

Case Report

## Progressive Systemic Sclerosis: A Case Report and Review of Literature

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### ABSTRACT

Systemic sclerosis (SSc) is a collagen disorder, characterized by vasomotor disturbances, fibrosis, and atrophy of skin, subcutaneous tissue, muscles, and internal organs (alimentary tract, lungs, heart, kidney, CNS) with associated immunologic disturbances. Dense collagen is deposited in the tissues of the body in extraordinary amounts. Systemic sclerosis can affect the facial and oral structures as well and can present as diagnostic dilemma. This paper reports a case of systemic sclerosis in a 24 year old female patient with hard, shiny skin, depigmentations, mouse facies, resorption of terminal phalanges, claw like hands, microstomia, reduced mouth opening, and fibrosis of oral mucosa. Orthopantomogram showed generalized PDL space widening and resorption of condyle. The characteristic presentations in this case enabled us to establish the diagnosis of progressive systemic sclerosis.

**Key words:** systemic sclerosis, collagen disorder, microstomia.

### INTRODUCTION

Systemic sclerosis is a chronic inflammatory disease of unknown origin. Systemic sclerosis is also known as Scleroderma, dermatosclerosis and hidebound disease. The autoimmune nature of this disease is characterized by excessive deposition of collagen and glycosaminoglycans in the connective tissue of the dermis and internal organs. (1) Genetic, environmental and vascular factors have been attributed to the pathogenesis. One theory states that HLA histocompatibility complex, including HLA-B8, HLA-DR5, HLA-DR3, HLA-DR52 and HLA-DQB2 are involved in SSc pathogenesis. (2) Some data suggest that apoptosis and generation of free radicals also are involved in the pathogenesis of SS. (2)

The age group most affected is between third and fifth decade of life. (3) The

prevalence of scleroderma is estimated to be between 4 and 253 cases per million persons and preferentially affects females. (4)

The disease can occur in 2 forms: (a) Morphea (circumscribed scleroderma), characterized by local thickening of the skin; (b) generalized or progressive scleroderma (diffuse form), characterized by stiffness of the skin with distinctive involvement of the lungs, heart, kidneys, gastrointestinal tract and osteolytic changes in the skeleton. There are two variants of morphea a) atrophoderma of Psasini and b) eosinophilic fasciitis of Shulman. (5) A variant of this disease is known as the "CREST SYNDROME", an acronym for calcinosiscutis / Raynauds phenomenon/ Esophageal dysfunction/Sclerodactyly and Telangiectasia. (6)

Oral manifestations of progressive systemic sclerosis (PSS) include limited

mouth opening; xerostomia, microstomia, periodontal disease; increased periodontal ligament (PDL) width; and osseous resorption of the mandible. (7) The radiographic manifestation of PSS, in about two-thirds of patients, is an increase in the width of the PDL, resorption of condyle and resorption of terminal phalanges. (8)

Hypergammaglobulinemia and elevated erythrocyte sedimentation rate are also noted in SCC. (9) Pulmonary complaints can be evaluated with chest radiography and pulmonary function testing. Cardiac symptoms may prompt an electrocardiogram, echocardiogram, a stress test, or cardiac catheterization. Several serological tests are also useful for confirmation of the diagnosis. (10) Microscopic examination shows collagen deposition in tissues and vascular changes. (2) Inflammatory and obstructive changes are seen in arterioles and capillaries. (3)

### CASE REPORT

A 24 year old female patient reported to the Department of oral medicine and radiology, Annoor Dental College, Muvattupuzha with a chief complaint of difficulty in opening the mouth since eight months.

Extra oral examination revealed tense, smooth and shiny skin with areas of depigmentation and expressionless mask like face. Skin was firm and could not be picked up. Nasal ala was atrophied giving a pinched appearance of the nose, resulting in a “mouse facies” (FIG.1). Oro-facial manifestations showed microstomia, circumoral fibrosis a long with restricted mouth opening (30mm), producing a purse string appearance. Lip thinning and incompetency was seen (FIG.2). Fixation of temporo-mandibular joint and deviation was noticed towards the right side. (FIG.3) On intra oral examination, oral mucosa showed blanching and paleness. Tongue was depapillated, atrophic and rigid with restricted movements. (FIG.4)

Medical history revealed generalized body weakness, other affected organs such as heart, lungs and kidney. Healed ulcers on the finger tips were present.(FIG.5) The patient exhibited stiffness during movement of extremities. Resorption of terminal phalanges was also observed, giving a shortened “claw like” appearance to the hands. (FIG.6) Terminal phalanges were stiff, and deformed. The skin over the hands was also firm and could not be picked up. She also reported dry cough associated with chest pain, heartburn and dysphagia.



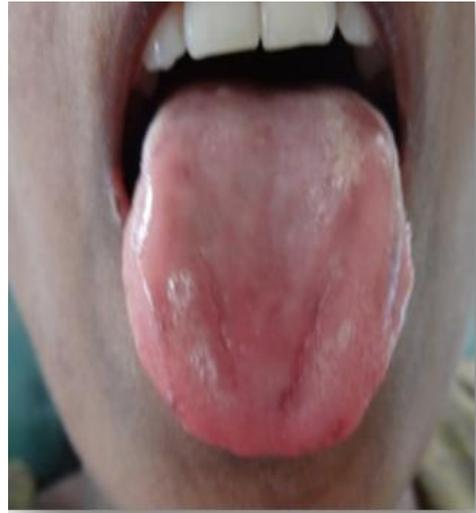
FIG. 1: pinched appearance of nose



FIG. 2: Smooth, tense, shiny skin with mask like facial appearance and thin lips



**FIG. 3:** restricted mouth opening and deviation towards right side



**FIG.4:** Atrophic and depapillated tongue



**FIG. 5:** Healed ulcers on finger tips



**FIG. 6:** claw like deformity and resorption of terminal phalanges



**FIG. 7:** OPG shows generalized widening of periodontal ligament and resorption of condyle



**FIG. 8:** Hand-wrist radiograph shows mild resorption of phalanges of middle and index finger

Orthopantomogram (OPG) showed generalized PDL space widening more pronounced in mandibular posterior teeth, interdental bone loss in maxillary and mandibular anterior teeth along with resorption of condyle.(FIG.7) Hand-wrist radiograph showed resorption of phalanges of index finger and middle finger.(FIG.8) The patient was subjected to ANA and Anti Scl 70 testing and showed positivity. This case was diagnosed as progressive systemic sclerosis.

## DISCUSSION

Scleroderma literally means “hard skin” and was first reported by William and Robert Watson in 1754. Goetz proposed the name progressive systemic sclerosis in 1945, when the systemic nature of the disease was proven. <sup>(11)</sup>

It affects 19 persons per million populations every year. One of the first sign of the disease is Raynaud’s phenomenon, a vasoconstructive event triggered by emotional distress or exposure to cold. In some patients, the first manifestation is active, often migratory, polyarthritis. In others, severe erosive digital osteoarthritis (related to CREST syndrome, particularly in females) occurs first. <sup>(12)</sup>

In progressive systemic sclerosis the disease extends to involve the upper extremities, trunk, face, and, finally, the lower extremities, which may sometimes be spared. In the early stages, the painless, slightly pitting edema often lasts several months before tightening of the skin occurs. Resorption of terminal phalanges (acro-osteolysis) and flexion contractures produce shortened, claw-like fingers. Vascular events and abnormal collagen deposition associated with ischaemic lesions, infarctions, necrosis produce ulcers on finger. <sup>(6)</sup> The subcutaneous collagen deposition of the facial skin result in loss of skin folds around the mouth giving a Mona Lisa or mask like appearance. Atrophy of nasal alae produces a mouse facies or bird face. <sup>(2)</sup>

Generalised hyperpigmentation, similar to that of Addison’s disease but without adrenal insufficiency can occur and may antedate the sclerosis. <sup>(13)</sup> Brown pigmentation of skin occurs as a late manifestation of the disease. <sup>(2)</sup>

A mild variant called localized scleroderma (linear scleroderma) affects a solitary patch of skin. Linear bands with a furrow and elevated ridge on one side are present on the face/forehead, chest and trunk which resemble the mark produced by the strike of sword termed as coupedesabre (strike of the sword). <sup>(6)</sup>

Systemic Sclerosis occurs in conjunction with other auto immune conditions such as rheumatoid arthritis, lupus erythematosus, dermatomyositis, and Sjogren's syndrome. Rheumatoid factor and antinuclearanti bodies are demonstrable in patients with scleroderma. <sup>(3)</sup>

In progressive disease, fibrosis of lungs, heart, kidneys, gastrointestinal tract leads to organ failure. Pulmonary fibrosis leads to pulmonary hypertension and heart failure, a primary cause of death in these patients. Renal crisis is characterized by increased blood pressure and rapidly progressing to renal failure if untreated, usually within the first 5 years of the disease. <sup>(6)</sup>

Oral manifestations include microstomia as a result of collagen deposition in perioral tissues leading to limited mouth opening. Characteristic furrows radiating from the mouth produce a “purse string” appearance. Loss of attached gingiva and gingival recession occur in some patients. Involvement of oral submucosa may cause the tongue to become narrow and stiff, giving rise to a “chicken tongue”, thus hindering swallowing. <sup>(14)</sup> Dysphagia develops later due to deposition of collagen in the lingual and esophageal submucosa. <sup>(6)</sup> These patients are subjected to increased risk of oral cancer, particularly squamous cell carcinoma of tongue. <sup>(15)</sup>

On dental radiographs diffuse widening of periodontal ligament space is

present. Varying degrees of resorption of posterior ramus of the mandible, coronoid process, chin, and condyle may be detected on panoramic radiographs in 10-20% of patients.

During the early phases it may be difficult to make a diagnosis of systemic sclerosis. The clinical sign of stiffened skin along with Raynaud's phenomenon are suggestive of diagnosis. Detection of anticentromere antibodies or anti-Sci70 (topoisomerase I) also adds on the diagnosis. Anti-Sci70 antibodies are often associated with systemic sclerosis; anticentromere antibodies with more limited forms of scleroderma or CREST syndrome. (6)

Histopathology reveals gingival thickening and hyalinization of the collagen fibers, epithelial atrophy and sclerosis of the walls of the blood vessels. The microscopic changes in the periodontal ligament demonstrate hyalinization with diminution in the number of connective tissue cells. (2)

Systemic medications, such as D-penicillamine, are administered to inhibit collagen production. Dental procedures in these patients may encounter difficulties due to limited mouth opening and rigidity of the tongue. The oral opening may be increased by an average of 5mm by stretching exercises or by a bilateral commissurotomy. Patients with extensive resorption of the angle of the mandible are at risk for developing pathologic fractures from minor trauma, including dental extractions.

When treating a patient with diffuse scleroderma, the extent of the heart, lung, or kidney involvement should be considered, and appropriate alterations should be made before, during, and after treatment. Patients with Sjogren's syndrome should have daily fluoride treatments and make frequent visits to the oral hygienist. (16)

Other management strategies are directed at controlling symptoms. Esophageal dilation is used to correct the

esophageal dysfunction and dysphagia. Calcium channel blockers help to increase peripheral blood flow and lessen the symptoms of Raynaud's phenomenon. Angiotensin-converting enzyme (ACE) inhibitors often effectively control hypertension if kidney involvement is prominent. (6) The prognosis is poor for those with diffuse involvement than for those with limited involvement.

## CONCLUSION

Systemic sclerosis is a complex disease with multisystem involvement. Detailed patient investigation is required, because of the possibility for generalized involvement of other organs. A proper clinical and laboratory analysis is important for the determination of the genetic risk and prognosis. Early diagnosis and individually tailored therapy help to manage this disorder which is treatable but not curable.

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