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Case Report

Systemic Lupus Erythematosus Patient Presenting With Acute Pulmonary Edema as the Initial Hospital Presentation

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ABSTRACT

Systemic lupus erythematosus (SLE) is a chronic inflammatory disease that occurs more commonly in women in the age group between 16 to 60 years. SLE most commonly affects skin but heart and cardiopulmonary circulation can also be significantly affected. [1] Acute non cardiogenic pulmonary edema is a rare initial presentation of SLE. [2]

Key words: systemic lupus erethymetosus, pulmonary edema, non cardiogenic.

Key message: SLE is a rare cause of non cardiogenic pulmonary edema. Patient of SLE presenting as pulmonary edema as initial hospital presentation is a very rare phenomenon. Such situations can me managed efficiently if diagnosis is made on time.

CASE REPORT

A 48 year old women, presented with history of breathlessness (of NYHA class 3), since 7 days and generalized weakness and easy fatigability since 5 days. Patient had past history of arthritis and erythematous butterfly shaped rash over the face since a year, and for which she was receiving symptomatic treatment from an Ayurvedic doctor. No history of fever, palpitations or chest pain. She had no past history of hypertension, diabetes mellitus or ischemic heart disease.

On physical examination at the time of admission– patient was well built and well nourished, conscious, cooperative and oriented with time, place, and person; heart rate- 110/min, Blood pressure- 140/90 mmHg (measured on both the arms in supine position), respiratory rate of 30 per minute and was using accessory muscles of respiration. On local examination patient had pallor, oral ulcers, alopecia, and multiple hyperpigmented and butterfly shaped rashes over the face.

Systemic examination: CVS-on auscultation, heart sounds were normal, no murmurs appreciated. Respiratory system-bilateral coarse crepitations were heard all over the lung fields with more in the infraaxillary and infrascapular regions.

Investigations: Electrocardiogram was done and showed sinus tachycardia with no ST-T changes. The chest radiography showed cephalization of veins, with perihilar venous congestion, presence of lung parenchyma with diffuse interstitial edema and left ventricular hypertrophy.

Trans-thoracic echocardiogram was done on day 5 which showed concentric left ventricular hypertrophy, left ventricular ejection fraction of 54%, grade one diastolic dysfunction, no region wall motion abnormality, and no other significant abnormality.

Skin biopsy of hyperpigmented, butterfly rash over face was taken-

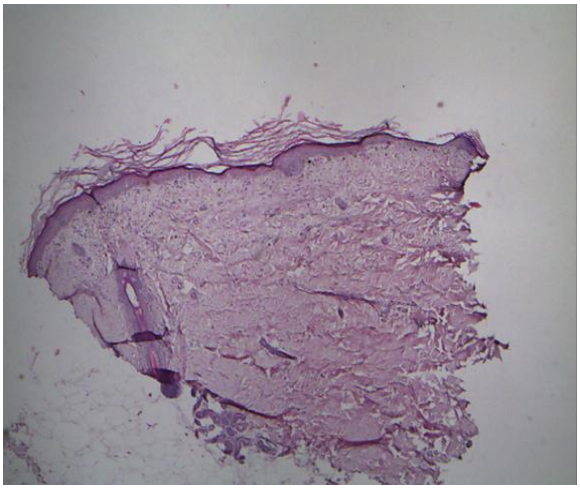
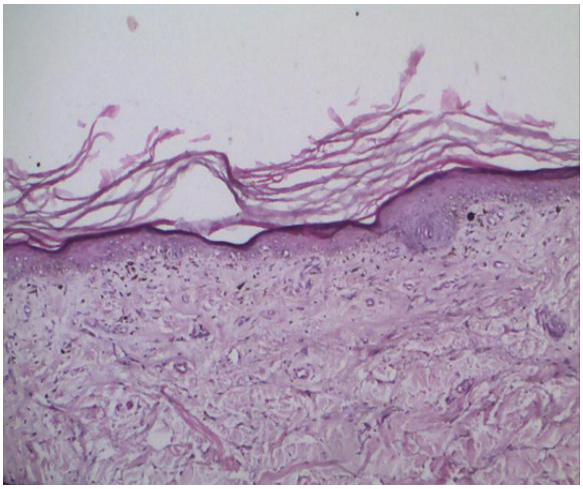

Figure 1 and 2

Impression: Acute Cutaneous Lupus Erythematosus

Laboratory tests favoured the hypothesis of active SLE; showing leucopenia (4000 leukocytes/mm^3), thrombocytopenia (platelets45000/mm^3), anemia (hemoglobin-8 mg/dl), direct coombs test- Positive, antinuclear antibodies (ANA) > 1:150, urine protein to creatinine ratio of 0.6, anti-dsDNA- Positive, erythrocyte sedimentation rate of 90 mm at the end of 1st hour, blood sugar (random)- 93mg/dl, serum creatinine 1.6 mg/dl.

Pulmonary function test: moderate restrictive lung disease FEV1/FVC % pred>95% FVC % Pred<64%.

Chest Xray: Before initiating treatment.

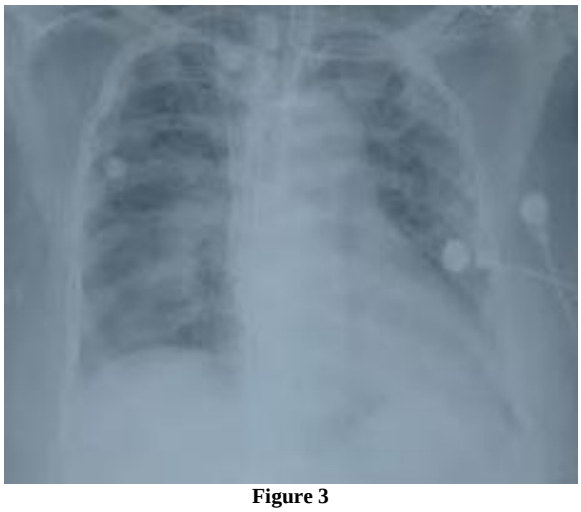

Figure 3

Chest Xray AP view: shows opacity in bilateral middle and lower zones, suggestive of pulmonary edema, with apparent cardiomegaly.

Patient was diagnosed with systemic lupus erythematosis with acute respiratory failure probably secondary to non cardiogenic pulmonary edema. Hence patient was intubated with endotracheal tube and was given mechanical ventilator support. Patient was managed with diuretics (furosemide), nitroglycerin, antibiotics and hydrocortisone 50 mg intravenous every 8 hourly for three consecutive days, azathioprine 50mg once a day. After four days the patient improved, was extubated and was given support of noninvasive ventilation intermittently. She remained afebrile, hemodynamically stable, preserved renal function, without any other complications.

Chest Xray: after 5 days of initiating treatment.

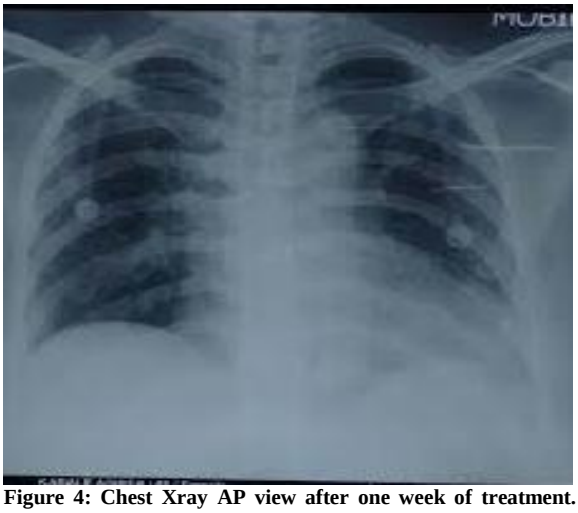

Figure 4: Chest Xray AP view after one week of treatment. Most of the infiltrates have resolved.

DISCUSSION

Based on the criteria of the *American College of Rheumatology* of 1997, [3] the diagnosis of SLE was made. It was established through the presence of acute cutaneous LE, oral ulcer, alopecia, anemia, positive direct coombs test, positive anti-ds DNA, positive antinuclear factor.

Pulmonary edema is accumulation of fluid in the air spaces and lung parenchyma. It leads to respiratory failure secondary to impaired gas exchange. [4] It is either due to left ventricular failure (known as cardiogenicpulmonary edema), or an injury to parenchyma or vasculature of the lung (known as noncardiogenic pulmonary edema). [4]

Pulmonary microvascular injury is caused either due to an initial and rapid increase in pulmonary vascular pressure either due to pulmonary vasoconstriction or increased pulmonary blood flow. An increase in vascular permeability results in edema formation (the blast theory). [5]

Systemic lupus erythematosus is a common chronic autoimmune disorder causing injury to many organ systems. Cardiac complications of lupus affect most parts of the heart. These include pericarditis, myocarditis, endocarditis and coronary artery disease. While many histopathological findings in lupus-related cardiac diseases are non-specific. [6,7]

Pathogenesis of Non cardiogenic pulmonary edema (NPE): Two major components contribute to it: elevated intravascular pressure and pulmonary capillary leak. [8,9] Therefore, hemodynamic cardiogenic and noncardiogenic components exist. These components often work in concert, as in pulmonary edema after epileptic convulsions or intracranial pressure elevation. The hemodynamic component is relatively brief and may unmask pure NPE, such as that seen in experimental seizures.

Diffuse pulmonary hemorrhage (DPH) is a syndrome characterized by the presence of widespread hemorrhage from the pulmonary microvasculature leading to hemoptysis, iron deficiency anemia, and a chest radiography showing bilateral airspace consolidation. Diagnostic imaging consists primarily of chest radiography, but CT and MR imaging may be helpful in selected cases. [10-12]

In order to avoid life-threatening complications, prompt recognition of NPE is important. The use of chest radiography and other tests is key to establishing the diagnosis and to distinguishing between the 2 types of pulmonary edema.

Our patient represented an interesting diagnostic challenge. She was an elderly woman who had predominantly bilateral radiographic abnormalities. Her initial manifestation of SLE was pulmonary edema with restrictive lung disease. Pulmonary embolism, cardiogenic pulmonary edema, and bleeding from coagulopathies were excluded.

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