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A case report of systemic lupus erythematous presenting with diffuse alveolar hemorrhage

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ABSTRACT

Systemic lupus erythematous (SLE), a chronic autoimmune disease with a wide range of manifestations such as, skin rashes, arthritis, fatigue, neurologic symptoms, stroke, glomerulonephritis, cardiovascular symptoms, and involvement of the lungs. Diffuse alveolar hemorrhage (DAH) is not common among SLE patients, but the mortality rate of DAH associated with SLE is reported to be significant. We report a 35 years old woman that diagnosed systemic lupus erythematous presenting with diffuse alveolar hemorrhage.

Keywords: systemic lupus erythematous, diffuse alveolar hemorrhage, hemoptysis, rituximab

INTRODUCTION

Systemic lupus erythematous (SLE), a chronic autoimmune disease with a wide range of manifestations such as skin rashes, arthritis, fatigue, neurologic symptoms, stroke, glomerulonephritis, cardiovascular symptoms, and involvement of the lungs [1]. This disease occurs mostly in adults, and 6-10 times more in females, although 15-20% of cases are diagnosed in childhood [2,3]. The underlying pathology is caused by antibodies which form cytosolic and nuclear antigen reactive immune complexes [4].

Respiratory tract involvement occurs in about 50% to 70% of cases, and co-morbidities such as pneumonia, bronchiolitis obliterans, pleuritis, pulmonary hypertension and diffuse alveolar hemorrhage (DAH) could be seen [5,6].

DAH is not common among SLE patients, but the mortality rate of DAH associated with SLE is reported to be significant [7].

Here we report a 35-year-old woman who presented with shortness of breath, hemoptysis, and skin lesions, and was diagnosed with DAH as a manifestation of SLE.

CASE PRESENTATION

The patient is a 35-year-old unmarried woman who came after a recent cold with complaints of shortness of breath at the level of MMRC II, cough, massive hemoptysis, skin lesions on the lower limbs and left elbow.

Associated symptoms include weakness and lethargy, an episode of oral ulcer 4 months before admission, mild fever in the last week, headache, feeling of congestion and stuffy nose. She did not mention the history of rheumatologic disease and other diseases in herself and her family.

During the examination, there was a 35-year-old woman who was lying in bed, the appearance of the patient was ill. She did not have a fever. Her blood pressure was 125/80. Heart rate was 100-95; respiration rate was 28 per minute.

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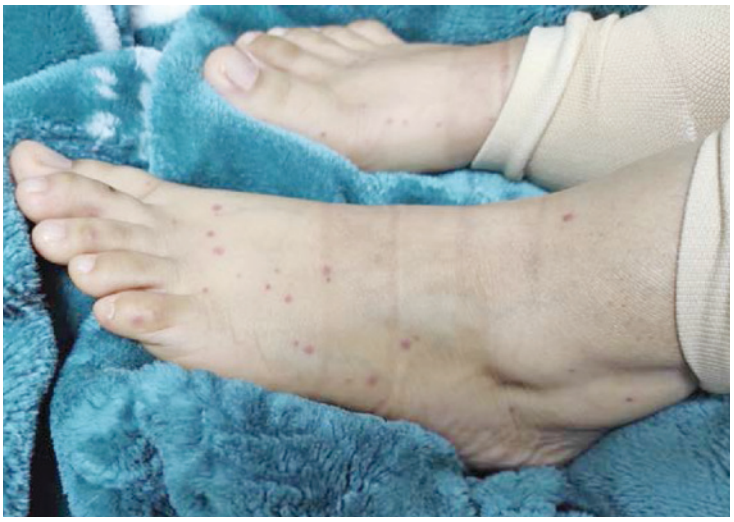

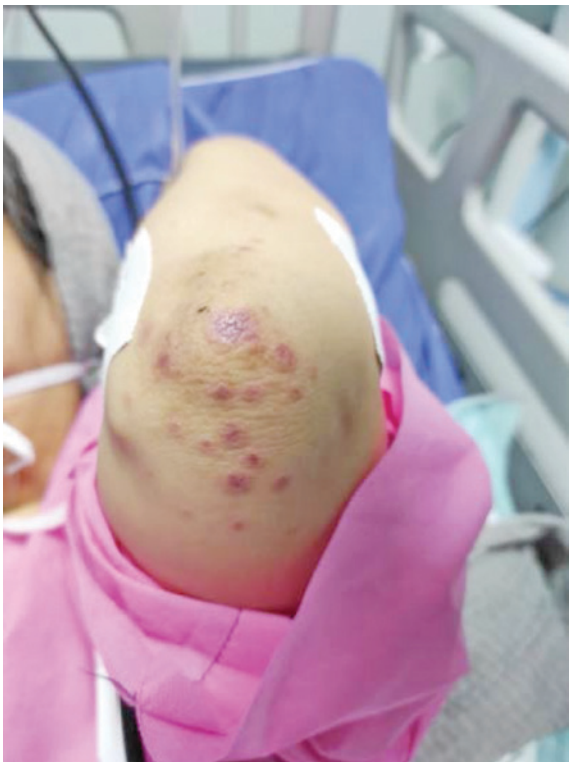

FIGURE 1. Skin lesions in the form of palpable purpura

SPO2: 88% without oxygen and 94% with oxygen mask.

BMI: 23.8, Tilt was negative. Skin lesions in the form of palpable purpura were evident in both lower limbs from fingers to ankles and in the elbow of the left hand (Figure 1). There was no clubbing. There was no hair loss, pulling test was negative, and the oral wound was not visible. There was no lymphadenopathy. JVP was not prominent. Thyroid examination was normal.

There was no chest deformity. Tachycardia was heard on auscultation without murmurs. Both halves of the chest were relatively involved in breathing. Lung auscultation did not have whizzing or crackles. Lung sounds were generally reduced. Abdominal and joint examinations were normal. The pulses were normal. There was no edema and cyanosis, and there was no difference in size in the lower limbs.

In the initial paraclinical examinations, the patient had a drop in white blood cells and hemoglobin.

WBC:3800, poly:79%, Hb:7.4, MCV:80, pletelet:176000, INR:1, HIV: Negative HBsAg: Neg, HCVAb:neg, ESR: 50, Retic count:5, albumin: 3.6, creatinine: 0.9, LFT: normal, electrolytes: normal, ferritin:1384, urine protein /24 h: 413/150, C-ANCA and P-ANCA: neg anti RO/LA:neg, APS tests:neg, C3, C4, CH50: Normal, RF and antiCCP: neg, anti G.B.M: Neg, FANA: 1/160 and DSDNA 250/18 and were positive. In the CT scan of the lung, bilateral involvement was evident in the form of diffuse alveolar hemorrhage (Figure 2).

The peripheral blood slide was normal; Echocardiography and kidney ultrasound was reported to be normal. The patient was transferred to the intensive care unit with this sign, and the prednisolone pulse as follow was started:

Amp Methyl Prednisolone 500mg + serum H/S 0.5 liter Q 1.5 Hour

It was prescribed for 5 days with the control of vital signs. Other treatment measures include: Amp Meropenem 1g IV TDS, Tab pantoprazole 40 mg po OD, Tab Acetaminophen Codeine POTDS,Syp Dextromethorphan 10 cc PO TDS, Tab Co-trimoxazole 80/400 mg Po daily, Cap Ferrodin PO BID, Amp Eprex 4000u s.c 3 times a week.

According to the rheumatology consultation and according to the results of the tests, the patient was treated with rituximab. Rituximab was slowly infused at a dose of 1000 mg into one liter of normal saline over 2 hours and repeated after 2 weeks. Finally, the patient received 5 doses of pulse methylprednisolone, 2 doses of rituximab at an interval of 2 weeks, antibiotics, supportive measures and prescription:

Prednisolone tablet 50 mg daily, the dose of which was gradually reduced, the Cellcept was started and the general condition was good, there was a significant improvement in the clinic and the tests (Figure 3), and the recommendation to continue the treatment was discharged.

DISCUSSION

Dr. William Osler first described DAH in 1904 [8].

DAH is a rare complication in SLE patients and is seen in 2-5.4% of cases, but despite the rareness it is associated with high mortality (about 50-80%) [7]. It is reported that SLE could be presented initially with DAH in 10-20% of patients [9]. DAH patients are mostly reported to have about 30 years of age, and the disease occurs at averagely 35 months after the onset of SLE [10,11].

DAH has a classic triad of hemoptysis, newly pulmonary infiltrates, and sudden fall of hemoglobin concentration; but not all cases present with this tri-

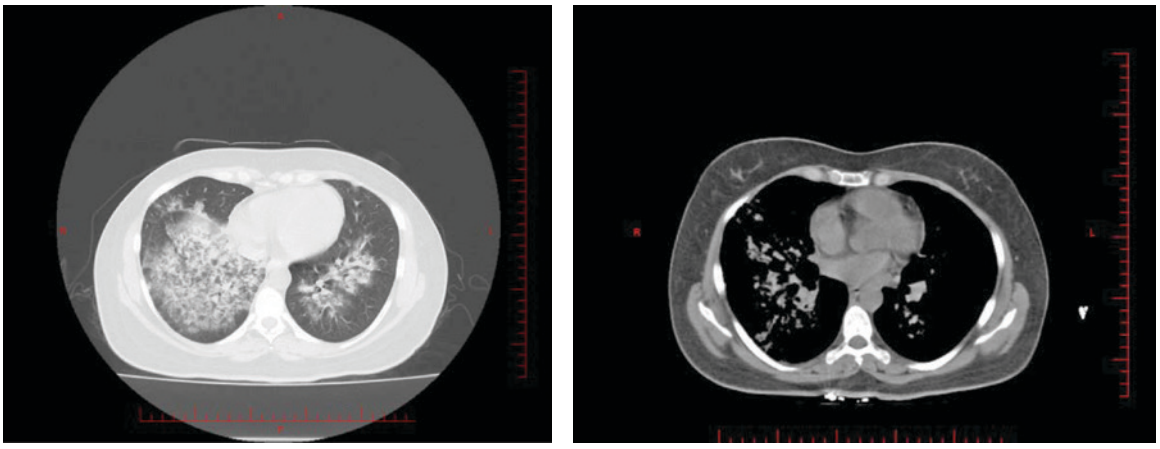

FIGURE 2.A. The CT scan of the lung, bilateral involvement was evident in the form of diffuse alveolar hemorrhage

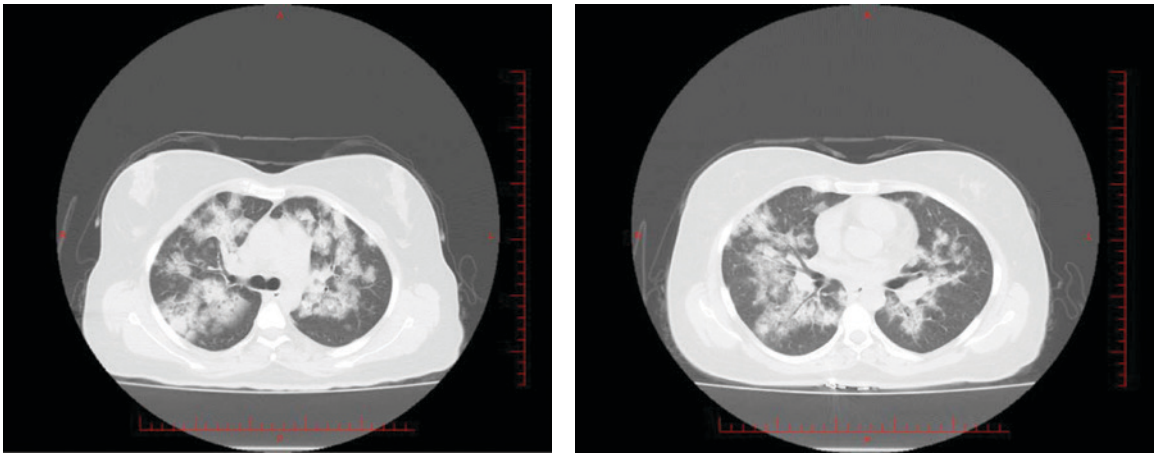

FIGURE 2.B. The CT scan of the lung, bilateral opacity that diagnosed as diffuse alveolar hemorrhage

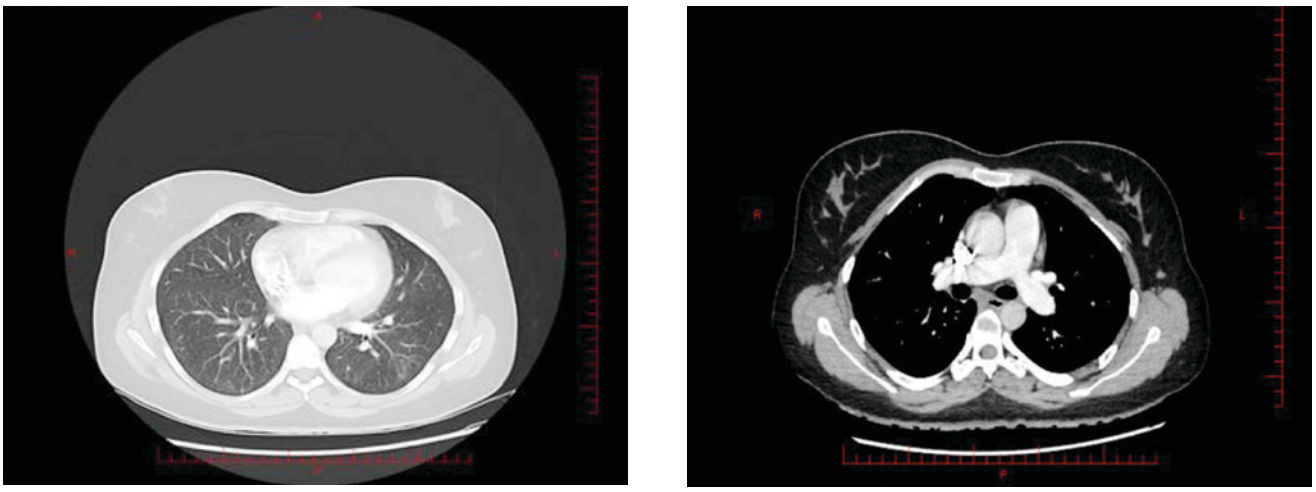

FIGURE 3. The lung CT scan 2 week later than beginning of treatment

ad, and other signs and symptoms include cough, dyspnea, chest pain, rales in auscultation, fever over 38 degrees Celsius, and pleural effusion [12,13]. The prevalence of hemoptysis has been reported to be around 25-100% [9,13].

CT scan and MRI are useful in diagnosis, and radiographic remarks include diffuse alveolar and lobar infiltrations [14,15]. It is also reported that High resolution CT (HRCT) could reveal consolidations, patches, and ground-glass opacities [16]. It is reported by several studies that a drop of hemoglobin concentration about 1-2 g/dl is also suggestive of DAH [17-19]. Other laboratory findings include thrombocytopenia, C3 hypocomplementemia, immune complexes in biopsy, and anti-dsDNA higher titers than SLE [17]. Macrophages containing hemosiderin, hemorrhages in bronchoalveolar lavage (BAL), and a rise in the carbon monoxide diffusing capacity (DLCO) are considered as criteria of DAH [20]. However, hemosiderin, which is a product of breakdown of hemoglobin is detectible after 48 hours of bleeding [21].

The risk of DAH has been reported to be increased during active renal involvement, which includes 80% of patients, and biopsies often reveal lupus nephritis

of class III or IV [9, 22]. However SLE activity could not be considered as the sole risk factor for DAH, in the study conducted by Haupt et al. [23] it was found that most DAH cases had other risk factors such as infection, renal failure, aspiration and heart failure. Martinez-Martinez et al. [17] reported that patients with ANCA (antineutrophilic cytoplasmic antibody) related vasculitis or APS (antiphospholipid syndrome) develop DAH more commonly.

Due to respiratory failure in the first days or even hours, DAH is considered as an emergency, hence the treatment must be started with no further hesitation [24]. The treatment of choice is intravenous methylprednisolone, but other therapies such as cyclophosphamide, plasmapheresis and rituximab could be added [25-27]. Esper et al. has reported that inhaling recombinant activated factor VII (rFVIIa) using jet nebulizer should be considered as an additive treatment to the main therapy (methylprednisolone boluses) [5].

The prediction of the outcome of DAH associated with SLE, OSF (organ system failure) and APACHE II (Acute Physiology, Age, and Chronic Health Evaluation) scores could be utilized [28].

Finally, for our patient with symptoms of shortness of breath, hemoptysis, arthralgia, leukopenia, Headache, anemia, High ESR, proteinuria, diffuse alveolar lung Hemorrhage FANA + and DsDNA + systemic lupus erythematous was diagnosed.

She was treated with corticosteroid and rituximab and other supportive measures and was discharged with significant improvement and she is under outpatient care.

Patient consent: obtained.
Conflict of interest: none declared
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