中文題目：全身性硬化症的自發性氣胸

英文題目：Spontaneous pneumothorax in systemic scleroderma

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Background:

Systemic sclerosis (SSc) is a complex autoimmune disease of unknown etiology that can affect multiple organ systems, with lung involvement occurring in over two thirds of patients. Spontaneous pneumothorax (PTX) remains a rare pulmonary manifestation of scleroderma, occurring only in patients with advanced pulmonary fibrosis associated with subpleural cyst formation and rupture. Physicians should be aware of patients with spontaneous pneumothorax in scleroderma. Initial management consists of chest tube insertion, but recurrence is high and may require pleurodesis or partial lung resections for localized pulmonary lesions. Tube thoracostomy or pig-tail insertion is generally performed initially for most patients based on their clinical symptoms and severity of pneumothorax. Nevertheless, recurrence rate is especially high in patients with secondary spontaneous pneumothorax. Partial lung resection for localized lesions or chemical pleurodesis might be required for the sequential management.

Methods:

SSc is a complex autoimmune disease in which abnormal endothelial resulting in a small and medium-sized vessels vasculopathy, dysregulation of fibroblast resulting of excessive collagen production and fibrosis, and abnormalities of the immune system.[1] The interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) are the most common manifestations of SSc, and are now the leading causes of 60% death in SSc.[1] SSc is often associated with various respiratory complications, including chronic respiratory failure, small airway disease, neuromuscular weakness, pleural effusion , pneumothorax and lung cancer. [2] Spontaneous pneumothorax is often associated with causued to multiple thin-walled lung cysts and likely secondary to subpleural cyst rupture in advanced pulmonary fibrosis. [3] We’ve reviewed the clinical features of these previous cases and discuss the management of this uncommon manifestation.

Case Report

This 44-year-old male has medical history of gouty arthritis and rheumatoid arthritis

more than 10 years without regular controlled which four extremities deformity and ulna deviation. His family history was unremarkable. He presented to our facility with 10-years history of severe progressive systemic sclerosis and manifested as sclera-dermatous skin changes, high titer of Scl -70 Ab and antinuclear antibodies, Raynaud's phenomenon, sclerodactyly (Figure 1 & 2), esophageal motility dysfunction (Figure 3), pulmonary fibrosis, and moderate pulmonary hypertension. His symptoms exacerbated of multiple arthralgia and he had been treated with D-penicillamine, prednisone, but no improvement in his symptoms.

He presented to our hospital who sudden onset difficulty of breath and right chest pain at past 2 days with productive cough but denied fever or chillness. Physical examination revealed reduced air entry on the right side of chest. The hemogram data showed no leukocytosis. Chest radiography showed right pneumothorax (Figure 4) and chest roentgenogram revealed a 50% spontaneous pneumothorax on the right side with severe honeycombing of both lungs, but no herniation across the midline (Figure 5). Meanwhile, we are afraid of acute coronary syndrome easily confused with pneumothorax. It was sudden onset with chest pain and short of breathing.

The Echocardiography revealed the moderate pulmonary hypertension and *Sildenafil* Citrate 20 mg was suggest by cardiologist. The Thallium scan (Figure 6) found mild to moderate ischemia on apical and lateral wall. The cardiologist suggest arrange coronary angiography post spontaneous pneumothorax resolved.

The chest tube was placed in his right thorax and subsequent lung re-expansion under Triflow training, but it was worsen after 2 weeks. He then underwent successful pleurodesis with OK432 and his symptoms resolved. The chest tube was removed 2 days later, and discharged. The chest tube was placed in the right thorax and persistent air-leak was presented with small amount of residual pneumothorax for weeks. Through spirometry training, the right lung got re-expanded, then chemical pleurodesis with OK432 was given with a successful result. The chest tube was removed days later and the patient was discharged uneventfully.

Discussion

Spontaneous pneumothroax is a rare complication of SSc. Patient presents with symptoms of exertional dyspnea and dry cough, common in SSc whom develop pulmonary involvement. [4]

Subpleural cyst rupture was associated with bronchopulmonary fistula formation in severe interstitial lung disease and potential caused PTX. [5]

SSc-associated ILD, chest high-resolution computed tomography (HRCT) usually revealed ground-glass opacities, peri-pleural honeycomb lesion, cyst and inter-lobular septal thickness, especially over bilateral lower lobes. [6]

SSc with pulmonary fibrosis increase mortality rate from 6% to 33%.[7]
Clinically, we should alert about SSc patient present with CREST syndrome (**C**alcinosis cutis, **R**aynaud phenomenon, **E**sophageal dysmotility, **S**clerodactyly, and **T**elangiectasia) [8] and dyspnea with hypoxemia. The PTX and PAH were under our differential diagnosis.
Those biomarker of autoantibodies included of Anti-Scl-70 antibodies, Anti-topoisomerase I (ATAs) and anticentromere antibodies (ANAs) were widely used and more than 95% of SSc patient was diagnosis at first time. [9]
We could careful about those symptoms, the further biomarkers and HRCT were used to confirm about SSc at beginning.
The chest tube replacement should be performed at chest radiography was found of PTX. The following management included of tac or OK432 pleurodesis to treat PTX. It may require partial lobectomy for localized lesions if spontaneous pneumothorax recurs. [4]

Conclusions:

Those advance pulmonary fibrosis and cyst formation, spontaneous pneumothorax may be more prevalent than previously recognized. Initial management consists of chest tube placed, but high recurrence and may require pleurodesis or partial lobectomy. Physicians managing patients with scleroderma should be aware of spontaneous pneumothorax as a cause of acute shortness of breath in those patients with advanced pulmonary fibrosis and subpleural cysts.