Radiological Evaluation of Systemic Sclerosis with Different Imaging Modalities

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Learning objectives

The collagen vascular diseases are autoimmune disorders which damage the connective tissue at different parts in the body. Systemic Sclerosis is a Connective tissue disease of unknown etiology. Our aim is to present a thorough review of the role of radiology in the assessment of systemic sclerosis and to describe the characteristic features of different imaging modalities in this disease.
Background

Systemic sclerosis or scleroderma is an unusual autoimmune disease which affects the skin, gastrointestinal tract, lungs, heart, musculoskeletal system, and peripheral circulation. ACR preliminary criteria for the classification of systemic sclerosis (The patient should meet the major criterion or 2 of the 3 minor criterias): Major criterion is proximal sclerodermatous skin changes (proximal to the metacarpophalangeal joints). Minor criterias are sclerodactyly, digital pitting scars of fingertips or loss of substance of the distal finger pad and bilateral basilar pulmonary fibrosis.

Gastrointestinal tract is the second most commonly involved system after skin. Pulmonary artery hypertension is the major cause of mortality in systemic sclerosis. This systemic disease may also affect the soft tissues, joints and bones. Radiology plays an integral part in diagnosis and management of this entity.
Findings and procedure details

In this review, we discuss the role of different imaging modalities such as upper and lower gastrointestinal tract radiography (barium enema), conventional enteroclysis, abdomen CT, high resolution thoracic CT, cardiac and extremity MRI, in the diagnostic management of systemic sclerosis.

Skin involvement

It is the most commonly involved organ. In systemic sclerosis 40% of patients with limited disease and a smaller fraction of those with diffuse disease are effected. Ultrasonography is a useful radiologic method which provides quantitative values for thickness of skin. The affected hypoechoic areas can be detected by this modality.

Gastrointestinal tract involvement

In systemic sclerosis, smooth muscle atrophy and fibrosis causes reduced primary peristaltic wave (fig 1). Also, stasis may predispose to candidiasis in esophagus. Because lower oesophageal sphincter is dilated, reflux may occur. Patients may have dysphagia or odynophagia. It may lead to esophagitis. Barrett's metaplasia may occur in one third of patients with systemic sclerosis. Patients with Barrett's metaplasia have an increased risk of stricture or adenocarcinoma (fig 2).

The potential appearances in the stomach and duodenum are distention and reduced motility.

Smooth muscle atrophy may effect motility of small bowel as seen with the esophageous.

Possible manifestations in small bowel are dilatation and prolonged transit time (fig 3). Anorexia, nausea, and vomiting are expected symptoms.

In systemic sclerosis, features of the affected colon are dilatation, haustration loss and wide-mouthed diverticulas (fig 4). Malabsorption may occur secondary to bacterial overgrowth.

Lung involvement

Chest radiography may appear normal or show only minimally changes. Finding of interstitial fibrosis (reticular-nodular pattern) has been reported in 20%-65% of patients. HRCT usually demonstrates evidence of fibrosis and interstitial pneumonitis in 44% of patients with normal chest radiography. Interstitial lung disease, pulmonary hypertension (major cause of mortality in these patients) and aspiration pneumonia, pulmonary haemorrhage, pleural effusions, organizing pneumonia, pulmonary embolism, thickening of pleura are common manifestations of respiratory system involvement.
Up to one third of patients with systemic sclerosis-associated interstitial lung disease exhibit usual interstitial pneumonia changes with a typically coarse pattern of reticulation and honeycombing with a little ground glass opacification (fig 5,6).

**Cardiovascular involvement**

Cardiovascular magnetic resonance imaging is a noninvasive technique without radiation which provides information about ejection fraction, myocardial fibrosis, congenital heart diseases, tumors, iron overload, myocardial infarction detection and follow-up, anomalous coronary arteries and myocarditis detection.

Patients with cardiovascular involvement have a bad prognosis. The prevalence of cardiac manifestations in systemic sclerosis is 10%, including pericardial involvement, arrhythmia, ventricular disease, congestive heart failure.

Cardiac MRI has been proposed as a non-invasive method of detecting.

**Musculoskeletal system involvement**

In systemic sclerosis myopathy or myositis occur in 70-96% of patients. Myositis is usually bilateral and symmetric, increased signal intensity on T2-weighted MR images in pelvic and thigh musculature is common.

Dystrophic calcification and contractures are common manifestations in systemic sclerosis. Dystrophic calcification may affect the hands, paraspinal soft tissues, tendons, joints, intervertebral discs. Soft-tissue atrophy is another appearance of systemic sclerosis.

In CREST syndrome, there are tumor-like deposits (fig 5,6). On magnetic resonance imaging, they have low signal intensity on T1 and heterogeneous signal on T2-weighted images.

Acroosteolysis (distal phalangeal resorption) is the characteristic bone involvement in systemic sclerosis which usually occurs in the hands (27% of cases).

On MR images, distal resorption is easily demonstrated; because fibrous tissue replaces the resorbed bone, low-signal intensity is observed and the residual bone does not show bone marrow signal intensity changes.
Fig. 2: Adenocarcinoma with irregular narrowing in the thoracic esophagus arising from Barrett's epithelium

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Fig. 4: Dilatation, haustration loss and wide-mouthed diverticulas in affected colon.

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Fig. 5: Reticulation and honeycombing with a little ground glass opacification in thorax CT

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Fig. 6: Reticulation and traction bronchiectasis in thorax CT.

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Fig. 7: Coronal T1 weighted MRI scan demonstrating the bilateral calcifications

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Fig. 8: Bilateral calcifications in pelvis ap x-ray.

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Fig. 1: Esophagus is dilated and has a fine reticulonodular mucosal pattern.

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Fig. 3: Enteroclysis showing dilated jejunum with reduced peristaltic activity.

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Conclusion

Herein we aimed to ensure a comprehensive review of the imaging workup of systemic sclerosis which is composed of heterogeneous group of disorders.
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