CASE REPORT

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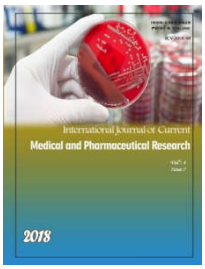

SYSTEMIC LUPUS ERYTHEMATOUS (SLE) WITH HYDRO PNEUMOTHORAX: A RARE CASE REPORT

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ABSTRACT

SLE an autoimmune disease, also known as lupus in which immune system affects healthy tissue of various regions of the body i.e. a multiorgan disease. Lung involvement is a common presentation in SLE; however life threatening pneumothorax is a rare occurrence in this disease. We describe a 26 year old young female diagnosed as SLE presenting with spontaneous pneumothorax. Intercostal tube was inserted and patient was given BIPAP ventilation and started on corticosteroids along with supportive treatment for underlying co-morbidities. Pneumothorax though not a common presentation in SLE can be life threatening with high mortality.

INTRODUCTION

Systemic lupus erythematous (SLE) is a multiorgan autoimmune disease involving the immune system affecting different parts of the body. Symptoms are as per the severity of disease ranging from fever, painful and swollen joints, chest pain, swollen lymph nodes, red rash on the face, mouth ulcers, hair loss, and fatigue. [1]. The disease is characterized by periods of flares and remission. Childbearing age group of women is at higher chance of developing SLE i.e. nine times more than men.[2]The presenting symptom in female is usually Reynaud's phenomenon, psychiatric symptoms and the white blood cell count is low, whereas males have seizures, kidney disease, serositis of lungs and heart, skin diseases, and peripheral neuropathy.[3] There is no cure for SLE.[1] Treatment may include NSAIDs, corticosteroids, immunosuppressant's, hydroxychlor oquine, and methotrexate. The most frequent complaint experienced in 90% of patients in due course of illness is pain of small joints especially of hand and wrist, though all joints are at equal risk. [4]

Case Report

A recently diagnosed 26 years old female of SLE with arthritis, lupus nephritis associated with co-morbidities diabetes mellitus and hypothyroidism presented to emergency room on 8.5.2018 with complaints of swelling over face & both lower limbs since 1 month shortness of breath and, dry cough since 4 days. There was no history of pulmonary tuberculosis or pneumonia.

On general examination, her higher functions were normal, respiratory rate was 30/minute, pulse 86/minute, regular, blood pressure 110/70mmHg, SPO2: 90% on room air. She had malar rash, discoid rash over the skin of upper & lower limbs and generalized anasarca. On respiratory system examination, air entry was absent on the right side with positive succussion splash. On cardiovascular examination, tachycardia was present, heart sounds were normal. Rest of the systemic examination was within normal limit. Her complete haemogram showed polymorphonuclear leukocytosis (84%), blood urea was raised (74mg/dl) with hyponatremia (120 mg/dl).Chest X-ray was suggestive of right sided hydropneumothorax (Legend1).HRCT scan of thorax was done on 2nd day post admission that was indicative of pneumothorax in right pleural cavity, collapse of right upper lobe, lateral segment of middle lobe & lateral basal segment of lower lobe of lung with intercostal drain in situ, pneumomediastinum and subcutaneous emphysema in anterior & lateral chest wall on right side & midline (Legend 2).She was diagnosed as class V Lupus Nephritis on kidney biopsy on previous admission. Patient was treated with intercostal drain (legend 3), corticosteroids, intermittent BIPAP and supportive therapy. However she succumbed to her disease on 20th day of admission.

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DISCUSSION

In 1982 the American College of Rheumatology (ACR) constituted criteria's for diagnosis of SLE, which were revised in 1997.In year 2012 the Systemic Lupus International Collaborating Clinics (SLICC) group revised and validated the ACR SLE eleven classification criteria [5]:

1. Malar rash
2. Red, scaly patches on skin causing scars known as Discoid rash
3. Serositis: Pleurisy or pericarditis
4. Oral ulcers
5. Arthritis: involvement of two or more peripheral joints characterised with tenderness, swelling, or effusion which are non-erosive.
6. Photosensitivity
7. On hematology SLE is usually associated with leukopenia <4000/µl, or low platelet count (<100000/µl), lymphopenia (<1500/µl), hemolytic anemia, in absence of drug causing it.
8. Renal disorder: Microscopic cellular casts observed in urine or more than 0.5 g per day protein in urine
9. Positive Antinuclear (ANA) test.
10. Anti-ds DNA, antiphospholipid antibody seen in 70% 0f patients, Positive anti-Smith, or false positive serological test for syphilis:Immunologic disorder
11. Neurologic disorder: Seizures or psychosis.

Besides these the patient may also have

- Fever
- Extreme fatigue
- Hair loss
- Reynaud's phenomenon i.e. on cold fingers discoloration to blue/white.

SLE may result in pleuritic pain and also one of the reasons for shrinking lung syndrome. Other associated lung conditions include pulmonary hemorrhage, pneumonitis, pulmonary emboli, pulmonary hypertension, and chronic diffuse interstitial lung disease. [6] Pneumothorax is rare in SLE, the cause may be because of 1) immune process of disease, 2) lung infections,3) glucocorticoids or cyclophosphamide therapy. Pneumothorax may occur as tissue may become fragile due to these drugs.[7]There are very few documented cases in literature showing association of pneumothorax/hydropneumothorax along with SLE.

Bilateral spontaneous pneumothorax incidence is 1–4%, as reported by Graf-Deuel and Knoblauch in their case series. Surgical intervention, especially with underlying pleuro-pulmonary disease, is strongly recommended for those cases, as they might fail the conservative management. [8] Sawkar and Easom [9], Richards *et al* [10], Masuda [11], have reported young female patient having SLE and recurrent pneumothorax. The renal involvement may be indicated by the presence of painless hematuria or proteinurea.This may result in acute or chronic renal damage developing in lupus nephritis resulting in acute or end stage renal failure. However, timely identification and management of SLE may prevent renal failure seen in less than 5%.[12]A case was reported by Wilhelm M *et al*[13] of Pneumothorax associated with nephritis in a patient with SLE, similar to our patient who was diagnosed with grade V lupus nephritis on renal biopsy.The international guidelines for the treatment of pneumothorax indicate the need for chest tube insertion if there is respiratory distress, hemodynamic compromise, or bilateral involvement. [8]Our patient was treated with chest tube drainage, corticosteroids & antibiotics. Yen JH *et al*[14] and Tanaka N *et al*[15] patients had received immunosuppressive/glucocorticoids therapy as ours. HRCT scan of our patient revealed right sided pneumothorax with pneumomediastum. R Maeda *et al* [16] in his patient of SLE with recurrent pneumothorax had multiple lung cysts.Paira S *et al* [17] reported a 36 year-old man with SLE, having recurrent pneumothorax complicated with pneumoperitoneum and mediastinal emphysema.

CONCLUSION

Pleuro-parenchymal manifestation in multiorgan SLE is very frequent, however spontaneous pneumothorax is very rare which is often life threatening. Our young female patient of SLE had hydropneumothorax which even on timely treatment proved to be fatal.

Figure Legends

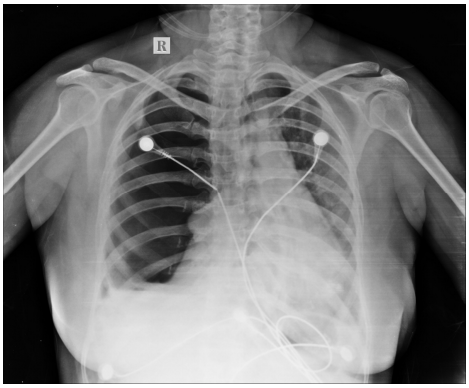

Figure 1 Chest X-ray PA view on admission

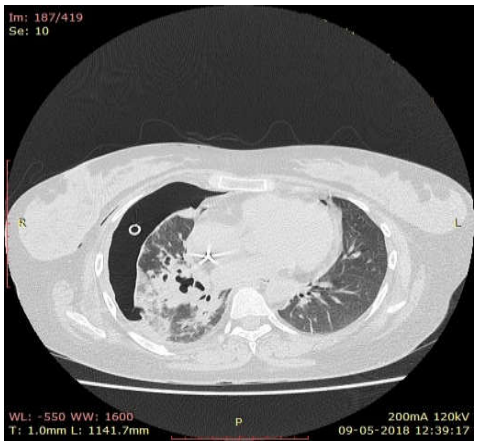

Figure 2 HRCT Scan

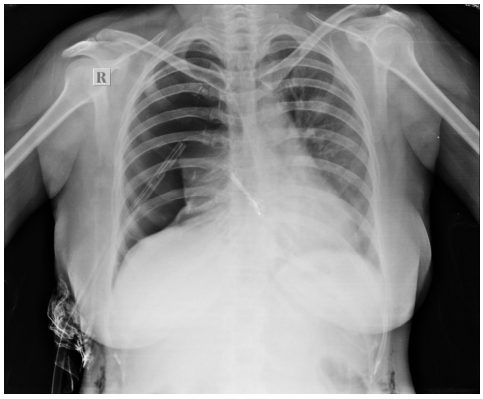

Figure 3 Chest X-ray PA view Post-ICD insertion

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