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Case Report

Interstitial lung disease in Indonesian adult with systemic sclerotic: A rare case

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ABSTRACT

Background: Systemic Sclerotic (SSc) with interstitial lung disease (ILD) is a rare autoimmune case. In Indonesia, SSc-ILD cases are still not widely reported.
Case presentation: A 43-year-old female complained fever, yellow phlegm, and shortness of breath for 1 week. The evaluation results based on the American College of Rheumatology and European League Against Rheumatism (ACR EULAR) classification criteria for systemic sclerosis showed a score of 17. The patient received Methylprednisolone 8 mg tab/8 hours, Meropenem 1 gr/8 hours, Ranitidine 50 mg/12 hours, Metoclopramide 10 mg/8 hours, KSR 600 mg tab/8 hours, Paracetamol 500 mg tab/8 hours, VIP albumin 500 mg tab/8 hours, and Folic acid 400 mg tab/8 hours. The patient underwent high resolution computer tomography (HRCT). A chest scan obtained SSc-ILD. Sputum culture results did not reveal Mycobacterium tuberculosis.
Discussion: SSc-ILD patients in endemic tuberculosis need to be examined for GeneXpert and sputum culture. The HRCT scan is more helpful in diagnosing SSc-ILD than bronchoalveolar lavage (BAL).
Conclusion: The HRCT is one of the gold standards in establishing the diagnosis of SSc-ILD.

1. Introduction

Systemic sclerotic (SSc) is a complex autoimmune disease with multiple manifestations, including vasculopathy, Raynaud's phenomenon, immune dysfunction, and fibrosis of the skin and internal organs. This is a rare disease with an estimated number of 3–24 patients per 100,000 people [1]. Interstitial lung disease (ILD) is the most common form of SSc [2], and it is estimated that SSc-ILD is >80% of SSc cases [1,2]. In Indonesia, SSc-ILD is a rare case that until now has only been reported in two cities, namely Bandung and Jakarta [3,4]. Based on this description, we are interested in reporting the SSc-ILD in Indonesian adults. We report based on Case Report Surgical (SCARE) 2020 guideline [5].

2. Case presentation

A 43-year-old female had fever, yellow phlegm, and shortness of breath for 1 week. The patient had a history of type 2 diabetes mellitus (T2DM) and SSc. The results of physical examination showed the patient was weak with pulse rate (126 ×/min) and respiratory rate (30 ×/min). On 2/3 of the lower right lung, percussion was dullness, and crackles auscultated. Meanwhile, in the lower 1/3 of the left lung, audible wheezing sounds were obtained. The results of chest x-ray showed similarly pulmonary tuberculosis (Fig. 1). The patient was given oxygen using a nasal cannula 3 L/min with an oxygen saturation of 98%. The results of the blood examination revealed leukocytosis (white blood count of 11.370/μL), anemia (Hemoglobin of 9.5 g/dL), hypoalbuminemia (Albumin of 3.0 g/dL), hypokalemia (potassium of 3.3 mol/L), and increased C-Reactive Protein (CRP of 22.3 mg/dL).

The evaluation results based on the American College of Rheumatology and European League Against Rheumatism (ACR EULAR) classification criteria for systemic sclerosis [5] obtained a score of 17, which consisted of thickening of the skin of the fingers on both hands from proximal to the metacarpophalangeal joints (point 9), sclerodactyly (distal to the metacarpophalangeal joints but proximal to the interphalangeal joints (point 4), telangiectasias (point 2) and ILD (point 2), contractures on digits 2, 3, 4 right and left hand, thickening of the skin on both arms. The patient received Methylprednisolone 8 mg tab/8 hours, Meropenem 1 gr/8 hours, Ranitidine 50 mg/12 hours, Metoclopramide 10 mg/8 hours, KSR 600 mg tab/8 hours, Paracetamol 500 mg

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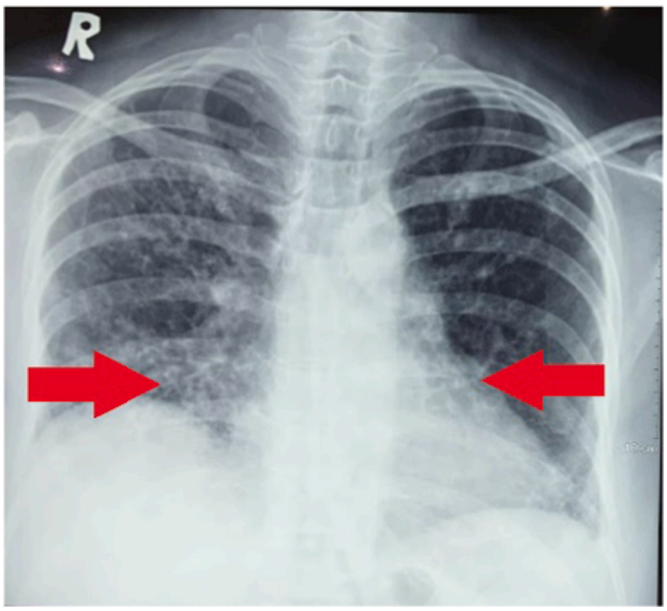

Fig. 1. X-Ray of the anterior-posterior showed honeycomb similarly pulmonary tuberculosis.

tab/8 hours, VIP albumin 500 mg tab/8 hours, and Folic acid 400 mg tab/8 hours.

On the third day, HbA1c was 6.3% and fasting blood sugar was 114 mg/dL. Meanwhile, on the fifth day, the patient got the results of GeneXpert RIF/MtB not detected and the sputum culture results obtained *Viridans streptococcus* which sensitive to used antibiotic (Meropenem). On the seventh day, the patient underwent high resolution computer tomography (HRCT) that revealed reticular opacity with bronchial dilatation and honeycomb appearance in both lung fields, supporting the manifestations of scleroderma in the lungs (Fig. 2). The patient, on the eighth day, showed clinical improvement and was advised to conduct outpatient treatment. The results of sputum culture did not show *Mycobacterium tuberculosis*.

3. Discussion

The ILD usually appears in the first 4–6 years after the onset of SSc, so aggressive and early screening is necessary. The use of HRCT-Scan thorax is essential to establish the diagnosis [6]. A study stated that HRCT-Scan was more effective than bronchoalveolar lavage (BAL) in a comparison (90.6% vs 70%) to identify SSc-ILD [7]. Pulmonary tuberculosis examination is considered because the patient is at risk of tuberculosis due to T2DM, lives in endemic tuberculosis country, and the chest x-ray shows typical feature of pulmonary tuberculosis as a differential diagnosis [8,9].

Management of SSc-ILD, based on some literature, recommends the use of mycophenolate mofetil (MMF) at a single dose of 3 g/day (orally). Should the patient is not tolerant to MMF, it is recommended to use 720 mg dose 3 ×/day [10,11]. In addition to MMF, cyclophosphamide (CYC) can be administered, in which the efficacy of administering CYC vs MMF in SSc-ILD is 42% vs 35% [11,12]. Previous studies showed that administration of CYC combined with prednisone can increase forced vital capacity (FVC) at 6 and 12 months of therapy [13]. MMF and CYC can be used simultaneously to improve lung capacity and skin problems in SSc-ILD patients [14]. VIP albumin is used to increase albumin levels which the minimum normal value is 3.5 g/dL [15].

The risk of mortality in SSc-ILC patients increases with various symptoms, so it greatly affects the quality of life of patients. This decrease in quality of life is characterized by chronic pain, fatigue, and sleep disturbances that get worse over time [12,16] which influenced by the presence or absence of pulmonary fibrosis, pulmonary hypertension, or heart disease in patients. Other diseases that can affect the risk of mortality are kidney disease, malignancy, gastrointestinal disease, and also infection [17]. Limited resources also cause SSc-ILD management to sometimes become a problem in itself.

4. Conclusion

A 43-year-old female is diagnosed with SSc-ILD, in which the diagnosis is confirmed through a chest HRCT scan. The HRCT is one of the gold standards in establishing the diagnosis of SSc-ILD and is non-invasive. It can detect the smallest possible parenchymal abnormality. Management of SSc-ILD currently uses corticosteroid therapy.

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None.

Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

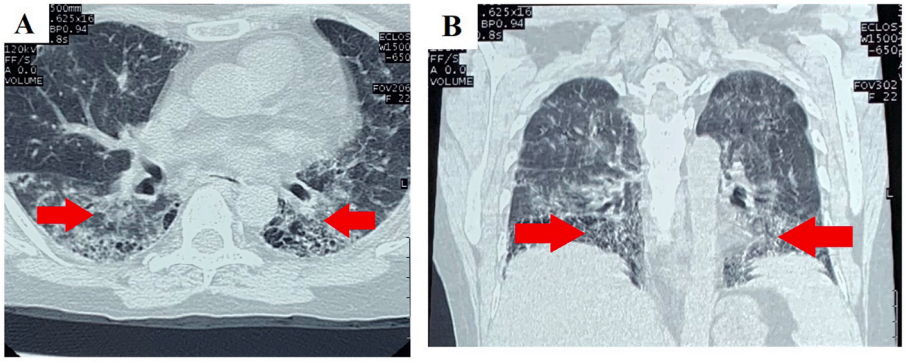

Fig. 2. High resolution computed tomography scan thorax indications of systemic sclerosis.

Author contribution

All authors contributed toward data analysis, drafting and revising the paper, gave final approval of the version to be published and agree to be accountable for all aspects of the work.

Registration of research studies

1. Name of the registry:-
2. Unique Identifying number or registration ID: -.
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): -.

Guarantor

Helmia Hasan.

Declaration of competing interest

The authors declare no conflict of interest.

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