

Imaging of Thoracic Manifestations of Scleroderma

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Abstract

Radiographs illustrating the various thoracic manifestations of systemic sclerosis are shown. These include pulmonary parenchymal disease e.g. interstitial pneumonitis and pneumonia, pleural disease (pneumothorax), cardiac involvement, oesophageal disease, bony changes and soft tissue calcification. The use of chest radiography and other imaging modalities such as high resolution computerised tomography (HRCT) of the thorax is discussed.

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Introduction

Systemic sclerosis (SScl) is a generalised disease of the connective tissue characterised by fibrosis and vascular changes affecting the skin, lungs, gastrointestinal tract, kidneys, and heart.¹ The American College of Rheumatology has proposed that for the purpose of clinical trials and surveys, the diagnosis is made if the patient has either one major or two of the three minor criteria listed below:²

Major Criterion

1. Proximal scleroderma—skin thickening proximal to the metacarpal or metatarsophalangeal joints

Minor Criteria

2. Sclerodactyly—skin thickening of the fingers
3. Digital pitting scars
4. Bibasilar pulmonary fibrosis

Although Raynaud's phenomenon is not a criterion for diagnosis, it is seen in almost all patients and its absence should make a doctor suspect other causes of skin thickening e.g. mucinosis.

Distinct subsets of this condition are recognised:³

1. Diffuse Scleroderma—where there is generalised skin thickening (including truncal sclerosis) and various visceral complications such as pulmonary fibrosis and renal crisis. The characteristic autoantibody is Anti-Scl 70 or anti-topoisomerase I.
2. Limited Scleroderma—where the skin thickening is often limited to the fingers. Other features include

prominent Raynaud's phenomenon, oesophageal hypomotility, telangiectasia and calcinosis. Synonym—CREST. The characteristic autoantibody is the Anti-Centromere antibody.

3. Overlap Syndrome—where the patient has clinical features fulfilling criteria for other rheumatic disease (e.g. systemic lupus erythematosus or polymyositis) in addition to features fulfilling criteria for systemic sclerosis.
4. Scleroderma sine Scleroderma—where the patient has no sclerosis of the skin but has characteristic visceral sclerosis e.g. pulmonary fibrosis.
5. Undifferentiated Connective Tissue Disease—where the patient is in a "pre-scleroderma" state with Raynaud's phenomenon and serologies of SScl but without fulfilling criteria for the condition.

Thoracic Manifestations of Systemic Sclerosis

The thoracic manifestations of SScl include:

pulmonary parenchyma

pulmonary fibrosis
pulmonary infections including lung abscess
lung cancer

pleural

spontaneous pneumothorax
pleural effusions

pulmonary vascular disease

primary pulmonary hypertension

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cardiac

pericardial
myocardial disease
conduction abnormalities

oesophagus

hypomotility and reflux

mediastinal

adenopathy

bony

bony resorption

soft tissues

calcification

Pulmonary Fibrosis

It is often reported that the lungs are the second most frequently affected viscera (after the oesophagus) in patients with SScl.⁴ In Singapore, interstitial lung disease was most frequently affected (56.2% of patients with SScl) while gastrointestinal involvement was second (42.5%).⁵ Pulmonary involvement has been shown in up to 100% of autopsies.⁶

The chest radiograph is an essential part of the routine multi-system evaluation of all patients with SScl. It is widely available, easy to perform and inexpensive. The chest radiograph may appear normal in very early cases. In advanced cases the lung volumes are small and there are interstitial opacities which predominate in the lower zones and the peripheral lung fields.

Plain radiography is however, inferior to computed tomography (CT) of the thorax [both standard and especially the high resolution computerised tomography (HRCT)] in showing early pulmonary abnormalities especially in asymptomatic patients. In a study by Schurawitzki et al,⁷ HRCT showed interstitial lung disease in 91% of patients compared to 39% of definite interstitial shadowing demonstrated by chest radiography. HRCT changes of interstitial lung disease in SScl

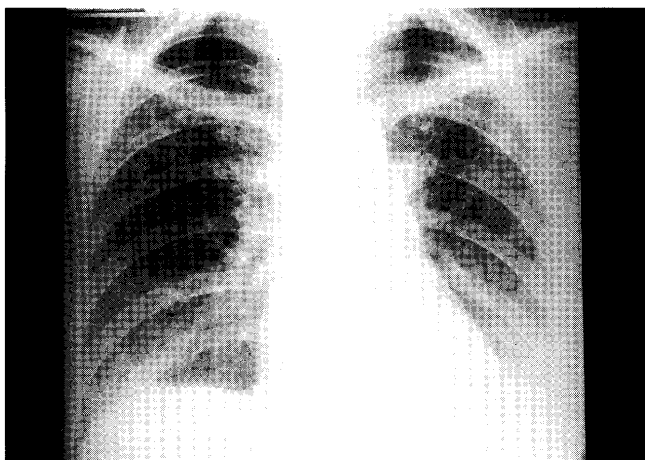


Fig. 1. Both lungs were slightly reduced in volume. There was reticular shadowing at both lung bases and more ill-defined shadowing in the upper/mid zones.

are similar to the changes seen in fibrosing alveolitis. These include ground glass opacities, parenchymal micronodules, linear opacities, subpleural micronodules and honeycombing.⁸ The lower lobes and peripheral and posterior lungs are most severely affected. The significance of ground glass opacities in scleroderma differs from that seen in fibrosing alveolitis where it is reportedly a good predictor of response to therapy; in scleroderma, ground glass opacities are thought to reflect fine fibrosis and may herald the development of honeycombing. In the scleroderma sine scleroderma subset, dyspnoea and abnormal lung function may be the presenting feature with minimal or no cutaneous involvement at initial presentation. These patients demonstrate interstitial pneumonitis and fibrosis and later develop skin manifestations.⁹

Case 1

AYH was a 46-year-old Chinese female with Overlap syndrome. She had clinical features of systemic lupus erythematosus (SLE) as well as SScl. Seven years after the initial presentation despite the absence of respiratory symptoms her chest X-ray (CXR) showed pulmonary fibrosis. The HRCT shows the changes in greater detail (Figs. 1, 2a & 2b).

Pulmonary Infections

Pulmonary infections are not uncommon in patients with SScl. Recurrent aspiration from reflux oesophagitis is an important cause of pneumonia. Malnutrition from chronic small bowel involvement can weaken immune function. The use of steroids and immunosuppressants further increase vulnerability to all types of infection. Lung abscess can complicate a severe lung infection. The radiological features of pneumonia are as in any other pneumonias. Sometimes the presence of underlying in-



Figs. 2a & b. HRCT performed one month after the chest radiograph. The upper zones were clearly abnormal with subpleural cysts (short arrow), honeycombing (long arrow), and marked thickening of the superior end of the oblique fissure on the right (broad arrow). At the bases (fig. 2b) there was marked honeycombing.



Figs. 2b.



Fig. 3. A chest radiograph taken in March 1993 showing decreased lung volumes and basal cystic lucencies compatible with honeycombing.

terstitial lung disease can confound interpretations of the chest radiograph. A follow-up film after adequate antibiotic therapy can help.

Case 2

PKY, a 63-year-old Chinese male with scleroderma of the diffuse variant had complications of pulmonary fibrosis as well as oesophageal hypomotility after 9 years of disease. Three years later, he was hospitalised for severe pneumonia and died despite aggressive treatment (Figs. 3 & 4).

Neoplasm

Lung cancer is a well described complication of SScl and it is postulated that pulmonary fibrosis predisposes to the development of lung cancer. An increased incidence of lung cancer in SScl patients which occurs in the setting of long-standing pulmonary fibrosis has been reported.^{10,11} A similar increased incidence of lung cancer was also seen in idiopathic pulmonary fibrosis.¹² Early studies reported that bronchioloalveolar type was the most prevalent,^{13,14} however, later studies showed a variety of histologic types but not bronchioloalveolar



Fig. 4. A chest radiograph three years later when the patient was admitted to hospital. There were patchy bilateral airspace opacities in the mid and lower zones compatible with aspiration pneumonia superimposed on basal honeycombing. There were small bilateral pleural effusions. A central venous catheter and endotracheal tube have been inserted.

type carcinoma.^{10,11} It is interesting to note that in the patient discussed below, there were only minimal fibrotic changes seen on the CT studies. However, this may be partly due to scan technique which was standard (10 mm collimation) and not high resolution (1 to 1.5 mm collimation) which would optimise visualisation of interstitial disease.

Case 3

LLH, a 68-year-old Chinese male had SScl of the diffuse variant for 20 years. He was hospitalised with a cavitary lung lesion. Barium swallow and meal showed gross gastroesophageal reflux and the clinical impression was that of a lung abscess complicating chronic aspiration. A CT of the thorax was done 2 weeks later when he did not respond to aggressive antibiotic therapy. This showed a right-sided cavitated mass and consolidation. There was also a large amount of mediastinal adenopathy and abnormal soft tissue surrounding the right lower lobe bronchus. Bronchoscopic biopsy showed a squamous cell carcinoma (Figs. 5 to 7).

Pleural Disease

Pleural reaction is more often asymptomatic and found at autopsy than is evident clinically. Pleural effusions can occur with cardiac failure from myocardial or secondary to renal involvement by SScl.⁴ Spontaneous pneumothorax in SScl was reported in 1954¹⁵ and bilateral pneumothoraces can also occur.¹⁶ These are secondary to rupture of subpleural cysts.

Case 4

LLH, a 60-year-old Chinese female had SScl of the diffuse variant for 25 years with pulmonary fibrosis. In July 1996 she was admitted to hospital for aspiration pneumonia. Two months later, she was re-admitted with cough and breathlessness. Chest radiography is as

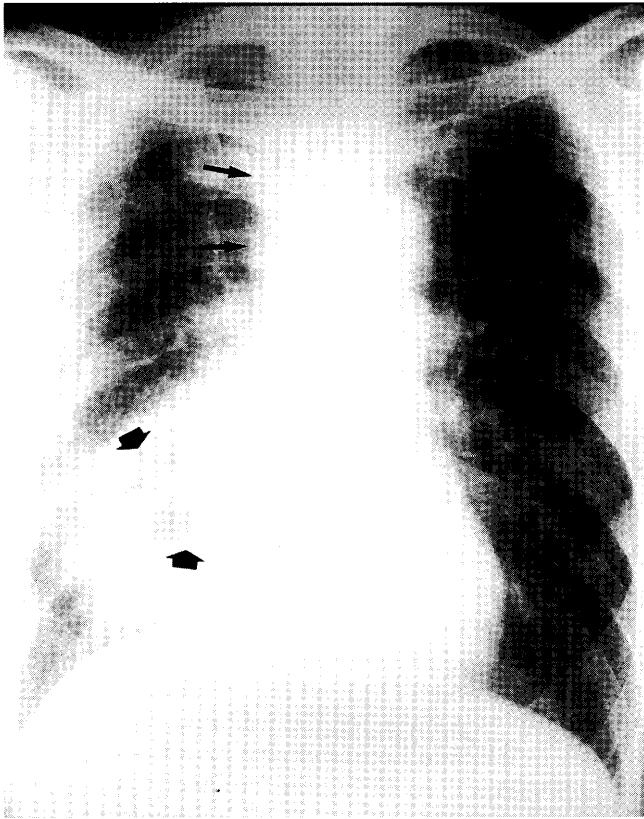


Fig. 5. Admission chest radiograph showing a right lower lobe cavity (broad arrows) with surrounding pulmonary consolidation. There was slight widening and lobulation of the right paratracheal stripe (thin arrows) compatible with adenopathy.

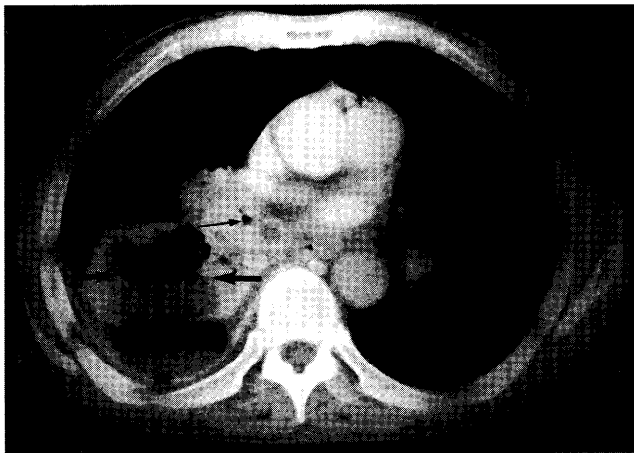


Fig. 7. Thoracic CT mediastinal setting demonstrating tumour surrounding the right lower lobe bronchus (thin arrow), cavity (arrows), and a loculated hydropneumothorax (broad arrow).

shown in Figure 8. She was treated conservatively and was discharged home one week later.

Pulmonary Vascular Disease

Pulmonary arterial hypertension (PAH) is an important complication of SScl. It can occur secondary to pulmonary fibrosis or to a direct vasculopathy from intimal fibroelastosis.¹⁷ Patients with the limited variant of scleroderma are particularly prone to the develop-



Fig. 6. Gastroesophageal reflux seen on a barium study. The oesophagus was noted to be aperistaltic (there were no gastroparetic agents used).

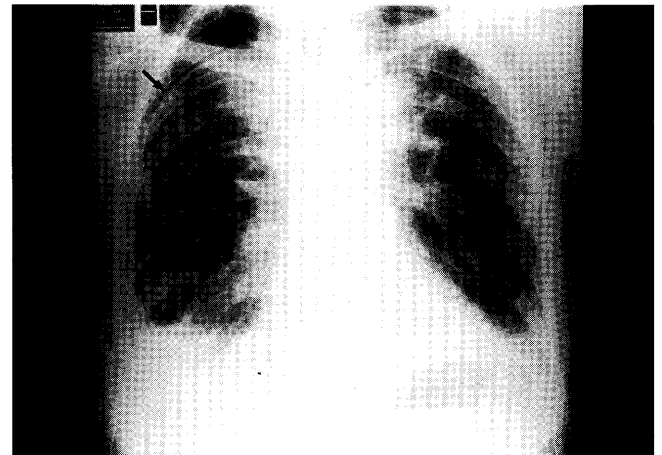


Fig. 8. Chest film showing a right-sided pneumothorax (arrow) with bilateral mild pleural reaction. There was background fibrotic change.

ment of primary pulmonary hypertension.¹⁷⁻¹⁹ The incidence of pulmonary vascular disease in scleroderma varies in different studies due to the different methods used to detect PAH. In Ungerer et al's study¹⁸ of 49 patients with scleroderma, 33% were found to have PAH by right heart catheterisation. Of the 10 patients in the study with limited disease, 50% had PAH. This study

demonstrated that the chest radiograph is the least sensitive but the most specific non-invasive method of demonstrating PAH based on the size of the right descending pulmonary artery. This measurement was described by Chang.²⁰ However, the size of the descending pulmonary artery is often difficult to measure from frontal radiographs because of vascular overlap and obscuration by the right cardiac border. Hence, measurement of the pulmonary arterial size by CT may be more useful. Kuriyama et al²¹ showed that CT could be used as a reliable indicator of pulmonary artery hypertension. Of the six sites studied, the highest positive correlation between pulmonary artery pressure and artery diameter was at the main pulmonary artery; its upper limit was found to be 28.6 mm. An abnormally low diffusing capacity (DLCO) of below 45%²⁰ and 43%¹⁸ is a sensitive indicator for PAH. Patients with limited disease and isolated PAH had a poor prognosis with a 2-year cumulative survival rate of 40%.¹⁹

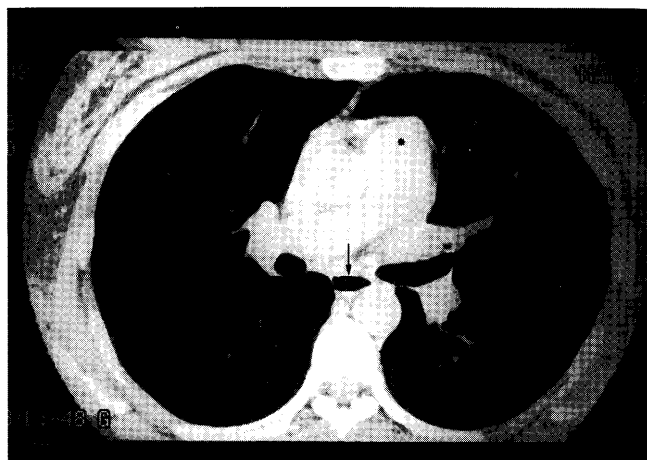


Fig. 9. A baseline HRCT of the thorax at the level of the main pulmonary trunk showing a mildly dilated oesophagus (arrow). The diameter of the main pulmonary artery (*) was slightly enlarged at 30 mm. The chest radiograph obtained at this time was normal (not shown).

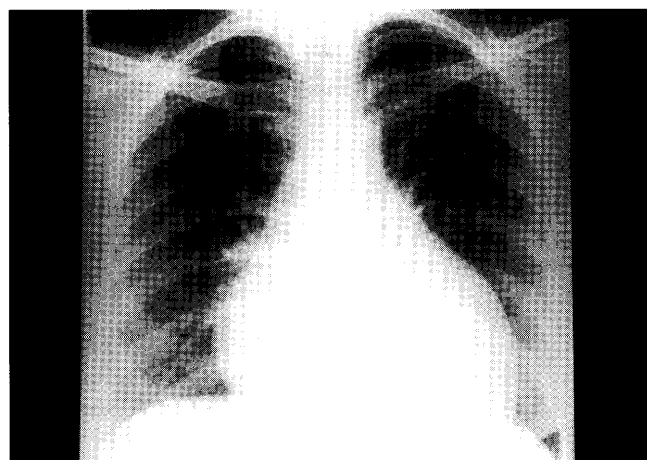


Fig. 10. A chest X-ray taken when the patient was acutely dyspnoeic showed moderate cardiomegaly. The right descending pulmonary artery was obscured by the right heart border. The lungs were clear.

Case 5

TCL, a 42-year-old Chinese female was diagnosed to have SScl of the limited variant in July 1995. Seven months later her DLCO fell to 28% of the predicted value but HRCT was unremarkable apart from a slightly enlarged main pulmonary artery and mild oesophageal dilatation. Another 7 months passed and she was hospitalised with pulmonary arterial hypertension. A chest radiograph obtained at this time demonstrated moderate cardiomegaly due to right ventricular enlargement. PAH was confirmed on 2D echocardiography. With treatment she improved and the heart size returned to normal on CXR. However, the diameter of the descending pulmonary artery remained enlarged indicating persisting pulmonary arterial hypertension (Figs. 9 to 11).

Oesophageal Hypomotility and Reflux

Oesophageal hypomotility and incompetence of the lower oesophageal sphincter often leads to reflux of the gastric contents. The oesophagus can be studied by manometry or imaged by a barium swallow (Fig. 6). It is particularly important for the referring physician to indicate to the radiologist performing a barium study that oesophageal motility is to be evaluated so that gastro and oesophageal parietic agents which are routinely given will be omitted. The oesophagus is also demonstrated on CT of the thorax (Fig. 9). A study of 25 SScl patients using CT showed that 80% had oesophageal dilatation despite the absence of symptoms. The

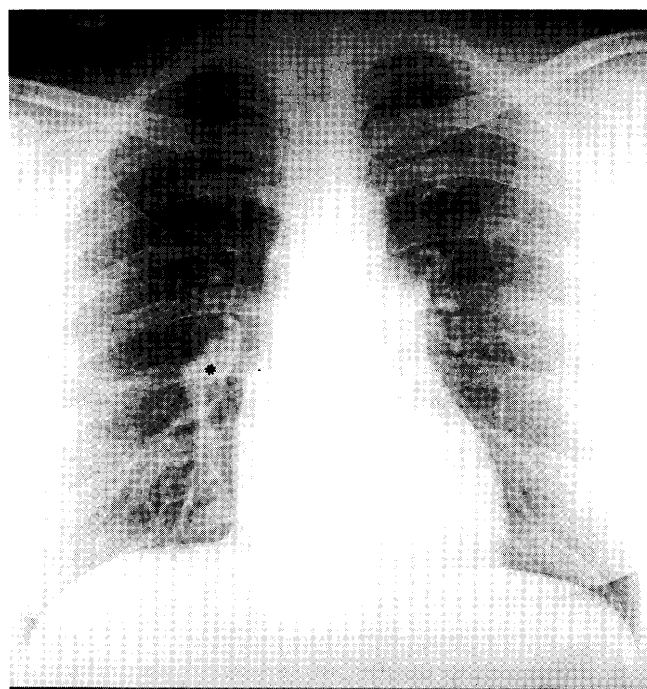


Fig. 11. Following treatment, two months after figure 9 the heart size returned to normal but the descending pulmonary artery (*) was enlarged at 20 mm (normal range 9 to 15 mm according to Chang).

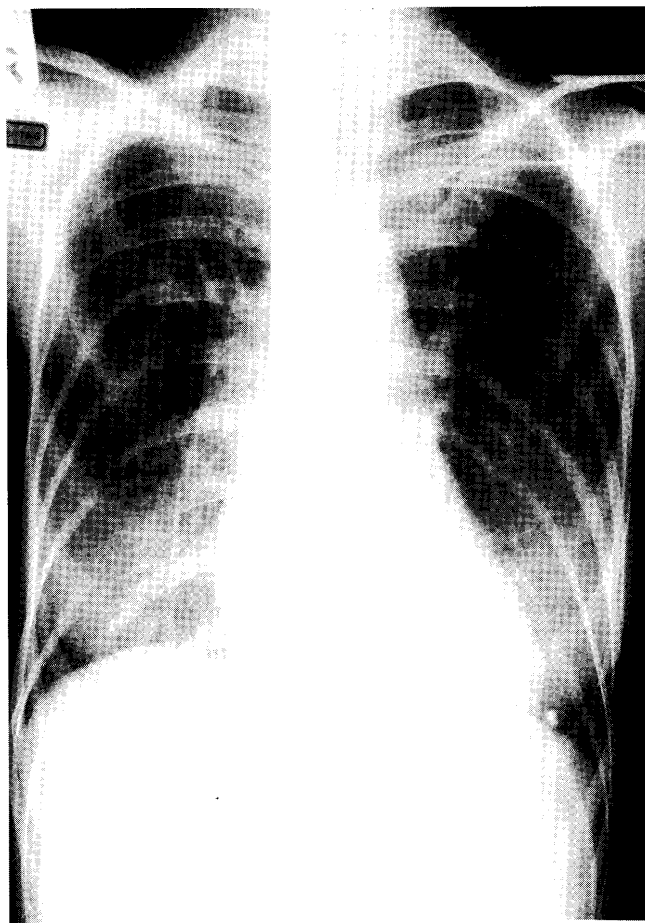


Fig. 12. Chest radiograph showing marked thinning of the ribs especially at their superior aspects. There was also resorption of the superior aspect of the mid clavicles.

oesophagus measuring more than 10 mm in the coronal plane on four or more consecutive axial slices below the aortic arch was considered to be dilated.²²

Lymphadenopathy

Mediastinal adenopathy per se can occur in SScl in the absence of complicating infection or tumour. Bhalla et al²² found adenopathy in 60% of SScl patients with interstitial lung disease on CT examination. Thoracic lymphadenopathy can also be due to tumour (Fig. 5).

Bony Resorption

Rib resorption was first described by Steigerwald et al in 1975.²³ Initially thought to be rarely seen, a later study by Owens et al²⁴ showed an incidence of 16% in diffuse disease. It is exclusively seen in diffuse disease. An increased incidence of rib notching was associated with terminal tuft resorption.²⁵ The cause of bony resorption is unclear.

Case 6

This 38-year-old lady has SScl of the diffuse variant for 12 years. These chest radiographs were taken 12 years



Fig. 13. Radiograph of the mandible showing marked thinning of the left ramus with pathological fracture (arrow).

after diagnosis (Figs. 12 & 13). She has very advanced disease with contractures and severe bone resorption.

Ectopic Calcification

Calcified granulomata are seen in both limited as well as diffuse SScl. A greater incidence is seen in the limited variant 67% as compared to 14% in diffuse disease.²⁴ It is not associated with an increased frequency of subcutaneous calcinosis²⁵ suggesting a different mechanism for the 2 types of calcium deposition.

Conclusion

This pictorial essay illustrates the various thoracic manifestations of scleroderma. Chest radiography still plays an important role in the screening and monitoring of patients with this disease. Other modalities especially HRCT of the thorax are valuable adjuncts.

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