โรคลูปัสที่มาด้วยภาวะปอดอักเสบเฉียบพลันเป็นอาการแรกเริ่ม: รายงานผู้ป่วย

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Acute Lupus Pneumonitis as the First Presentation in Systemic Lupus Erythematosus Patient: A Case Report

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บทคัดย่อ

หลักการและวัตถุประสงค์: โรคลูปัส (Systemic lupus erythematosus : SLE) เป็นโรคที่เกิดจากความผิดปกติของระบบภูมิคุ้มกันต่อระบบต่างๆ ของร่างกายโดยมีอาการแสดงในระบบทางเดินหายใจที่หลากหลาย ภาวะปอดอักเสบเฉียบพลันจากโรคลูปัส (acute lupus pneumonitis) ถือเป็นภาวะที่พบได้ยากและเป็นภาวะแทรกซ้อนที่รุนแรงถึงแก่ชีวิตได้ การศึกษานี้ทำขึ้นเพื่อศึกษาลักษณะทางคลินิกของผู้ป่วยโรคลูปัสที่มีภาวะปอดอักเสบเฉียบพลันเป็นอาการนำและผลการรักษา

วิธีการศึกษา: เป็นรายงานผู้ป่วย 1 ราย โดยเก็บรวบรวมข้อมูลจากเวชระเบียนผู้ป่วย โรงพยาบาลสุรินทร์

ผลการศึกษา: รายงานผู้ป่วยหญิงอายุ 17 ปี มีอาการไอแห้งๆ ร่วมกับหอบเหนื่อยมากขึ้นมา 1 เดือนได้รับการวินิจฉัยภาวะปอดอักเสบเฉียบพลันเป็นอาการแสดงแรกของโรคลูปัส หลังได้รับการรักษาด้วยสเตียรอยด์ขนาดสูงพบว่าอาการทางคลินิก ผลตรวจทางห้องปฏิบัติการและภาพรังสีของผู้ป่วยดีขึ้นอย่างชัดเจน

สรุป: การวินิจฉัยภาวะปอดอักเสบเฉียบพลันจากโรคลูปัสเป็นสิ่งที่ท้าทาย หากวินิจฉัยเร็วและรักษาอย่างทันท่วงที จะช่วยให้ผลการรักษาดีขึ้นได้

คำสำคัญ: ปอดอักเสบเฉียบพลันจากโรคลูปัส, อาการนำ, อาการแรกเริ่ม, โรคลูปัส

Abstract

Background and Objective: Systemic lupus erythematosus (SLE) is a multisystem autoimmune disease that has broad pulmonary manifestations. Acute lupus pneumonitis is one of its rare and mortal complications. This study aimed to describe the SLE patient with clinical features of acute pneumonitis as the leading symptom and treatment outcome.

Materials and Method: A case report, data was collected from medical records of Surin hospital

Results: We report a 17-year-old woman, who presented with dry cough and progressive dyspnea for a month. She was diagnosed with acute lupus pneumonitis as the initial presentation of SLE. Her clinical, biological and radiological improvements were noticed after receiving pulse methylprednisolone.

Conclusion: Diagnosing lupus pneumonitis is challenging. Early diagnosis and timely treatment can improve the disease outcome and its prognosis.

Keywords: lupus pneumonitis, initial, first presentation, systemic lupus erythematosus

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Introduction

Systemic lupus erythematosus (SLE) is a chronic systemic autoimmune connective tissue disease with multi-organ involvements. Pleuropulmonary manifestation has been reported wide-ranging between 16 and 90% of SLE patients[1-3]. Pulmonary involvement includes pleuritis and/or pleural effusion (most common), pulmonary arterial hypertension, pulmonary thromboembolism, pulmonary vasculitis, chronic interstitial lung disease, acute lupus pneumonitis, diffuse alveolar hemorrhage bronchiolitis obliterans and shrinking lungsyndrome[1]. Acute lupus pneumonitis is an uncommon presentation with an incidence of 1-4%[1,4]. We describe the case of a 17-year-old woman with subacute non-productive cough with progressive dyspnea for a month. Acute lupus pneumonitis was diagnosed as the first presentation.

Materials and methods

The medical records of the SLE patient with lupus pneumonitis as an initial presentation were reviewed. Her clinical features, laboratory and radiography investigations, treatments and outcomes were collected.

Results

A 17-year-old Thai female with no previous underlying diseases was referred to the tertiary hospital due to intermittent high-grade fever, dry cough, and progressive dyspnea for 4 weeks. She has been previously initial workup at a community hospital in the first week of illness. Laboratory findings showed pancytopenia with leukocyte count of $2.2*10^3$/µL (4.5-11 10^3/µL), hemoglobin of 8.6 g/dL (12-16 g/dL) and platelet count of $99*10^3$/µL (140-400*10^3/µL). Renal and liver function tests were normal. The initial chest X-ray was a negative study. Tropical infections such as dengue, scrub typhus, leptospirosis and COVID-19 infection were also excluded. Her chest symptom was worsening with oxygen saturation (SpO_2) of 94% (on oxygen cannula 5 L/min). Due to lack of improvement despite intravenous antibiotics therapy, she was referred for further investigation.

At our hospital, she had fever (38.6 °C), tachycardia (104 beats/min), tachypnea (22 breaths/min) and hypoxia on room air (SpO_2 of 83%). Discoid lupus erythematosus was observed on the right ear. Fine crepitation was heard at both lungs. Other physical examinations were unremarkable. The chest X-ray showed bilateral peripheral ground-glass opacity (Figure 1). On HRCT thorax showed bilateral multiple patchy peripheral and peribronchovascular ground-glass opacity with subpleural predominance (Figure 2).

Pulmonary function test was not done due to her clinically dyspnea. Laboratory tests showed pancytopenia again with leukocyte count of $1.7*10^3$/µL, hemoglobin of 8.9 g/dL and platelet count of $53*10^3$/µL. Direct Coombs' test was positive (1+), without hemolytic feature on the peripheral blood smear. Her creatinine was 0.7 mg/dL. Urinalysis showed proteinuria (urine protein 2.65 g/24 hr). She also had abnormal liver function tests with albumin of 2.7 g/L, globulin of 3.2 g/L, total bilirubin of 0.67 mg/dL, direct bilirubin of 0.23 mg/dL, AST of 469 U/L, ALT 52 U/L and ALP of 136 U/L. HBsAg, anti-HCV and anti-HIV were negative. Serum complements showed decrease of both C3 and C4; C3 was 0.21 g/L (0.9-1.8), C4 was lower than 0.08 g/L (0.1-0.4).

The patient had been treated as multilobar pneumonia with piperacillin/tazobactam intravenous (IV) since the admitted time. However, she remained febrile, dry cough, dyspnea and progressive hypoxia. Two days later, the bronchoscopy with bronchoalveolar lavage (BAL) and transbronchial biopsy was performed by a pulmonologist to exclude other pulmonary infections. BAL demonstrated leukocyte of 1,323 cells/µL (90% lymphocyte, 10% neutrophil). Later, all microbiological cultures including tuberculosis and fungus were negative. Cytology revealed chronic inflammation and negative for malignancy. Transbronchial biopsies showed chronic inflammation with numerous histiocytes and the absence of hemosiderin-laden macrophage.

Autoimmune markers revealed high antinuclear antibodies(ANA), titre 1:640, with a homogenous pattern and anti-dsDNA was positive, anti-Smith and antiphospholipid antibodies were all negative.

She was diagnosed as SLE with acute lupus pneumonitis according to the ERLAR/ACR 2019 criteria[4]. Hematological, mucocutaneous and renal manifestations were also present (pancytopenia, discoid rash and proteinuria/lupus nephritis) as well as immunological domains (positive anti-dsDNA and low C3, C4).

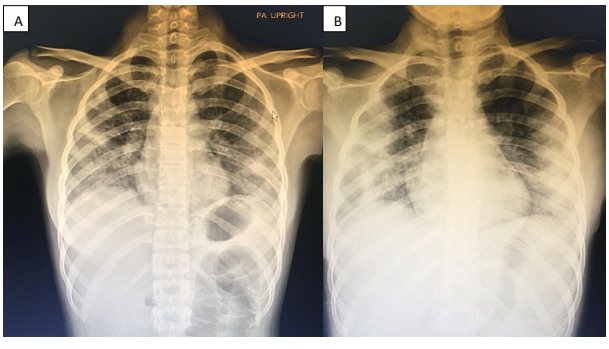

Figure 1 Chest X-rays on admission (A) and after bronchoscopy with BAL (B) show increased ground-glass opacity of both lungs

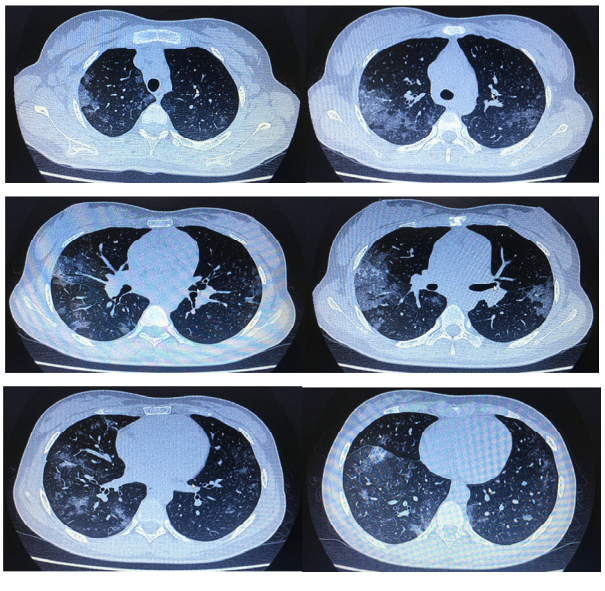

Figure 2 HRCT thorax shows bilateral multiple patchy peripheral and peribronchovascular ground-glass opacity with subpleural predominance

After underwent bronchoscopy, our patient was immediately started on pulse methylpredisolone 1 gm IV daily for three days due to worsening to impending respiratory failure. Her clinical status significantly improved; she was afebrile and could wean off the respiratory support from high-flow nasal cannula to room air within a few days later. As a result, steroid treatment was tapper back and mycophenolate mofetil and hydroxychloroquine were added.

Two weeks after being discharged, she came back for a follow-up visit without any dyspnea. Her oxygen saturation was 98% on room air. Chest X-ray revealed a significant decreased of bilateral pulmonary opacities (Figure 3). Her blood analysis also showed improvement with leukocyte count of $8.7*10^3$/µL, hemoglobin of 9.8 g/dL and platelet count of $119*10^3$/µL. The urine protein/creatinine ratio (UPCR) was 1.12. She also had improved liver function tests with albumin of 3.5 g/L, globulin of 3.1 g/L, total bilirubin of 0.59 mg/dL, direct bilirubin of 0.09 mg/dL, AST of 38 U/L, ALT of 16 U/L and ALP of 123 U/L. Lupus hepatitis was probably diagnosed in this patient because of response to the treatment.

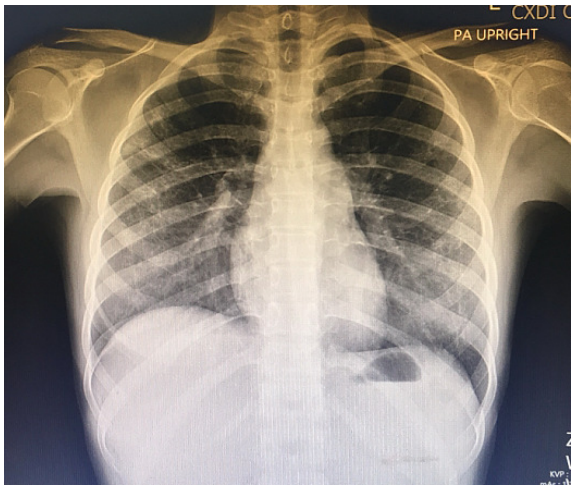

Figure 3 Chest X-ray on 2-week follow-up visit shows improvement of bilateral ground-glass opacity

Discussion

This was the first case report in Thailand of acute lupus pneumonitis which is an uncommon manifestation of SLE with a poor prognosis and high mortality rates. Therefore, early diagnosis and treatment are essential in these patients. At the time the patient was referred to us, it was not difficult to diagnose because she had many systemic symptoms that are compatible with SLE such as pancytopenia and proteinuria. A thorough physical examination also found a discoid rash, which is a pathognomonic sign of this disease, made it easier to diagnose and investigate immediately to support the diagnosis.

However, acute lupus pneumonitis is rarely to be the initial presentation in SLE patients[1,2]. Its clinical feature is similar to pneumonia, characterized by

fever, cough with or without hemoptysis, hypoxemia and even acute respiratory failure, as it found in our patient. Therefore, pulmonary infection is the most concern in the differential diagnosis and it is necessary to initiated empirical antibiotics and continued until the infection is ruled out.

Typical radiographic image of acute lupus pneumonitis is bilateral alveolar infiltrations with predominance at the lung bases. Chest computed tomography commonly showed ground-glass opacities and bilateral patchy consolidation[6].

Wan et al. reported a case series of five patients in which acute lupus pneumonitis was the initial presentation of SLE[7]; all were females, aged 14-26 years old, all had the additional manifestations of SLE that enable the diagnosis: fever and mucocutaneous in 100%, hematological in 80%, serositis in 40%, positive for ANA in 100% and positive for anti-dsDNA in 60% which are seen to be found in our patient.

Neither bronchoscopy with BAL nor transbronchial/ lung biopsy have value in establishing the diagnosis of lupus pneumonitis but they are useful to exclude infection and diffuse alveolar hemorrhage, another rare and mortal pulmonary complication in SLE patients.

There are no clinical trials for this treatment; the current regimens are based on case reports[1]. The cornerstone of treatment is high-dose glucocorticoid adjustment to the severity and clinical response. Adding immunosuppressive agents (such as cyclophosphamide, azathioprine, mycophenolate mofetil), intravenous gammaglobulin (IVIg), plasmapheresis and rituximab had been reported in the treatment of acute lupus nephritis. In spite of this, the outcome is often poor with a high mortality rate of 40%-50% from the previous series[2,7].

Conclusion

Acute lupus pneumonitis is a rare and mortal manifestation in SLE patients. Early diagnosis, appropriate investigation and prompt treatment are the success keys to improving the disease outcome.

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