Case 9074

Scleroderma: A focus on pulmonary findings
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Section: Chest imaging
Area of Interest: Oesophagus Lung
Imaging Technique: Digital radiography
Imaging Technique: CT
Imaging Technique: CT-High Resolution
Special Focus: Dilatation Case Type: Clinical Cases
Patient: 58 years, female

Clinical History:

A 58-year-old woman with chronic dyspnoea and reflux undergoes evaluation for worsening shortness of breath. The patient has a chronic medical condition and is on immunosuppressive medication. Chest radiograph, CT, and high-resolution CT (HRCT) are performed.

Imaging Findings:

The chest radiograph shows increased interstitial markings and decreased lung volumes. Associated esophageal dilation with an air-fluid level, as seen in this case (Fig. 1), suggests the diagnosis of scleroderma. Colonic dilation, an additional manifestation of the disease’s smooth muscle involvement, is also seen (Fig 1). Although not appreciated on the chest radiograph, pulmonary arterial dilation from pulmonary hypertension is seen on CT (Fig. 2b).

HRCT shows the classic pulmonary findings in scleroderma, which consist of a non-specific interstitial pneumonia (NSIP) pattern. These findings are most apparent in the lower lobes and include reticulonodular interlobular septal thickening, subpleural ground-glass opacities, and traction bronchiectasis (Fig. 3a-c). There is notable absence of pleural-based honeycombing, which is seen in usual interstitial pneumonia (UIP) / idiopathic pulmonary fibrosis (IPF), and not NSIP. Predominant involvement of the lower lobes, with extensive volume loss, can be appreciated on the coronal reformat (Fig. 4). Esophageal dilation is also noted (Fig. 2a, b and Fig 3c).

Discussion:

Scleroderma is a collagen vascular disease of unknown etiology that leads to inflammation and fibrosis in multiple organ systems including the skin, GI tract, heart, and lungs. Pathogenesis involves inflammatory mediators from alveolar macrophages causing fibrotic changes in the lung. Similar to other immune-mediated diseases, it presents most commonly in middle-aged women. Two main variants are the limited and systemic forms, with the limited form obsoleted described as CREST syndrome (Calcinosis, Raynaud phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia) [1]. Clinical pulmonary findings include dyspnea on exertion and dry cough.

The most common radiographic findings in scleroderma are those of the respiratory tract. Interstitial lung disease occurs in approximately 80% of patients and pulmonary hypertension occurs in up to 50% of patients [2]. Pulmonary
Radiographic findings include a basilar irregular reticular or reticulonodular pattern with interlobular septal thickening, ground-glass opacities, traction bronchiectasis, and pleural thickening. High resolution CT (HRCT) in scleroderma more closely resembles non-specific interstitial pneumonia (NSIP) with pulmonary fibrosis that is less coarse with more ground glass opacification compared to usual interstitial pneumonia (UIP) / idiopathic pulmonary fibrosis (IPF). Scleroderma rarely has multiple subpleural cysts termed “honeycombing” or findings of upper lobe fibrosis characteristic of UIP. Histopathologic and survival data in scleroderma patients with pulmonary findings also more closely resemble those seen in patients with idiopathic NSIP rather than UIP/ IPF. [3]

HRCT is superior to chest radiograph in detecting pulmonary findings; however, the degree of fibrosis does not correlate to clinical presentation [4]. There is asymptomatic esophageal dilatation in 80% of cases and mediastinal adenopathy in 60% [5]. Subpleural cysts, seen in the lower lobes, can cause spontaneous pneumothorax. Alveolar infiltrates are occasionally seen and can be secondary to aspiration from esophageal dysmotility.

Differential diagnosis includes other collagen vascular diseases, such as systemic lupus erythematosus (SLE), rheumatoid arthritis, Sjogren’s syndrome, dermatomyositis-polymyositis, and mixed connective tissue disease. Lung disease secondary to rheumatoid arthritis is associated with air trapping, mosaic perfusion, and cavitating subpleural rounded opacities. Lung disease associated with SLE is more frequently seen with pleural and pericardial effusions. Less likely differentials include asbestosis which would have calcific pleural plaques with less ground-glass opacities and bronchiectases; or drug-toxicity which would have more ground-glass opacities [6].

Clinical diagnosis is supported with serologic markers of specific autoantibodies, bronchoalveolar lavage, and rarely, lung biopsy. Patients with scleroderma have increased risk of lung cancer [7]. The disease progresses towards fibrosis and respiratory failure. Steroids and other immunosuppressants are used to manage disease progression which may lead to further pulmonary complications of drug-toxicity and opportunistic infections. Pulmonary disease has become the leading cause of death in scleroderma patients with the advent of improvements in the treatment of renal complications [8].

Teaching Point:
Radiographic findings of a dilated esophagus and HRCT findings consistent with an NSIP pattern are suggestive of scleroderma.

Differential Diagnosis List: Scleroderma, Systemic sclerosis, Systemic lupus erythematosus (SLE), Rheumatoid arthritis, Sjogren’s syndrome, Dermatomyositis-Polymyositis, Mixed connective tissue disease, Nonspecific interstitial pneumonitis (NSIP), Usual interstitial pneumonia (UIP), Asbestosis, Drug-toxicity

Final Diagnosis: Scleroderma, Systemic sclerosis

References:
Maffessanti M, Dalpiaz G, eds. (2006) Diffuse Lung Diseases: Clinical Features, Pathology, HRCT. Springer, Milan,
Italy
Description: Posteroanterior chest radiograph shows a large, dilated, air-filled esophagus, reticular interstitial lung markings, bilateral lower lobe volume loss, and dilated loops of bowel. No enlargement of the pulmonary arteries can be appreciated. Origin:
Description: The 5 mm thick coronal reformat shows peripheral, subpleural groundglass opacity, prominent reticular markings, and traction bronchiectasis with lower lobe predominance consistent with nonspecific interstitial pneumonia (NSIP) and scleroderma. Origin:
Description: Sequential 1.25mm axial HRCT images. Peripheral, subpleural groundglass opacity, prominent reticular markings, and traction bronchiectases are seen with lower lobe predominance consistent with nonspecific interstitial pneumonia (NSIP) and patient's history of scleroderma. Origin:
Description: Sequential 1.25mm axial HRCT images. Peripheral, subpleural groundglass opacity, prominent reticular markings, and traction bronchiectases are seen with lower lobe predominance consistent with nonspecific interstitial pneumonia (NSIP) and patient’s history of scleroderma. Origin:
Description: Sequential 1.25mm axial HRCT images. Peripheral, subpleural groundglass opacity, prominent reticular markings, and traction bronchiectases are seen with lower lobe predominance consistent with nonspecific interstitial pneumonia (NSIP) and patient’s history of scleroderma. Origin:
Figure 4

a

Description: Initial CT on lung windows, at the level of the aortic arch demonstrates a dilated oesophagus with an air-fluid level and normal appearing lung parenchyma. Origin:

b

Description: Soft tissue windows demonstrate mild dilation of the pulmonary arteries. This is evidenced by the diameter of the main pulmonary artery equaling the adjacent aortic root. Air-fluid level in the oesophagus is seen. Origin: