

Case Report

A rare case of hidebound disease with dental implications

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ABSTRACT

Systemic sclerosis (also called as Scleroderma or hidebound disease) is a chronic sclerotic disease of unknown etiology which causes diffuse, increased deposition of extra cellular matrix in connective tissue with vascular abnormalities, resulting in tissue hypoxia. The disease is characterized by diffuse fibrosis; degenerative changes; and vascular abnormalities in the skin (scleroderma), articular structures, and internal organs. Aesthetic and facial dysfunctions are followed by important oral and facial manifestations. Most oral manifestations begin with tongue rigidity and facial skin changes. Bone resorption of mandibular angle and widening of periodontal ligament space on periapical radiographs are important radiological findings. Other systemic changes include the involvement of internal organs, which lead to serious complications as well as disorders in the cardiac muscle and Raynaud's phenomenon. This is a case report of 30-year-old female patient with the classical features of this disease. This case is reported for its rarity and variable expressivity. The main aim of this article is to describe thorough presentation of the case report, various forms of scleroderma, pathogenesis, oral, extraoral, periodontal manifestations of scleroderma, and its treatment options. A brief review of the literature, focusing on dental alterations is also presented.

Key Words: Acroosteolysis, connective tissue, crest syndrome, hidebound disease, periodontitis, Raynaud's phenomenon, scleroderma

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INTRODUCTION

Scleroderma is a connective tissue disorder characterized by tissue fibrosis, obliterate microangiopathy, and immune abnormalities. The term scleroderma is derived from the Greek word *scleros* (hard) and *derma* (skin), hence the meaning hard skin. This term describes the pathognomonic clinical appearance of the skin seen in this disease. It is also called "Hidebound disease" since hidebound skin is the hallmark of the disease.^[1]

Etiology is unknown, but several risk factors such as age, sex, genetic background and environmental

exposure are thought to influence susceptibility. Vascular injury and autoimmune factors are thought to play major role in the etiology of the disease.^[2] Defects in cell mediated immunity increase collagen production which results in fibrosis.^[3] In the early phase of this condition, there is skin infiltration by T lymphocytes and abnormal fibroblast activation, which leads to increased production of extracellular matrix in the dermis, primarily type I collagen.^[4] This results in symmetrical thickening, tightening, and induration of the skin. There is also arterial and arteriolar narrowing due to intimal proliferation and vessel wall inflammation.^[5]

Systemic sclerosis is a fatal, multi-system disease with an uncertain prognosis. The involvement of the skin together with the quality of its mobility, particularly in the distal portions of the extremities is by far the most obvious symptom.^[6] Cutaneous manifestations include thickening of skin, starting with pitting edema that, over a period of time, is replaced by tightening

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and hardening of skin.^[7] Raynaud's phenomenon, a paroxysmal vasospasm of fingers, which results in change in color of fingertips as a response to cold or emotion and resorption of terminal phalanges is usually the first symptom. Oral and facial tissues are often affected, presenting characteristic features. Most clinical manifestations begin with tongue rigidity and classic facial skin hardening, which gives it a classic mask-like appearance. Limited opening of the oral orifice besides bone resorption at the angle of the mandible are commonly reported feature.^[7,8] Abnormal movement of mandible and subluxation are other possible dental manifestations. Progressive vascular fibrosis and deficient wound healing make any surgical procedure difficult and hazardous in these patients.

DIAGNOSIS

The diagnosis of systemic sclerosis is based on the identification of features that distinguish it from other disease, and thus a detailed history and careful physical examination are required. The American College of Rheumatology has proposed criteria to assist in identifying those affected with the condition.^[9]

Major criteria

Proximal scleroderma

Symmetric thickening, tightening and induration of the skin of the fingers and the skin proximal to the metacarpophalangeal or the metatarsophalangeal joints.

Minor criteria

Sclerodactyly

The above mentioned skin changes are limited to the fingers.

Digital pitting scars

Loss of substance from the finger pad as a result of ischemia.

Basilar pulmonary fibrosis

Bilateral reticular pattern of linear or lineonodular densities most pronounced in basilar portions of the lungs or standard chest roentgenogram, not attributable to primary lung disease.

To fulfil a diagnosis of systemic sclerosis, either one major or two minor criteria must be present.

Clinical evidence of active disease in systemic sclerosis must be investigated. Pulmonary complaints should prompt an evaluation with chest radiography, pulmonary function testing, or high resolution computed tomography of the chest. Cardiac symptoms

may prompt an electrocardiogram, echocardiogram, a stress test, or cardiac catheterization. Serial echocardiograms are routinely done in systemic sclerosis to screen for pulmonary hypertension, and if positive, right heart catheterization is performed. Symptoms of gastrointestinal involvement may require evaluation with endoscopy. Capillary microscopy may be indicated for complaints of Raynaud's phenomenon.

This article reports a case with oral and facial manifestations in systemic sclerosis. A brief review of the literature is presented, focusing on dental manifestations.

CASE REPORT

A 30-year-old female patient reported to the Department of Dentistry in our hospital with a complaint of difficulty in chewing due to decreased mouth opening since past few months. Extraoral examination of the patient revealed multiple ill defined white patches on the forehead and cheeks [Figure 1]. Facial skin was smooth, taut, and mask like. It was firm and could not be picked up. Temporomandibular joint was stiff on palpation. Paraoral examination revealed circumoral fibrosis resulting in microstomia with characteristic furrows radiating from the mouth giving rise to 'purse string appearance'. The lips had become thin and nasal alae were atrophied giving a pinched appearance to the nose, classically known as "mouse facies" [Figure 2].

Intraoral examination was difficult due to microstomia and limited mouth opening (28 mm). On palpation, tongue was rigid with decreased mobility. Periodontal examination revealed generalized mild chronic periodontal disease, with an average probing depth of 3-5 mm. Periapical intraoral radiographs showed normal lamina dura with widening of periodontal space, which was most prominent on the posterior teeth [Figure 3].

Stiffening of phalangeal joints in both fingers and toes was observed. Flexure contractures producing shortened 'claw like' fingers were also present [Figure 4]. Roentgenographic examination of hands showed resorption of terminal phalanges (acro-osteolysis) [Figure 5].

The patient was treated symptomatically, with scaling and root planning and was advised oral hygiene instructions. The periodontal pockets responded well to scaling and root planning. Other recommended aids were to eat a balanced diet and using electrical tooth brush and anti-bacterial mouth washes.



Figure 1: White patches on forehead and cheeks



Figure 2: Lips had become thin giving mouse facies appearance



Figure 3: Radiograph showing widening of periodontal ligament



Figure 4: Claw like hands with symmetric thickening and tightening of skin



Figure 5: Radiograph showing resorption of terminal phalanges

DISCUSSION

Systemic sclerosis is a chronic connective tissue disease that causes widespread microvascular damage and excessive deposition of collagen in the skin

and internal organs. The first convincing description of the disease appeared in monography written by Carlo Curzio, in Naples in 1752.^[10] It was given the first name Sclerodermie by Gintrac in 1847. Goetz proposed the name progressive systemic sclerosis in 1945, when the systemic nature of the disease was proven.^[11]

The etiology is unknown, however, vascular injury and subsequent over production of collagen has been proposed as a possibility. The pathological findings indicate that fibroblasts are activated to produce excess amounts of collagen and other components of cellular matrix. Moreover, an autoimmune mechanism is involved because these patients show high levels of specific and nonspecific circulating auto antibodies i.e., against deoxy ribonucleic acid topoisomerase, ribonucleic acid polymerase II, centromeric protein B, laminin S, and Vimentin.^[12]

Clinically, women between the ages of 30 and 50 are predominantly affected.^[13] Raynaud's phenomenon (an episodic change in color of the fingertips in response to cold or emotions) is the most common early manifestation. This is followed by early skin changes starting with pitting edema. In several months, the edema is replaced by a tightening and hardening of the skin, which results in difficulty in movement of the affected parts.^[14] Cutaneous involvement usually starts peripherally on the hands and face, and gradually extends centripetally.^[2] A hallmark of the disease is involvement of hands causing such changes as atrophy or of ischemic damage to the tips of the fingers and contractures preventing straightening of the fingers (claw like fingers). Hyperpigmentation, telangiectases, and subcutaneous calcifications may occur leading to deformity and severe cosmetic problems.^[15]

The disease has been divided into two major categories: Diffuse and localized.^[16] The diffuse form has generalized skin involvement and rapid progressive internal organ involvement, whereas the localized form shows limited cutaneous involvement generally confined to the distal aspects of the fingers and the face. A variant of this disease known as the "CREST syndrome" is an acronym for calcinosis cutis, Raynauds phenomenon, esophageal dysmotility with dysphagia, sclerodactyly, and telangiectases.^[17]

Localized scleroderma is characterized by localized fibrosis resembling that of scleroderma, but without the systemic involvement of serological changes. It occurs in two forms circumscribed form and linear form. In linear localized form, the face is frequently the chief site and the area of fibrosis resembles the scar from a sabre cut (*en coup de sabre*) and may result in hemiatrophy of the face. The circumscribed form (morphea) begins with violaceous patches on the skin. Later in the course of the disease the lesion "burns out" and appears as hypo or hyperpigmented area depressed below the level of skin. Scleroderma localized to the hands is called acrosclerosis.^[18]

In multi-organ involvement gut is affected commonly. Smooth muscle atrophy and fibrous replacement of the muscularis may develop in the esophagus leading to reflux with erosive esophagitis and dysphagia.^[19] In lung, fibrosing alveolitis mainly affects patients with

diffuse form. Renal involvement in scleroderma is due to diminished blood flow in the afferent arterioles and is called scleroderma renal crisis.^[20] Death is from renal disease, heart failure, or severe intestinal malabsorption.

Oro-facial manifestations include narrowing of eyes and loss of skin folds around the mouth giving a characteristic mask-like appearance of face (Mona Lisa face). The lips may be constricted (Fish mouth) or become pursed with radiating furrows. Involvement of periarticular tissues of temporomandibular joint together with microstomia may greatly limit opening of mouth, causing a pseudoankylosis.^[21] Involvement of oral submucosa may cause the tongue to become stiff and narrowed (chicken tongue). The mandibular angle may be resorbed or rarely there is gross extensive resorption of the jaw.

In dental radiographic findings, the main changes are seen in periodontium characterized as widening of periodontal ligament space (PDL space) at the expense of alveolar bone especially around the posterior teeth.^[22] The exact mechanism for widening still remains unclear. This is because when muscles of mastication are involved they become more bulky due to fibrosis, there is an increase in masticatory occlusal force especially on posterior teeth.^[23] Another plausible reason is that increased collagen synthesis in PDL leads to increase in bulk of the ligament, which is accommodated at the expense of the alveolar bone.^[24,25] Additional radiographic changes reported include resorption of the condyle, coronoid process and the angle of the mandible.^[26] These osseous changes are apparently related to pressure atrophy or ischemia. Resorption in the apical part of the roots and complete destruction of the lamina dura was reported in a case report of scleroderma.^[27] Nagy, *et al.*, found that some patients had salivary hypofunction, keratoconjunctivitis sicca, or both.^[28]

No agent has demonstrated efficacy in arresting/improving skin changes. Self-management to maintain core body temperature and avoid peripheral cold exposure is important. Infections of the ulcerated skin should be treated with prompt antibiotic therapy.

Maintenance of existing dentition is important because microstomia and tongue rigidity can make the prosthetic replacement difficult. The preventive nature of proper oral hygiene habits and the need

for good permanent restorative dentistry should be stressed to these patients because the progressive constriction of the oral cavity eventually limits access to perform adequate dental treatment. Patients with extensive resorption of the mandible are at risk for pathologic fractures from dental extractions. Oral exercise techniques like the use of an increasing number of tongue blades between the posterior teeth to stretch the facial tissues may help to increase oral opening. Surgical procedures like bilateral commissurotomy to enlarge the oral orifice have been described.^[29] Implants have lately been used successfully for extensive prosthetic reconstruction in such patients.^[30]

DIFFERENTIAL DIAGNOSIS OF SYSTEMIC SCLERODERMA

Based on the vascular changes

1. Primary Raynaud's phenomenon
2. Physical trauma
3. Chemical exposure (polyvinyl chloride disease)
4. Drugs/toxins (toxin oil syndrome, drug induced pseudoscleroderma)
5. Other autoimmune connective tissue disorders (Systemic Lupus Erythematosus (SLE), Polymyositis/Dermatomyositis (PM/DM), Vasculitis, Rheumatoid Arthritis (RA), Sjogren's syndrome (SS), Cryoglobulinemia)

Based on the skin changes

1. Localized scleroderma (Morphea, linear scleroderma with atrophy of the affected extremity)
2. Scleroderma like skin changes
3. Undifferentiated connective tissue disease (UCTD)
4. Eosinophilic fasciitis with eosinophilia
5. Eosinophilia myalgia like syndrome
6. Overlap syndromes (scleroderma with SLE, or SS or DM/PM, or RA)
7. Metabolic – genetic disorders
8. Scleroedema generalized Buschke
9. Scleromyxoedema
10. Insulin dependent diabetes mellitus (scleredema, digital sclerosis)
11. Amyloidosis

Based on visceral involvement

1. Ageing and diabetes mellitus
2. Idiopathic pulmonary fibrosis
3. Idiopathic pulmonary hypertension
4. Sarcoidosis

5. Amyloidosis
6. Infiltrative cardiomyopathies
7. Malignant hypertension
8. Other autoimmune connective tissue disease

CONCLUSION

In systemic sclerosis, all oral tissues are affected, causing limited oral function such as decrease in the maximal oral aperture, impaired healing, widened periodontal ligament spaces and neurological symptoms, which may be presented to varying degrees. Dentists should monitor these patients periodically, performing clinical and radiological examination so as to follow the course of the disease and prevent poor oral hygiene, loss of teeth, and periodontitis.

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