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SYSTEMIC LUPUS ERYTHEMATOSUS WITH MULTISYSTEM INVOLVEMENT: A DETAILED CASE REPORT

General Medicine

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ABSTRACT

Reports of sudden and potentially fatal presentations in connective tissue disorders (CTDs) are uncommon. Early identification and targeted treatment can significantly alter disease progression. Systemic lupus erythematosus (SLE) is an inflammatory multisystem disease that can involve various organs and systems, including the lungs, kidneys, and hematologic system.

KEYWORDS

Systemic Lupus Erythematosus (SLE)

INTRODUCTION:

Systemic lupus erythematosus (SLE) is an inflammatory connective tissue disease that primarily affects women. Antibodies (anti-nuclear, anti-dsDNA, anti-histone) and immune complexes play a role in the pathophysiology of SLE, causing damage to various tissues and organs.

Case Presentation:

A 41-year-old female patient with a known history of hypothyroidism presented with a 48-hour history of progressive dyspnea, non-productive cough, and fever persisting on and off for 20 days. She also reported fatigue, myalgia, arthralgia, oral ulcers, and photosensitivity for three months. On examination, the patient appeared very ill, with tachycardia, afebrile, tachypnea, and a peripheral oxygen saturation of 94% on room air, necessitating oxygen support. Her blood pressure was 120/80 mmHg, pulse rate 100 bpm, and she had bilateral non-pitting pedal edema and an erythematous rash over her cheeks. Auscultation revealed diffuse coarse crackles in both lungs. Abdominal examination showed mild hepatomegaly. Electrocardiogram showed no significant abnormalities.

HRCT revealed patchy consolidations, interlobular septal thickening, and peribronchial wall thickening in bilateral lung fields, minimal bilateral pleural effusion, minimal pericardial effusion, and cardiomegaly. Chest X-ray confirmed cardiomegaly and mild bilateral pleural effusion. Blood and urine cultures were sent, and empirical treatment with antibiotics, IV diuretics, and other supportive medications was initiated. 2-D echocardiography showed mild pericardial effusion without signs of tamponade, warranting only monitoring as advised by the consulting cardiologist. Multislice CT pulmonary angiogram ruled out pulmonary artery thrombosis and confirmed minimal bilateral pleural effusion, cardiomegaly, minimal pericardial effusion, and patchy consolidation. Abdominal ultrasound showed hepatosplenomegaly.

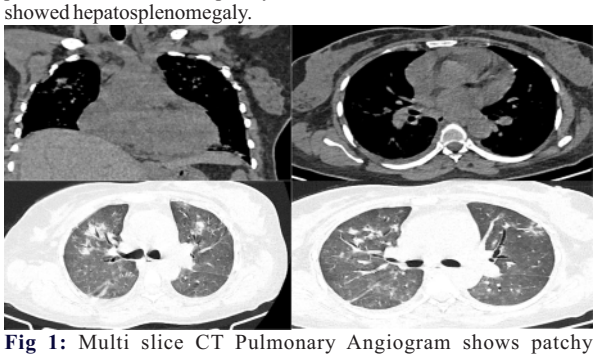

Fig 1: Multi slice CT Pulmonary Angiogram shows patchy consolidation, Minimal bilateral pleural effusion, Cardiomegaly, Minimal pericardial effusion.

Laboratory results revealed WBC count of 2800 cells/cumm, Hb 6.5 g/dL, RBC 2.50, platelet count 1.10 lakh/cumm, ESR 140 mm/hr, CRP 2.5, SGOT 50 IU/L, SGPT 31 IU/L, serum albumin 2.4 g/dL, urea 86 mg/dL, creatinine 2.3 mg/dL, sodium 137 meq/L, potassium 3.5 meq/L, urine albumin +2, RBC 20-22, PCT 0.32, ANA positive, and ANA profile positive for anti-SSA, anti-SM, RNP, Ro52, and dsDNA-19. A 24-hour urine collection demonstrated 3 g proteinuria. Based on a rheumatologist's recommendation, IV methylprednisolone 500 mg/day for three days was initiated, and blood parameters were corrected. The patient was discharged on the 10th hospital day with a therapy regimen of 60 mg/day prednisone, 200 mg/day hydroxychloroquine, and 500 mg/day mycophenolate mofetil, with follow-up arrangements for a renal biopsy at the nephrology clinic.

DISCUSSION:

This case highlights the complexity and severity of systemic lupus erythematosus (SLE) and its potential for acute, life-threatening presentations. The 41-year-old female patient exhibited a wide range of symptoms and clinical findings that underscored the multisystemic nature of SLE. Her history of hypothyroidism.

The patient presented with progressive dyspnea, non-productive cough, fever, fatigue, myalgia, arthralgia, oral ulcers, and photosensitivity, which are all indicative of an active SLE flare. Her clinical examination revealed significant findings such as tachycardia, tachypnea, bilateral non-pitting pedal edema, and an erythematous malar rash, suggesting systemic involvement. Laboratory results further supported the diagnosis, with positive ANA, anti-SSA, anti-SM, RNP, Ro52, and dsDNA antibodies, along with significant proteinuria.

Imaging studies, including HRCT and chest X-ray, showed patchy consolidations, interlobular septal thickening, peribronchial wall thickening, pleural and pericardial effusions, and cardiomegaly. These findings indicated widespread inflammation and fluid accumulation, common in SLE patients with active disease.

The management of this patient involved a multidisciplinary approach, including empirical treatment with antibiotics and IV diuretics to address potential sepsis and supportive care measures. The decision to initiate high-dose IV methylprednisolone was crucial in controlling the acute inflammatory process and preventing further organ damage. The differential diagnosis was broad, including sepsis and catastrophic antiphospholipid syndrome, both of which can present with similar clinical features. The aggressive initial treatment with immunosuppressants was justified given the severity of the presentation and the potential for rapid deterioration.

This case underscores the importance of early recognition and comprehensive management of acute SLE flares. It also highlights the

challenges in distinguishing between sepsis and SLE exacerbations in emergency settings. The use of glucocorticoids was pivotal in managing the active disease and preventing further complications.

The patient's discharge on a regimen of prednisone, hydroxychloroquine, and mycophenolate mofetil, along with follow-up for a renal biopsy, reflects a tailored approach to long-term management and monitoring of SLE. The outcome underscores the effectiveness of prompt and adequate immunosuppression in controlling severe SLE manifestations

CONCLUSION:

The combination of anamnestic characteristics with the malar rash, polyserositis, and positive immunology met the criteria for an SLE diagnosis. The patient exhibited a highly dynamic clinical course. The short time span between the patient's initial need for medical attention and the voluntary cessation of medication suggested either a real flare or the first full presentation of SLE. Given the initial focal urologic complaints, evidence of multiorgan dysfunction, and a picture resembling distributive shock, this explosive clinical presentation warranted a low threshold for sepsis and empirical antimicrobial treatment until infection was definitively ruled out. The aggressive initial measures were justified by the understanding that prognosis in sepsis varies with the timeliness and aggressiveness of intervention. Differentiating between sepsis and an acute SLE episode in such emergency scenarios is challenging. In this patient, catastrophic antiphospholipid syndrome was also considered due to similar clinical presentations seen in young women with the condition. Complete immunosuppression with glucocorticoids was critical in managing the active SLE and preventing potentially harmful outcomes. The question of whether this episode was an actual flare-up or the full manifestation of developing lupus remains unanswered. Treating connective tissue disease and its complications with organized reasoning and careful consideration is crucial in urgent clinical scenarios. Acute, multisystemic, life-threatening disorders in the emergency room should prompt consideration of autoimmune diseases like SLE. The complex clinical and laboratory expression of SLE is driven by circulating immune complexes and autoantibodies with diverse specificities that initiate a comprehensive inflammatory response. Adequate, early, and complete immunosuppression can effectively manage the various manifestations, despite their severe and alarming nature.

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