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Case Report

Variation of usual interstitial pneumonia using HRCT in Scleroderma Patients [☆]

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ABSTRACT

Progressive Systemic Sclerosis or Scleroderma is a systemic inflammation marked by endothelial damaged, fibrosis, and inflammation in skin, joints, and visceral organs. Pulmonary hypertension and interstitial pulmonary disease (Ssc-ILD) are the most reported pulmonary complications in scleroderma patients. The pathogenesis of SSc-ILD is not well understood and the spectrum of SSc-ILD ranges from minimal lung involvement, which is often non-progressive, to severe illnesses. Usual Interstitial Pneumonia's (UIP) is one pattern of Ssc-ILD, marked in one-third of the patients with characteristic of honeycomb appearance and bronchiectasis. It is very important to determine the UIP based on radiology imaging especially with the presence of a poor prognosis in patients with UIP. This case report will discuss the importance of finding UIP-type ILD patterns based on HRCT in patients with scleroderma and different outcome.

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Introduction

Progressive Systemic Sclerosis or Scleroderma is a systemic inflammation marked by endothelial damaged, fibrosis, and inflammation in skin, joints, and visceral organs including lungs, kidneys, and digestive system.¹ Pulmonary hypertension and interstitial pulmonary disease (Ssc-ILD) are the most common pulmonary complications in scleroderma, with up to one-third of patients experiencing pulmonary fibrosis.¹ Pulmonary parenchyma involvement often in the very beginning after the diagnosis of Ssc-ILD was made, with 25% of the patients experience significant deterioration of the dis-

ease within 3 years, and this progression was seen on clinical manifestations, radiological evidences, and bronchoalveolar lavage (BAL).²,

The pathogenesis of SSc-ILD is still not well understood. It is thought to be associated with abnormal interactions between endothelial cells, lymphocytes and/or monocytes and fibroblasts that cause extracellular matrix overproduction by fibroblasts due to the hypoxic tissue regulation and vascular hyperreactivity.³ Patients may show elevated levels of pro-inflammatory cytokines such as interleukin IL-8, TNF-alpha and macrophage-1 depicted in BAL fluid. B-cells are also thought to be involved in the pathogenesis since patients with SSc-ILD usually have higher levels of anti-topoisomerase antibodies and anti-fibroblast antibodies, the latter of which have

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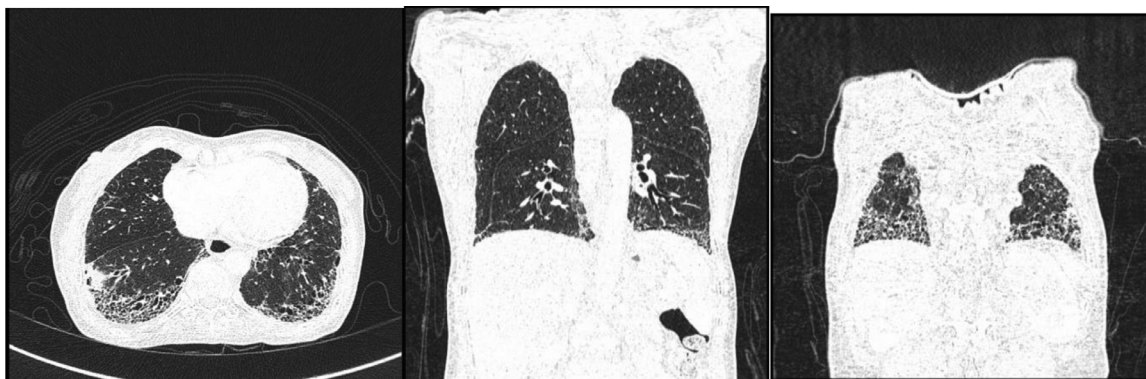


Fig. 1 – HRCT Chest of the patient showed honeycombing picture with reticular opacity, ground glass opacity in the bilateral pulmonary basal (segment 5-10) accompanied by bronchiectasis in segment 7-10 bilaterally

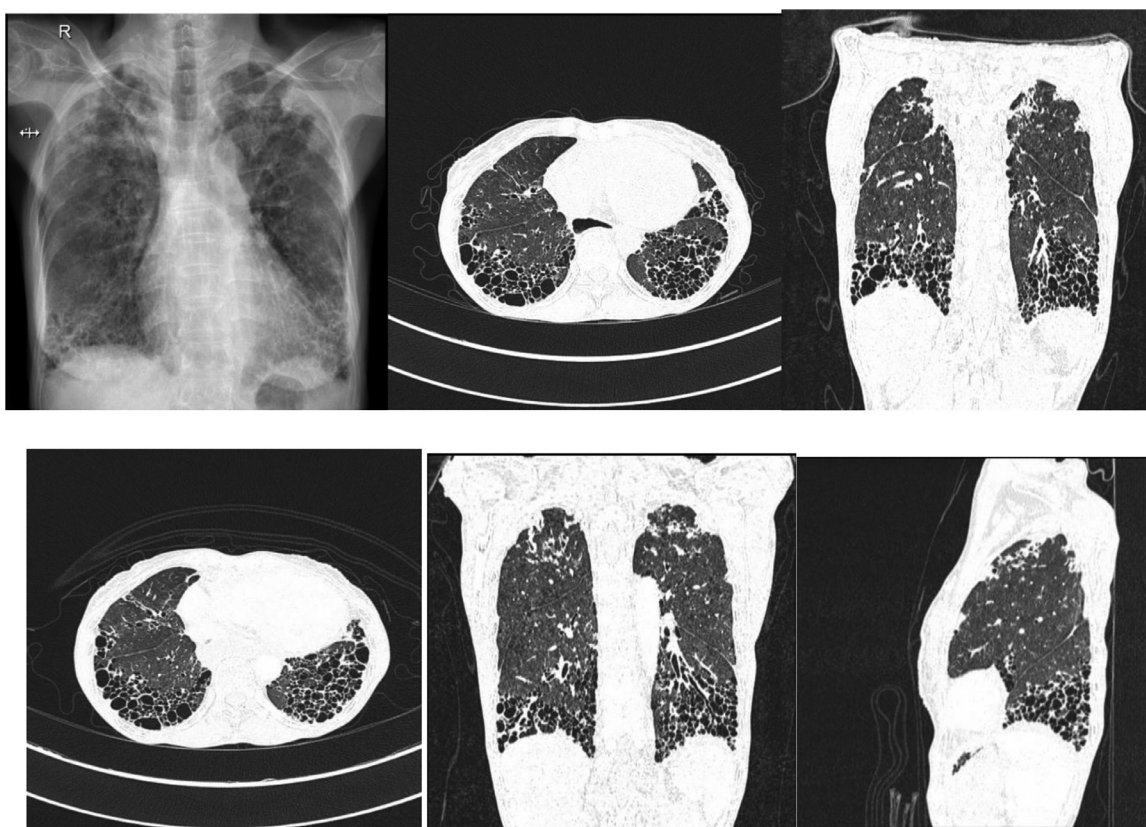


Fig. 2 – HRCT chest of the patient showed honeycombing picture with reticular opacity, ground glass opacity in the bilateral pulmonary basal (segment 5-10) with bilateral bronchiectasis in segment 7-10

been shown to induce fibroblasts activation and increase the production of extracellular matrices.³

The spectrum of SSc-ILD varies widely from minimal lung involvement, usually non-progressive, to severe illnesses especially during in the first year of the disease and would develop into respiratory failure and eventually death.² Symptoms are not specific, including dyspnea on effort, cough, and fatigue. Since SSc-ILD can develop without any symptoms, high resolution CT (HRCT) should be performed at initial diagnosis, along with a pulmonary capacity examination (PFT) for

early detection.² The degree of pulmonary fibrosis from HRCT are considered to be a strong predictor of mortality. HRCT is an important role of diagnostic evaluation for patients with SSc-ILD, accompanied by lung function tests that show restrictions.²

Usual Interstitial Pneumonia's (UIP) pattern marked by honeycomb appearance and bronchiectasis are found in one-third of patients with SSc-ILD.⁴ Ground glass-opacity could also be an indicator of progressive lung parenchyma fibrosis.⁴ Compared to pulmonary biopsies of patients with idiopathic

pulmonary fibrosis, SSc-ILD patients have more germinal centers and less fibroblast focus.⁴ However, pulmonary biopsies are invasive and not routinely performed in most healthcare facility for this purpose.

Therefore, it is very important to determine the UIP based on radiological evidence to ensure the best treatment given. Moreover, UIP is one predictor of poor outcome for patients and therefore its detection become vital. This case report will discuss the importance of finding UIP pattern as a marker for ILD using HRCT in patients with scleroderma.

Case Report

Case 1

A 60-year-old female came to the hospital with chief complaint of skin stiffness since 5 months prior the stiffness progressed as time goes by. In the last 2 months the patient also experienced cough and severe shortness of breath. Patient also felt a cold sensation on the tip of the fingers and sometimes the skin turned to blue. On physical examination, stiffen and hardened skin were observed, matches to the Modified Rodnan Skin Score (MRSS) 47. Spirometry resulted in FVC 35% which depicted pulmonary restriction. HRCT was performed and they showed honeycomb appearance with reticular opacity, ground glass opacity in the bilateral basal lung region (segment 5-10) accompanied by bronchiectasis in segment 7-10 bilaterally. Reticular opacities were also found in left lung parenchyma, matches with UIP's appearance. Patient had taken 500 mg of cyclophosphamide and 960 mg cotrimoxazole (Figure 1).

Case 2

A patient came into the clinic complaining shortness of breath and cough accompanied by weight loss. There were also fatigue since 2 months prior. Patients were having a history of scleroderma since 1998. Chest x-ray and CT scan were performed. HRCT findings showed honeycomb appearance with reticular opacity in the basal of lung parenchyma bilaterally (segment 5-10) accompanied by bronchiectasis in lung segment 7-10 and fibrosis in outer half of left lung parenchyma (Figure 2).

Patient were scheduled for spirometry but were cancelled due to patient's condition. Two months later, patient came to the ER complaining severe shortness of breath since the last 2 days. No symptoms of shortness of breath, cough, and fever before. There were also no vomiting and nausea. Patient also experienced reduced urine production since the last 1 month. Patient had previously treated by hemato-oncologist, blood transfusion was administered, and haemodialysis was advised. A pale conjunctiva and rhonchi were found on physical examination. Laboratory test obtained with Hb 4 mg/dL, ABG: 7.26/26.4/176.9/12.8/98.8, ureum and/or creatinine: 249 of 11.1, magnesium: 1.37, and calcium: 7.5. On HRCT, there is traction bronchiectasis in segment 1,2,3,4, honeycomb appearance accompanied by fibrosis, pleural thickening and minimal infiltrate was found. Patient was diagnosed with scleroderma and

renal crisis, and ILD. As the patient's condition was unstable, the chemotherapy cannot be initiated and the patient just received oral prednisone.

Discussion

The pattern of Usual Interstitial Pneumonia (UIP) on HRCT is one finding suggesting Idiopathic Pulmonary Fibrosis (IPF).² However, not every ILD with UIP will be classified as IPF. If there are signs of systemic disease involvement, such as connective tissue disease, then it would classified into connective tissue disease associated with ILD.⁵ Based on a study by Chung et al, there are some typical findings of patients with CTD who have ILD appearances in the form of UIP, which are: exuberant honeycombing sign, anterior upper lobe sign, and straight-edge sign. These findings have a high specificity in distinguishing UIP caused by IPF and CTD-ILD.⁵

In this case report, both patients have a concomitant disease, which is scleroderma, a connective tissue disease. The findings of UIP pattern are also typical, because HRCT showed a heterogeneous picture and distribution of abnormalities, as well as the presence of honeycomb appearance and diffuse infiltrate. Honeycomb appearance is a major sign of the UIP, with characteristics that tend to be macrocytic, and the size varies between 2 to 20 mm, and progressively increasing in general over time and are followed by fibrosis components.⁵

However, there are differences in both cases, where honeycomb appearance in the second case is more typical because it is focused on the anterior aspect of the superior lobe of the lung and followed by honeycombing in the pulmonary basal, matches with the definition of the anterior upper lobe sign.⁵ In the first case, honeycomb appearance is more dominant in the pulmonary basal and more minimal than the second case. This condition is influenced by the severity of the disease and the treatment of the patient. In the second patient, the patient's clinical condition is more severe and has not had time to undergo other diagnostic tests such as spirometry to determine the type of the treatment.

These results show that there is a variations in the HRCT images of the UIP, especially in the case of SSC-ILD accompanied with UIP. The presence of 3 signs such as exuberant honeycombing sign, anterior upper lobe sign, and straight-edge sign can improve the specificity of diagnosis.⁵ However, disease severity and progression might also influence radiological findings. The presence of honeycombing and traction bronchiectasis remains a major marker of UIP diagnosis.

Patient Consent Statement

Along with this letter, we would like to confirm that our patients have agreed that their medical history can be published as a scientific paper. In order to protect the patient's privacy, we did not include the physical appearance of the patient. Furthermore, we focused on imaging, so we did not violate any patient's privacy.

We hope that this letter is enough to comply the regulation of patient's privacy in this journal. Thank you and looking forward for your response.

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