New-onset systemic lupus erythematosus presenting with pseudo-pseudo Meigs’ syndrome

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Abstract

Systemic lupus erythematosus is a complex disease and can present with almost any organ involvement, including serosal inflammation. Our patient is not known to have any medical problems, presented for the first time with pseudo-pseudo Meigs’ syndrome (PPMS), and after extensive workup to rule out other possibilities like infection and malignancy, she was found to have systemic lupus erythematosus. Several other cases have been reported in the literature; our patient had to have a pleural biopsy for completion of workup. She responded to prednisone and Immune suppression therapy (including mycophenolate mofetil (MMF) and Plaquenil).

Keywords: Inflammation, pseudo-pseudo Meigs’ syndrome, systemic lupus

Introduction

Tjalma was the first to describe Pseudo-pseudo Meigs entity; hence the name Tjalma syndrome.[1] The diagnosis is made based on the following criteria: pleural effusion, ascites, and elevated CA-125 after ruling out benign or malignant tumors. Polyserositis is common in systemic lupus. In one report,[2] the prevalence was about 17.9%.

There is a specific differential diagnosis when we approach a patient with generalized anasarca, like cases of decreased oncotic pressure or elevated hydrostatic pressure; systemic rheumatological disease is not one of them.

The number of pseudo-pseudo Meigs syndrome is on the rise especially as a presenting feature of Systemic Lupus Erythematosus and other connective tissue diseases.

Our case report aims to increase the awareness of this entity and its association with Systemic rheumatological diseases. We also aim to encourage early referral to a rheumatologist after ruling out malignancy to establish an early diagnosis and avoid the complication of delayed management.

Case Report

A 41-years-old female presented to the hospital complaining of progressive shortness of breath in January 2018.

She denied any history of orthopnea or PND. No past medical problems or history of medication use. She was not associated with urinary problems, skin rash, Raynaud’s, bald spots, discoid rash, sicca features, Central nervous system, hematological, or kidney disease. She also denied a history of Diarrhea, abdominal pain, or lower limb swelling. No history of constitutional symptoms. She is a, not married, with no children. There was no family history of similar or other problems. She does not consume alcohol or smoke cigarettes.

On Examination

She generally looked well. Her blood pressure was 120/80. Chest examination revealed decreased air entry on the right side with

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decreased vocal resonance. She also had massive abdominal distention and positive fluid thrill. There was no lower limb swelling or periorbital edema; no active arthritis, rashes, or bald spots. On mouth examination, no ulcers with adequate salivary pool were found.

Investigations revealed low hemoglobin 98 mg/dl, MCV was 92 fl (normocytic normochromic anemia), platelets were 426 $10^9$/L, and normal white cell count. Urinalysis was negative for blood and protein with a negative protein to creatinine ratio. Renal function test showed creatinine of 61 umol/L. The hepatic profile showed an alkaline phosphatase of 80 U/L; Alanine transaminase (ALT) was 14 U/L.

Creatine kinase (CK) was 74 U/L. Serum Albumin level was 38 g/L. We also requested serum immunoglobulin level, which was unremarkable. Inflammatory markers screening showed an erythrocyte sedimentation rate (ESR) of 31 mm/h with a normal C-reactive protein (CRP).

The autoimmune profile revealed an antinuclear antibody (ANA) of 1:640 (+); rest, including rheumatoid factor, extractable nuclear antigen antibodies (ENA), and Antiphospholipid serology, were negative except for Anti Chromatin which was more than 8. Complement levels: C3 0.85 g/L (low) and C4 0.3 (normal) g/L.

Plain chest radiograph and abdominal Ultrasound showed Rt-sided pleural effusion and ascites. Abdominal paracentesis showed an exudative effusion with negative cytology. Cultures and acid-fast bacillus (AFB) staining were negative.

Pleural fluid analysis was primarily exudative effusion which was also negative for both cytology and cultures as well as tuberculosis (TB). ANA of pleural fluid was positive. Screening for tumor markers revealed an elevated cancer antigen 125 (CA-125) level 121 U/ml. Computed tomography (CT) of the chest showed multiple black pleural nodules, which were thought to be malignant and the cause of effusion. For that reason, a pleural biopsy was done and showed benign cytology with no malignant cells.

CT of the abdomen and the pelvis showed a large bulky uterus of 20 × 18 × 10 cm (which raised the possibility of uterine fibroid). The right ovary was 4.9 × 4 × 3 cm, with a partially collapsing hemorrhagic cyst of 3.8 cm, the left ovary was 2 × 2.9 × 1.8 cm. There was no radiographic evidence of peritoneal nodularity enhancement or thickening.

Obstetrics and gynecology were consulted for evaluation of a possible uterine malignancy. After extensive evaluation, she was found to have uterine fibroid, which contributed to her anemia. After surgical removal of the fibroid (total abdominal hysterectomy with a bilateral salpingectomy), anemia significantly improved. She tolerated surgery very well with no complications. Her hemoglobin went back to normal, and the iron store stabilized.

After extensive investigation and after ruling out malignancy, the diagnosis of SLE presenting with pseudo-pseudo Meigs’s syndrome (PPMS) was made. The patient was started on methylprednisolone 500 mg for three days; then she was switched to regular oral prednisone. We added Plaquenil and MMF (Mycophenolate mofetil) afterward as a steroid-sparing agent. Her ascites subsequently improved during her hospital stay, and she was discharged. Two months later, there was no evidence of pleural effusion or ascites on examination. We started to taper off steroids gradually to a dose of 10 mg daily with plaquenil and MMF 2 gm daily. Unfortunately, she presented again with pleural effusion, so steroids were increased, and MMF was maximized with a plan to switch to either cyclophosphamide or rituximab if reaccumulated.

Discussion

There are several cases of pseudo-pseudo Meigs’s syndrome in lupus patients, some as initial manifestations of lupus and others in patients with an established diagnosis. Interestingly, it can present in a patient with mixed connective tissue disease as well.

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.

In lupus, we treat according to the organ involved; our patient responded very well to high doses of steroids and traditional therapy of major organ involvement.

PPMS is increasingly reported in the literature, especially in patients with autoimmune diseases like systemic lupus erythematosus (SLE). Ruling out malignancies and infections is crucial. Interestingly as in our report and several others, it can be the presenting manifestations, even in the absence of a prior known diagnosis of lupus. Systemic lupus is a complex autoimmune disease that can affect almost any organ, and these entities are reported more frequently, which might raise the question for future larger studies to assess the nature, pathogenesis, and management.

Our patient is the first to have a pleural biopsy as part of a diagnostic workup. After extensive evaluation, multiple ER visits for pleural fluid aspiration as well as multiple radiographic images, she was found to have Systemic Lupus Erythematosus based on serositis and positive serology. This entity is increasingly reported in the literature as a presenting manifestation of SLE. Despite the fact that it is not part of diagnostic criteria, it is still a valid differential in a patient presenting with serous cavity effusion and elevated CA 125 after ruling out malignancy. Our patient responded very well to prednisone and immune suppression therapy (including MMF and plaquenil).

More studies are needed to explore the pathogenesis and therapeutic options for pseudo-pseudo Meigs’ in the context of Autoimmune rheumatological diseases.
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Conflicts of interest

There are no conflicts of interest.

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