied ovulation induction and in vitro fertilization in systemic lupus erythematosus and antiphospholipid syndrome. During ovulation induction (OI), ovarian stimulation is accomplished by hormonal manipulation, which includes administration of gonadotropins, gonadotropin-releasing hormone agonists, follicle-stimulating hormone, and luteinizing hormone. In in vitro fertilization (IVF), progesterone is often added. Because of the possibility of hormone-associated flare or thrombosis, patients with systemic lupus erythematosus (SLE) and primary antiphospholipid syndrome (primary APS) undergoing OI/IVF are potentially at increased risk. Postpartum complications included nephritis flare, costochondritis, and suicidal depression. It was concluded that although OI/IVF can be successful in SLE and primary APS patients, rates of fetal and maternal complications are high

Conclusion

SLE may be present along with persistent anemia in the presence of positive ANA for prolonged periods of time. It is recommended that patients with a positive ANA and persistent anemia serially be looked for clinical and laboratory manifestations of SLE in other organs.

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