

Systemic Sclerosis

A case report with review of literature.

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Abstract

Scleroderma is a multisystem, autoimmune disease disorder, affecting young females, involving the immune system , blood vessels and connective tissue. Dermal manifestations include stiff, tight, shiny skin usually of the hands and feet due to swelling and thickening of the connective tissue as they become fibrotic or scarred. It may be localized (SSc) i.e, affecting extremities beyond elbow , tibia and face ,or progressive systemic sclerosis (PSSc). We present a case report of progressive systemic sclerosis in a 28-year-old female patient with characteristic systemic and oral manifestations.

Keywords: PSSc - Progressive Systemic Sclerosis, SSc - Scleroderma.

Introduction

Scleroderma comes from a greek word “sclero” meaning hard and “derma” meaning skin.

SSc is a multisystem, autoimmune disease affecting small arteries, micro vessels and fibroblast resulting in vascular obliteration, collagen accumulation, and scarring of the skin and internal organs such as gastrointestinal tract, lungs, heart, and kidneys. SSc is otherwise called Hidebound disease because of hide bound skin.

Case report

28 year old female presented to our department for barium swallow examination for dysphagia since one week.

The patient gave a history of loss of appetite and blue coloration of fingertips as a response to cold.

The patient was poorly built and nourished. General physical examination revealed tightening of the skin of extremities, claw-like the appearance of hands, extra oral examination revealed smooth, taut mask-like appearance of the facial skin and atrophied nasal alae giving rise to mouse facies appearance with loss of facial wrinkles.

Intraoral periapical radiographs revealed widening of periodontal ligament (PDL) space in the molars.

Anteroposterior view of hands Figure 1 and feet [Figure normal] revealed shortening and resorption of terminal

phalanges with ulcers and discoloration. Esophageal constriction could be appreciated in barium swallow, with complete esophago-gram with loss of primary and secondary peristalsis. BMFT did not show any of the abnormality like dilatation pseudo-obstruction, hide bound or stack of coin sign, sacculatation nor diverticula. Except long transit time in the stomach. HRCT lung and MRI Brain were normal.

Based on clinical examination hidebound skin, acro-osteolysis and esophageal changes, the diagnosis of progressive systemic sclerosis was made and presented a rare case.



Fig. 1: Claw like appearance of right hand with acro-osteolysis in both hands.

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Fig. 2: Taut mask like and mouse facies appearance



Fig. 3: Reduced mouth opening.

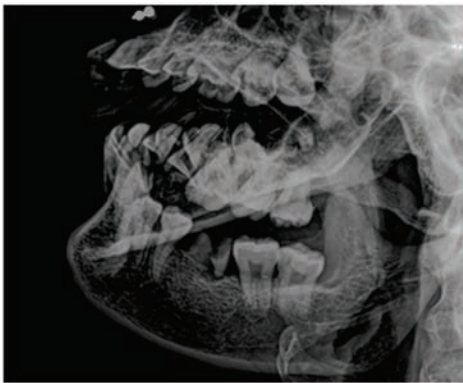


Fig. 4: Intra oral periapical radiograph of region reveals periodontal ligament space widening

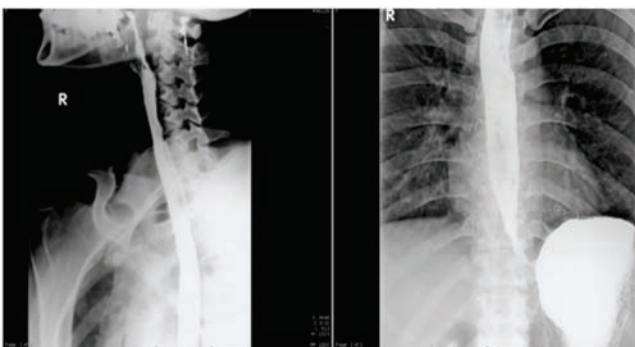


Fig. 5: Barium swallow radiograph reveals complete oesophagogram and esophageal constriction at lower end and on fluoroscopy no peristalsis noted.

Discussion

Curzio of Naples in 1752 reported the first case of PSSc. The name PSSc was proposed by Goetz in 1945 [1], when systemic nature of the disease was proven. SSc has a worldwide distribution and affects all races. Factors such as age, sex, genetic background, and environmental exposure may influence its susceptibility. Resorption of the terminal phalanges, short and claw-like fingers because of acro-osteolysis, ulcers on finger tips are common in patients with scleroderma fig 1. Dysphagia and gastroesophageal reflux are frequently reported complaints in this group of patients as in our case [4].

widening of the periodontal space, decrease in facial wrinkles owing to fibrosis of skin fig and orofacial telangiectasia, resorption of mandibular angle, Excessive collagen deposition in the cutaneous tissues a Fibrosis of salivary glands leads to xerostomia, dysphagia and subsequently periodontal infections round the mouth causes microstomia which prevents the patient from opening and closing the mouth [2,3].

Fibrosis of salivary glands leads to xerostomia, dysphagia, and subsequently periodontal infections. Radiographic changes in scleroderma patients are widening of the periodontal space, loss of lamina dura, resorption of the mandibular angle, (tail of the whale sign) zygomatic arch, digastric region, caput mandible, and coronoid process [3,4].

The American College of Rheumatology (former American Rheumatism Association) has defined criteria, that are 97% sensitive and 98% specific for SSc as follows:- Major criterion-Proximal diffuse (truncal) sclerosis (skin tightness, thickening, non pitting induration). Minor criteria - Sclerodactyly (only fingers and/or toes) Digital pitting scars or loss of substance of the digital finger pads (pulp loss) Bi-basilar pulmonary fibrosis.

Raynaud's phenomenon is observed in 90–98% of SSc patients.

The exact etiology of the osteolysis is unknown, but there are three proposed theories: Tightening of facial skin may exert excessive pressure on the mandible and induce bone loss, The vasculopathy associated with this disease may diminish the blood supply to the mandible resulting in bone ischemia and necrosis, Atrophy of the muscles of mastication may lead to bone necrosis [2].

Localized or limited cutaneous scleroderma (SSc) is limited to the distal extremities and face and also shows features of the CREST syndrome (Calcinosis, Raynaud's phenomenon, Oesophageal dysmotility, Sclerodactyly and Telangiectasia).

Majority of patients with scleroderma have GI tract involvement with oesophagus as first organ to be involved [5]. Hypotonia or atony with hypokinesia or aperistalsis in lower two-third of oesophagus, may lead to complications like stricture formation, aspiration, Barrett's oesophagus and adenocarcinoma. Small bowel involvement leads to malabsorption due to delayed intestinal transit time and bacterial overgrowth, dilatation, pneumatosis intestinalis, pseudo obstruction, sacculation changes.

Bibasilar pulmonary fibrosis - basal reticulo-nodular shadowing with progressive pulmonary volume loss are the main pulmonary manifestations. Scleroderma Renal Crisis is a severe life threatening renal disease found more commonly in diffuse form of Systemic Sclerosis than limited form. Cardiac involvement includes pericarditis, congestive heart failure and conduction problems.

CONCLUSION

Scleroderma is a multisystem organ involvement disorder with oral, cutaneous and gut manifestation. Leprosy, Achlasia cardia, Chagas disease, ILDs are the main differential diagnosis.

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