

Case Report

A multisystem disease scleroderma – A case report

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ABSTRACT

Systemic sclerosis (SSC) is an autoimmune, connective tissue disorder with involvement of various systems of the body. There are also various oral and para-oral manifestations too. It is described under localized and diffused forms as mentioned in various literatures. Patients with SSC have prevalent clinical manifestations involving all the organ systems of the body with various oral and para-oral manifestations, which are of foremost concern for the dentists to maintain hygiene of the oral cavity and to elevate the quality of life. This paper aims to present a case report of a 36-year-old female patient with scleroderma who presented with signs of skin stiffness, fishlike facies, purse string appearance, etc., with review of literature.

Keywords: Antinuclear antibody, Collagen accumulation, Gingival, Periodontal diseases, Skin fibrosis, Systemic sclerosis

INTRODUCTION

The term scleroderma derived from the Greek words for hard and skin, describes a group of clinical disorders characterized by thickening and fibrosis of the skin. Hippocrates was the first one to describe scleroderma as a thickening of skin. It was first described by Carlo Curzio in 1752 and the systemic nature of the disease by Robert Goetz.^[1] It is defined as a multi-systemic, autoimmune disease in which there is increased fibroblast activity, which results in vascular damage, collagen accumulation and scarring of skin and fibrosis. Its etiology is still unknown but there is genetic predisposition with various factors playing roles such as environmental, viruses (cytomegalovirus), chemicals (vinyl chloride, pesticides, and silica), and drugs (cocaine, appetite suppressants, and Vitamin K). It is acquired sporadic disease which affects all races, with an incidence of 9–19 cases/million/year, having a female predominance of 4–5:1. The peak age is 35–64 years.^[2–4]

Scleroderma can be classified into two different forms depending on the extent of skin involvement

1. Diffuse cutaneous form and
2. Limited cutaneous form.

There are several oral manifestations which include microstomia, xerostomia, telangiectasia, increased decayed, and filled teeth. There is difficulty in eating swallowing, and making speeches. As a result of this, there is gingivitis

and periodontitis. Since systemic sclerosis (SSC) is a very rare, systemic disease having several oral and periodontal implications which have not been described in the literature very well.^[4,5] This article is aimed to explain various systemic and oral manifestations of scleroderma with its diagnosis and management according to the signs and symptoms.

CASE REPORT

Patient aged 36 years visited the hospital with the chief complaint of yellowish deposits in the teeth for 2 months. There is no significant past dental and family history. Patient's medical history revealed that she is suffering from hypothyroidism and is on Thyronorm 50 mg OD for 5 years. Patient was diagnosed with scleroderma at a Private Medical College, Bangalore and was on medication Tab Depin 10 mg and Omnacortil 5 mg OD for 3 months. There were no deleterious habits and the patient was moderately built and moderately nourished with normal gait and erect posture.

On general examination, the skin over the face, neck, and extremities appeared to be stretched and shiny with hyperpigmented hidebound skin [Figure 1]. She was having difficulty in eating, speaking, and swallowing. There was a claw like deformity of hands with puffed fingers. There were pitted scars with hyperpigmentation in both the digits [Figure 2]. All the vital signs and vital parameters were normal.

On extra-oral examination, it was found that there is loss of facial expression lines resulting in a mask like appearance or

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a mouse-like facies. There is a fish mouth like appearance due to the thinning of lips. There is a beak shaped nose. There was circumscribed morphea seen around both the eyes [Figure 1]. There was approximately 2 cm of mouth opening with a purse string appearance [Figure 3].

Intraoral examination reveals presence of calculus present in lower anteriors and molars up to cervical 1/3rd of the teeth with inflamed marginal gingiva in molars. There was bleeding on probing present. There was stiffness and paleness of the oral mucosa. Diagnosis of chronic generalized gingivitis was given along with the oral manifestations of scleroderma was given. Other conditions such as oral submucous fibrosis (OSMF) and anemic stomatitis can be considered as differential diagnosis. Blood investigations

reveal hemoglobin levels of 14 mm of Hg with total white blood cell count of 7700 cells/cmm. Differential count reveals 49% of polymorphs and 58% of lymphocytes with normal erythrocyte sedimentation rate. Bleeding time of 2 min 10 s and clotting time of 6 min 15 s were noted. Rheumatoid arthritis factor appears to be negative. Antinuclear antibody reveals a positive correlation. Orthopantomogram does not give any relevant information whereas hand wrist radiograph reveals erosion of terminal phalanges [Figure 4a and b]. OSMF can be ruled out as diagnosis as there was no history of deleterious habits, anemic stomatitis can be ruled out after blood investigations. All the investigations were directed towards diagnosis of scleroderma. Patient was advised to maintain better oral hygiene so as to avoid further



Figure 1: The skin over the face, neck appeared to be stretched and shiny with hyperpigmented hidebound skin.



Figure 3: A mask such as appearance or a mouse-like facies with thinning of lips. There is beak shaped nose. There was circumscribed morphea seen around both the eyes.



Figure 2: Claw like deformity of hands with puffed fingers.



Figure 4: (a) Orthopantomogram with does not give any relevant findings. (b) Hand-wrist radiograph reveals erosion of terminal phalanges.

oral manifestations like periodontitis, hence was referred to department of periodontics for scaling. Patient was also advised to perform regular mouth exercises for maintaining adequate mouth opening, healthy lifestyle, and eating habits. This article aims to discuss and highlight the oral manifestations of scleroderma with investigations and treatment planning.

DISCUSSION

As discussed in introduction, scleroderma is described as an auto-immune disease having clinical disorders characterized by thickening and fibrosis of the skin and oral mucosa.^[1,2]

Epidemiology/risk factors

1. The incidence of the disease varies regionally and is approximately 0.5–2/100,000 individuals. The prevalence of the disease is approximately 17.6 cases/100,000 individuals. It affects women beyond the 4–5th decade.^[4]
2. In India, the prevalence of the disease was 25.9/100,000 people till 2016 with incidence rate of 15.1/100,000 individuals. The possible risk factors quoted were female sex, African origin, and exposure to various metals like silica, vinyl chloride, solvents like trichloroethylene, and drugs like bleomycin, etc. About 66.7% of patients had joint involvement whereas 52.6% had internal organ involvement.^[5,6]
3. Up-to 80% of cases manifests with oral manifestations of SSC but it usually gets ignored due to several other systemic symptoms. Yet, the oral and dental manifestations

of scleroderma, which can occur in up to 80% of patients, generate a significant quality-of-life burden.^[4,6]

Pathogenesis

The pathogenesis of the disease is as follows [Table 1].^[7,8]

Clinical features

1. Skin manifestations – As name itself suggests that there is mandate skin involvement in each patient having different degree of severity. Hence, the symptoms are classified based on its involvement as two subtypes as limited and diffused cutaneous [Table 2].^[4,5,9]

Table 2: Subtypes of systemic sclerosis

Limited cutaneous systemic sclerosis	Diffuse cutaneous systemic sclerosis
1. Characterized by skin involvement seen in proximities not involving trunk.	1. Characterized by involvement of proximities with trunk involvement
2. Facial involvement	2. Facial involvement

Skin involvement occurs in two phases discussed below:

- a. Initial puffy finger phase – there is inflammation of hands with non-pitting edema which lasts longer. As there is inflammation, so there will erythema, pain, and pruritus. From all these manifestations, there will be compression of certain nerves leading to compression neuropathies, for example, carpal tunnel syndrome.
 - b. Second prolonged fibrotic phase – skin thickening and resultant fibrosis that develops during the initial phase is later followed by the fibrotic phase. It begins distal to the metacarpophalangeal joints, a condition termed sclerodactyly which later progresses proximally. Fibrosis of subcutaneous structures causes contractures and decreased mobility of the peripheral joints. Telangiectasias seen on the peripheries, chest, and face. Digital ulcers are very common which is seen on the tips of finger or over calcinosis nodules.^[9,10]
2. Nail manifestations – there are abnormal capillary and microvascular loops which are seen in nails by ophthalmoscope.^[11]
 3. Musculoskeletal manifestations – Due to fibrosis, there are various flexion deformities of fingers and various body joints. Polyarthralgia's may or may not be present. Flexion contractures can develop in the fingers, wrists, and elbows.^[12,13]
 4. Gastrointestinal (GI) manifestations – First GI symptom occurs as dysphagia which is present in almost all patients. As a result of this, there is acid reflux which later causes heartburn and stricture. Barrett esophagus occurs most commonly in patients with SSC which can predispose it to adenocarcinoma.^[14,15]

Table 1: Pathogenesis of scleroderma

Vasculopathy	Autoimmunity	Fibrosis
1. Arterioles and the microvascular system are affected	1. innate and adaptive immune system gets activated	1. Myofibroblasts secrete high levels of ECM components
2. Endothelial cells swells up causing cell apoptosis	2. B cells secrete autoantibodies which is directed against the nuclear and several other antigens	which results in excessive stiffening and rigidity of the involved tissue
3. Expression of adhesion proteins and cytokines gets altered causing chronic hypoxia	3. Activation of inactivated fibroblasts is caused by fibrogenic cytokines	2. The increased stiffness is perceived by integrins expressed at the surface of myofibroblasts
4. Hypoxia and reactive oxygen species together cause continuous damage	4. Leads to the trans-differentiation of preadipocytes, endothelial cells and mesenchymal stem cells into myofibroblasts	3. Smooth muscle cell proliferation, Intima thickening
5. All the factors together causes complete loss of capillaries		4. Duplications of the basement membrane
		5. Endothelial mesenchymal transition

ECM: Extracellular matrix

5. Cardiopulmonary manifestations – There is increased buildup of collagen fibers in the smaller arteries, resulting in vasculopathy, which predisposes to death ultimately. In the lungs, there was fibrosis, which causes impairment of gaseous exchange and interstitial lung diseases.^[15-17]
6. Renal manifestations – Renal manifestations are sudden causing scleroderma renal crisis in cases of diffuse cutaneous cases not all. It is often indicated by severe hypertension and thrombotic hemolytic anemia.^[17,18]
7. Oral and para-oral manifestations – There are several oral and para-oral manifestations present in patients with SSC.^[3-7,19-21]
 - Jaws – maxilla and mandible shows varying degrees of bone resorption. There are some explanations which are given for the same are: (1) Tightening of the facial skin exerts excessive pressure on the mandible which, in turn, brings the bone loss; (2) there are small arteries vasculopathy which reduces the blood supply to the mandible causing ischemia and necrosis of the bone; and (3) muscles of mastication also gets affected, there atrophy causes ischemia and necrosis.
 - Mouth opening – fibrosis of buccal mucosa causes limited mouth opening. This, in turn, causes difficulty in eating food, day to day speech too.
 - Gingiva – Maintenance of oral hygiene is difficult in these patients due to limited mouth opening. Hence, there are gingival and periodontal diseases. Vasculopathy causes absence of bleeding on probing during clinical examination despite having gingivitis and periodontitis.
 - Tooth – Inability in opening mouth causes deteriorated oral hygiene giving rise to increased incidence of decayed, missing, and filled teeth.
 - Tongue – Increased fibrosis causes microstomia with loss of papillae. The tongue also becomes rigid making difficulty in speech and swallowing.
 - Temporomandibular joint (TMJ) – The soft tissues related to TMJ is affected, which results in pseudo-ankylosis.
 - Others – xerostomia and telangiectasia.

Evaluation/investigations

SSC is usually diagnosed after obtaining detailed case history with thorough clinical examination and assessment. Early

Score	Involvement
0	Normal skin thickness
1	Mild skin thickness
2	Moderate skin thickness
3	Severe skin thickness

diagnosis of the disease always aids in early evaluation and extent of involvement of various organ system.

Firstly, and fore mostly, skin is assessed using the modified rodman skin score which includes scoring from 17 specific body areas including: Face, chest, abdomen, arms, forearms, hands, fingers, thighs, legs, and feet [Table 3].^[9,10]

These are supervised time to time to assess the progress of disease. A complete physical examination of these patients is done to detect involvement of multiple organ systems and to detect subtypes.

Autoantibody tests

Several autoantibodies are an essential diagnostic tool in evaluating the progression of the disease which is detected through direct immunofluorescence. These tests are usually positive in almost 90% of patients having SSC. Anti-centromere antibody and anti-RNA polymerase III antibody also found to be positive in patients having diffuse cutaneous SSC.^[22,23]

Laboratory evaluation

Complete blood counts, and renal and liver functions tests are performed to assess involvement of various organs.

Radiographic evaluation

Calcinosis can be seen on the X-rays of the extremities. There is loss of distal phalanges. To assess the muscle involvement, electromyography/nerve conduction velocity test are performed to confirm. Lung diseases like interstitial diseases can be detected by pulmonary function tests. Arrhythmias can be diagnosed by electrocardiography. To detect esophageal and upper GI involvement, endoscopy and esophageal manometry are advised.^[17-19,23]

Treatment

It starts with the education and motivation of the patient before start investigations and treatment planning. They are promoted and encouraged to engage themselves in regular exercises, maintaining a healthy lifestyle and eating habits. Nor there is specific treatment for SSC, neither drug which interrupts the progression of the disease. The treatment should be comprehensive and holistic to improve the quality of life for patients to prevent further complications.

Specific therapies in scleroderma

Below are the specific therapies for various disease manifestation.^[14-18,24-27]

Raynaud phenomenon

This treatment aids in preventing digital ischemia and ulcers, which are easier to prevent. Conservative management for the Raynaud phenomenon is followed in patients, in which it is advised to keep the extremities and body warm. Vasodilator drugs such as calcium channel blockers are first-line drugs to be used, namely, nifedipine or amlodipine. In

addition, other vasodilators such as pentoxifylline, nitroglycerine, and sildenafil can be used.

Skin disease

Various immunosuppressive agents are used such as methotrexate, mycophenolate mofetil, and cyclophosphamide. Topical applications with corticosteroids, Vitamin D analogs, and tacrolimus are usually providing relieve from skin discomfort.

Musculoskeletal involvement

Mild-to-moderate arthralgias are mostly present in the patients and for which low-dose of glucocorticoids are prescribed. Physical and occupational therapy are prescribed for avoiding contractures.

Pulmonary involvement

It is leading cause of mortality which necessitates various health professionals such as rheumatologists, pulmonologists, and cardiologists to intervene together. Mycophenolate mofetil is commonly used drug to treat interstitial lung diseases associated with SSC.

Cardiac involvement

Arrhythmias are managed with antiarrhythmic agents and if needed pacemaker placement.

GI involvement

To avoid heartburn and gastroesophageal reflux, lifestyle and dietary adjustments are taken up by the patients to elevate quality of life. For the GI involvement, drugs such as proton-pump inhibitors and H2 blockers are widely used.

Scleroderma renal crisis

ACE inhibitors usually captopril is widely used to control the occurrence of renal crisis.

CONCLUSION

The disease SSC is highly associated with increased mortality mostly in cases of having collagen vascular diseases. Over the past few years, the prognosis of this disease has been increased to 5-years survival rates which is up to 80%. Understanding the etiology, clinical manifestations and their definitive treatment helps improve the prognosis of the disease. Furthermore, it is guided to the patients to visit the dentist at regular interval of time to maintain good oral health.

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