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SYSTEMIC LUPUS ERYTHREMATOUS INDUCED PULMONARY FIBROSIS AND PANCREATITIS: A CASE REVIEW

Dermatology

Vishnu Das Clinical Pharmacist, Apollo Main Hospital, Chennai, India- 600006

ABSTRACT

Systemic lupus erythrematous is an auto immune disease in which organ and cells undergo damage initially mediated by tissue binding auto antibodies and immune complexes. It is known to affect women more frequently than men, with a ratio of approximately six women to every one man. here was worldwide variation, with the highest incidence reported in North America This 23 year old lady, a known case of SLE (on treatment with immunosuppressants), Emergency with low blood pressure of 90/60mmHg and tachycardia, 118/min. on physical examination, she had severe left hypochondriac tenderness.

KEYWORDS

Immunosuppressants, Hypochondriac, Pericaditis

INTRODUCTION

Systemic Lupus Erythrematous is a chronic, inflammatory, variable autoimmune disease of connective tissue that occurs chiefly in women and is typically characterized by fever, skin rash, fatigue, and joint pain and often by disorders of the blood, kidneys, heart, lungs, and brain (such as hemolytic anemia, nephritis, pleurisy, pericarditis, cognitive dysfunction, or meningitis).[1] It is known to affect women more frequently than men, with a ratio of approximately six women to every one man. here was worldwide variation, with the highest incidence reported in North America (23.2/100 000 person-years, 95% CI: 22.4, 24.0) and the lowest incidences reported in Africa (0.3/100 000 person-years).[4]

CASE PRESENTATION

This 23 year old lady, a known case of SLE (on treatment with immunosuppressants), admitted with complaints of abdominal pain, in left hypochondriac region, which started 4 days back. Pain was progressively increasing in nature and radiated to her back. Pain increased in intensity in lying down position and after eating food. She prefers sitting position.

She was admitted to the Emergency with low blood pressure of 90/60mmHg and tachycardia, 118/min. on physical examination, she had severe left hypochondriac tenderness. While asking to the patient it was found that her father have SLE. She was managed with adequate analgesia and IV fluids and was shifted to ward for further management.

Laboratory investigation was done. Her hematologic parameters has showed anemia, 9.7g/dl, along with high ferritin level of 3515ng/ml, elevated C-reactive protein level of 33.6mg/dl and raised Erythrocyte Sedimentation rate, 88mm/hr. Liver Function test showed that her alkaline phosphatase -310U/L, Alanine Aminotransferase (ALT)-131U/L, Gamma Glutaryl Transpeptidase- 124U/L and Lipase- 824 U/L are elevated her albumin level is in the normal range of 3.8g/dl. Her Renal Function parameters such as urea, 14mg/dl and serum creatinine, 0.4mg/dl were found to be normal.

Two sets of blood cultures were sent and both showed no growth and the samples from anterior segment of left lower lobe and other segment were sent for bacteriology pack and X-PERT MTB, which were all negative. Pleural fluid protein levels were 4.5g/dL and glucose was 111mg/dL. Pathologist was consulted, who reconfirmed that AFB and Fungal stains from lung lesions were negative.

Chest X-Ray showed ill defined patchy opacities in left lung lower zone. 320 Slice CT W/A with triphasic protocol was done and it showed findings suggestive of mild cholangitis with left pleuritis, left lingular consolidation and mild pericarditis with normal kidneys. Echocardiography was done and it was normal with EF 60%.Histopathology Examination of the tissue showed variable airspace organization (organizing pneumonia pattern) with intra-alveolar hemorrhage, interstitial chronic inflammation and patchy (multifocal) interstitial fibrosis.

These finding raises the possibility of Non Specific Interstitial Pneumonia pattern secondary to SLE. Her repeat Serum lipase level i.e. 221U/L was found to be in decreasing trend. She is abdominal pain was gone, but she started experiencing left sided pleuritic type of pain, most likely due to the left pleural effusion. Interventional Radiologist team did left side pleural therapeutic and diagnostic tapping, about 500ml of fluid was aspirated and samples were sent for various tests. Pleural fluid cytology, X-pert MTB, ADA and LDH were normal. Pleural fluid protein levels were 4.5g/dL and glucose was 111mg/dL.Bilateral lung parenchyma ground glass opacities with septal thickening and suggestion of fine nodules associated patchy area of dense consolidations.

Mild degree of associated bronchiolitis possible in view of small centrilobular nodules.Hepatomegaly with small hyper dense lesions in the VIII and II segment of liver.Non specific Bulky pancreas with mild fat stranding correlate with amylase / lipase, to rule out interstitial pancreatitis. MRI shows Bilateral sacroilitis - possibility of seronegative spondyloarthropathy to be excluded.

She is discharged with the following advice, Tab. Prednisolone 30mg po 1-0-0 after food till review, Tab. Rifaximin 400mg po 1-0-1 after food till review, Syp. Lactulose 20 ml 0-0-1 sos if constipation present.

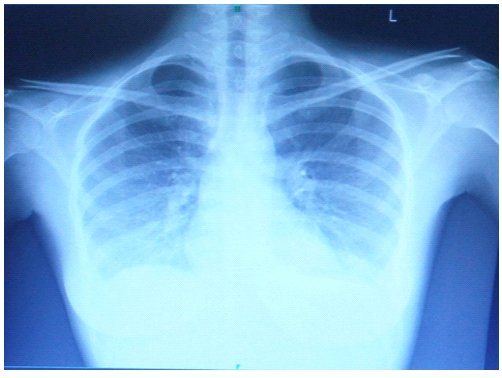

Figure 1: X ray image of ill defined patchy opacities in left lower lobe.

DISCUSSION

Systemic lupus erythrematous is an auto immune disease in which organ and cells undergo damage initially mediated by tissue binding auto antibodies and immune complexes. In most patients auto antibodies are present for a few years before the initial clinical symptoms appear, clinical manifestations are heterogeneous.[1]

Pancreatitis in the setting of SLE has an estimated annual incidence of 0.4–1.1 per 1000 patients. Patients are typically female, with clinically active SLE developing within the first 2 years of disease onset.

Interaction between susceptible genes and environmental factors triggers abnormal immune response which varies among the individual. The response may include

1. Activation of innate immunity by CpG DNA, DNA in immune

complexes, viral RNA/RNA protein self antigens.
2. Lowered activation thresholds and abnormal activation pathways in adaptive immunity cells(T and B lymphocytes).
3. In effective regulatory CD4+ and CD8+ Tcells
4. Reduced clearance of immune complexes and apoptotic cells

The self antigens are available for the recognition of immune system in surface blebs of apoptotic cells, thus antigen, antibody and and immune complexes persist for prolonged period of time causing inflammation and disease development. Immune cell activation is followed by the production of pro inflammatory type I and II interferon's, tumour necrosis α and interleukin, IL-17 and IL-10. These result in the production of auto antibody and immune complexes. And these cause the release of cytokines, vasoactive peptides, oxidants and destructive enzymes. As a result of the chronic inflammation followed by accumulation of growth factor contribute to irreversible tissue damage including fibrosis, sclerosis in lungs, glomerulus, pancreas and other tissues.[1]

Some drugs also act as triggering factor, include
- Sulfa drugs, which make a person more sensitive to the sun, such as: Trimethoprim-sulfamethoxazole, sulfisoxazoletolbutamide, sulfasalazine.
- Sun-sensitizing tetracycline drugs such as minocycline.
- Penicillin or other antibiotic drugs such as amoxicillin,ampicillin, cloxacillin [2]

Fatigue, fever, arthralgia, and weight changes are the most common symptoms in new cases or recurrent active SLE flares. Fatigue, the most common constitutional symptom associated with SLE, can be due to active SLE, medications, lifestyle habits, or concomitant fibromyalgia or affective disorders. Pancreatitis is an uncommon initial presentation of SLE and occurs in approximately 5% of patients with SLE, often secondary to vasculitis. [3] Patient presented with abdominal pain (88%), followed by nausea and/or vomiting (67%). Pancreatitis was diagnosed on laboratory evidence of elevated serum amylase or lipase in 97% of patients. .The most common pulmonary manifestation of SLE is pleuritis with or without pleural effusion. And the life threatening manifestations include interstitial inflammation leading to fibrosis, shrinking lung syndrome and intra-alveolar hemorrhage. The active presence of lupus symptoms among pancreatitis patients considerably increases mortality risk. One series reported no mortality events among patients without SLE symptoms at the onset of pancreatitis, compared to 40% mortality among those with SLE manifestations.[5]

In 2007, the European League Against Rheumatism (EULAR) released recommendations for the treatment of SLE.In patients with SLE without major organ manifestations, glucocorticoids and antimalarial agents may be beneficial. NSAIDs may be used for short periods in patients at low risk for complications from these drugs. Consider immunosuppressive agents (eg, azathioprine, mycophenolatemofetil, methotrexate) in refractory cases or when steroid doses cannot be reduced to levels for long-term use. [4]

CONCLUSION

Pulmonary fibrosis and Pancreatitis are the rare complications of Systemic Lupus Erythrematous, which increases the morbidity of the patients. The female patient with familial history of the disease which require steroid therapy for the reliving of the symptoms and it should be monitored regularly for preventing the complications irreversible.

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