Systemic sclerosis with hemoptysis and a huge lung cavity

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Abstract

Systemic sclerosis or scleroderma is associated with distal vasculitis, Raynaud's phenomenon, and inflammation of internal organs and the skin. We present on a 58-year-old Thai woman with systemic sclerosis who came to the 10th Zonal Tuberculosis and Chest Disease Center, Chiang Mai, Thailand in 2009 and presented with hemoptysis and a solitary huge-lung cavity as the predominant clinical manifestations which spontaneously resoluted 2 months later. This case demonstrates a solitary huge-lung cavity with hemoptysis and looked like from non-tuberculous Mycobacterial infections or malignancy with spontaneous resolution of hemoptysis and the lung cavity, which does not need invasive investigations.

Case Report

Hemoptysis for one year developed in a 58-year-old Thai woman with medical history of thickening of the skin, face, neck, and both upper extremities with flexion deformity of both hands (Figure 1), cool-exposure-cyanotic skin changes and difficulty in the mouth opening for 10 years presented to the 10th Zonal Tuberculosis and Chest Disease Center, Chiang Mai, Thailand in 2009. Ten years previous, she was performed the chest radiograph, high-resolution computed tomography (HRCT) of the chest, three consecutive sputum acid-fast bacilli (AFB) examinations, and bronchoscopic examinations which revealed reticular and small cystic infiltration with fibrotic change and some ground glass infiltration at the both lower lobes, more at the basal lungs and periphery of the upper lobes (Figure 2). Tubular bronchiectasis was seen at bilaterally perihilar regions while small cystic bronchiectasis was shown at the both lower lobes (Figure 2) without evidence of tuberculosis (TB) or other Mycobacterium species from sputum and bronchoscopic examinations. She has received a diagnosis of systemic sclerosis (SSc) with lung involvement without evidence of family history of SSc and has received prednisolone and plaquenil. Cyclophosphamide was added between 2000 and 2008 due to partially clinical response to prednisolone. Five-years follow-up of the chest radiograph and the HRCT of the chest still revealed reticulocystic lesions indicated bronchiectasis and bronchiolectasis, honeycombing, subpleural bands and septal thickening at the both lower lobes with new patchy-cystic lesion at the right midlung field, new small fibronodular infiltration at periphery of the posterior aspect of the both upper lobes and lower lobes, and new minimal right apical pleural thickening without pleural effusion while the mediastinum was within normal limits (Figure 3). Five years previous she was also performed the echocardiography due to dysnea on supine position for four years and revealed moderate mitral valve regurgitation with progression to severe form 1 year later without pleural effusion and pulmonary arterial hypertension (PAH). She has been added ferrous sulphate therapy. Other laboratory results were unremarkable. Two years previous she developed sclerodactyly of both hands with severe anemia. Increased dosage of ferrous sulphate had been prescribed for 2 years. She is a non-smoker. On physical examination, the patient's vital signs were within normal limits. There was mildly thickening of the skin of the face, neck, and extremities without dysnea. Evidence of sclerodactyly of both hands was noted.

Investigations

On the first day of the patient's presentation, we hypothesized that chronic receiving of prednisolone can allow development of pulmonary TB with presenting hemoptysis due to drug-induced immunosuppression. Thus, the three consecutive sputum examinations for the AFB were performed including sputum cultures for *Mycobacterium tuberculosis* and other Mycobacterium species, which revealed negative results. A chest radiograph showed a huge thin-wall cavity with 8 cm in diameter in the right lower lobe, bilaterally diffuse fibrotic infiltration, bilateral pleural effusion, and bilateral basal pleural thickening (Figure 4).

Discussion

SSc is systemic and complex collagen vascular disease of unknown etiology,[1] associated with excessive tissue fibrosis of the skin and various internal organs and small-vessel vasculopathy.[2,3] Numerous known agents induce SSc-like pulmonary disease; for examples, benzene, toluene, trichloroethylene, bleomycin, pentazocine, trytophan, and D-penicillamine[4] including silica[5] and rapeseed oil denatured with aniline.[6] The patient's one major criterion of thickening of both hands and two minor criteria of sclerodactyly of both hands and bilaterally basilar idiopathic pulmonary fibrosis (IPF) made the diagnosis of SSc.[7] She has had a history of Raynaud's phenomenon which is a common clinical manifestation (91%)[8] and more than 10 years of IPF which we hypothesized the early clinical sign which was found between 52%-70%.[8,9] Hemoptysis found in this patient without evidence of PAH, TB and D-penicillamine[10,11] and other known-induced agents[4-6] exposure is a rare complication which may complicate bronchial telangiectasia or carcinoma.[12] Wangkaew S. *et al.* reported their study among Thai SSc patients with 59.1% (39 of 275 patients) of PAH and 92.3% (36 of 275 patients) of dyspnea on exertion[13] while Panicheewa S. *et al.* reported of 43.3% with pulmonary involvement among Thai SSc patients.[14] Only 26.5% (22 of 83 Thai SSc patients) were diagnosed PAH by echocardiography.[15] A study reported in 2004 of only one case of complicated pulmonary TB among 11 patients with SSc-associated interstitial lung disease treated with azathioprine.[16] Serum concentrations of insulin-like growth factor (IGF)-1[17] and IGF binding protein,[17] interleukin-15[18] and chest ultrasonography[19] can early and simply predict the development of pulmonary telangiectasia[17] and IPF,[18,19] respectively which were not early done in this case. Fortunately, only 0.2% of cases with pulmonary hemorrhage or hemoptysis or both are regis-

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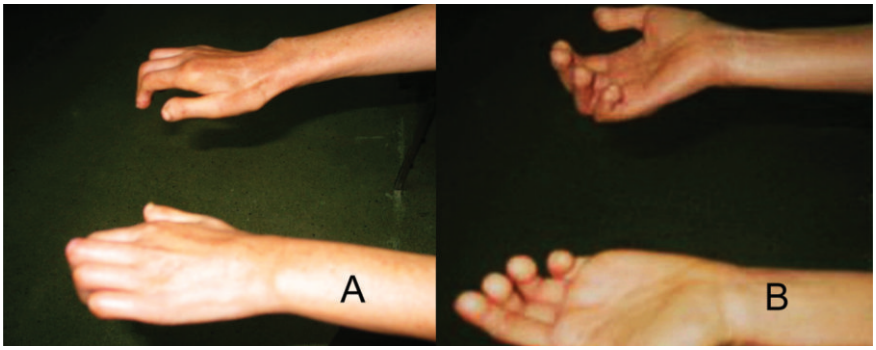

Figure 1. Showing the thickening of the skin of both upper extremities with sclerodactyly of both hands (A, B).

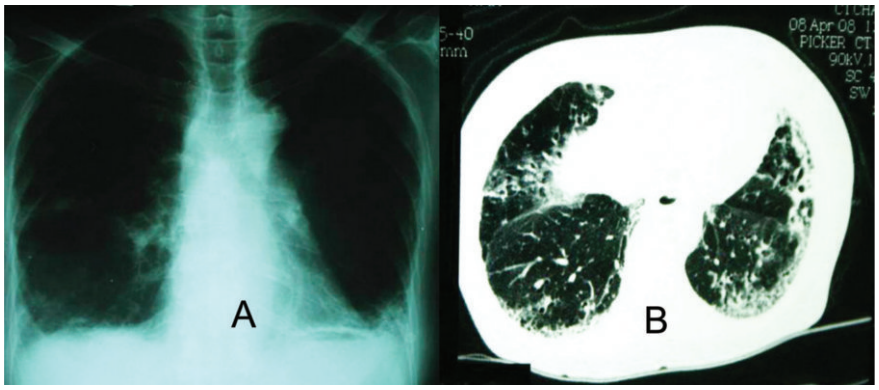

Figure 2. Posterior-anterior chest radiograph (A) and high-resolution computed tomography of the chest (B) at ten years previous showing reticular and small cystic infiltration with fibrotic change and some ground glass infiltration in the both lower lobes.

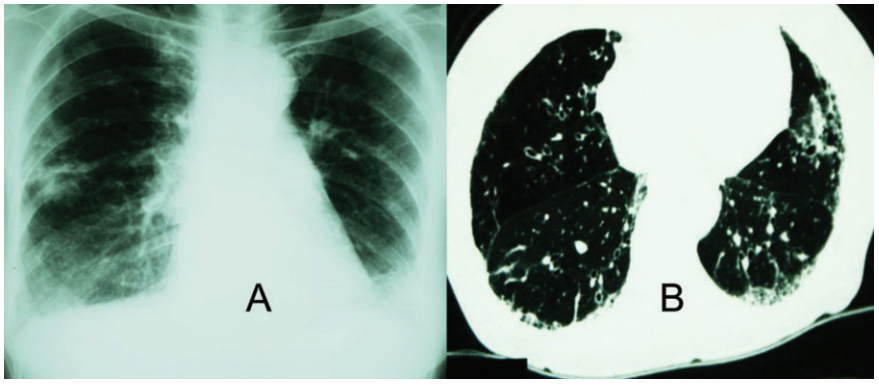

Figure 3. Posterior-anterior chest radiograph (A) and high-resolution computed tomography of the chest (B) at five years previous still showing brochiectasis and bronchiolectasis, honeycombing, subpleural bands and septal thickening in the both lower lobes.

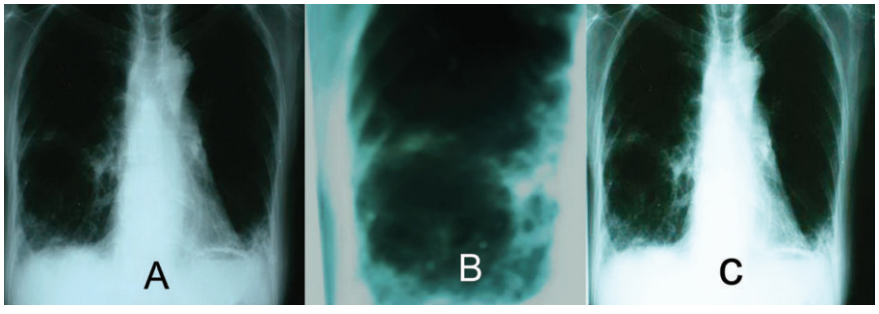

Figure 4. Posterior-anterior chest radiograph on the first day of the patient's presentation at our Center still showing bronchiectasis, honeycombing in the both lower lobes and bilaterally pleural effusion (A,B) with a new huge-thin-wall cavity in the right lower lobe (A,C), which spontaneously resolved 2 months later (C).

tered as a leading cause of death.[10] Honeycombing[20] of the lungs on the last chest HRCT indicated more advanced disease and serum anti-topoisomerase autoantibody[21,22] measurement could strongly predict the patient's 10-year previous IPF before progression to this pathological lung appearance. On the chest radiograph of the first attendance at this Center, it demonstrated a huge thin-wall cavity at the right lower lobe with acute pneumonitis at the right midlung field and bilaterally pleural effusion which could be SSc-associated-infective brochiectasis and this lung cavity spontaneously resolved 2 months later excepted the bilaterally pleural effusion (Figure 4). This lower lobe huge-solitary cavity may be complication of the SSc-associated-cystic bronchiectasis seen on the patient's 10-year previous chest radiograph and HRCT of the chest and it can cause hemoptysis which disappeared at the same time of this lung cavity resolution. The patient's 10-year and 5-year previous chest radiographs and HRCTs of the chest showed usual patterns of the disease with previously normal bronchoalveolar lavage examinations and only bilaterally minimal-pleural effusion on the last chest radiograph indicated not necessary to perform lung biopsy.[23] On the last attendance the patient revealed very dramatic clinical responses excepted the difficulty in open her mouth, thickening of the skin, and the Raynaud's phenomenon.

Conclusions

This case does not need invasive investigations for diagnosis. Our case highlights a huge lower-lobe-thin-wall cavity in SSc patient, which has been reported as SSc-associated non-tuberculous Mycobacterial pulmonary infections in the medical literature in Thailand. This case demonstrates the importance of the chest radiographic follow-up of the SSc-associated-lung cavity with hemoptysis instead of performing the lung biopsy or investigations for non-tuberculous Mycobacterial infections.

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