Review Article

Review of orofacial considerations of systemic sclerosis or scleroderma with report of analysis of 3 cases

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Abstract

Scleroderma (skleros; hard, and derma; skin), is currently known as systemic sclerosis due to its progressive nature and widespread tissue involvement. It is a rare connective tissue disorder with a wide range of oral manifestations. Thickening of the skin is the hallmark of the disease. The patient education for self-care and multidisciplinary approach would be needed to manage the condition. The article presents the review of orofacial considerations in scleroderma with a report of analysis of orofacial manifestations 3 cases.

Key words: Connective tissue disorders, reduced mouth opening, scleroderma, widened periodontal ligament space

INTRODUCTION

Progressive systemic sclerosis (PSS), conventionally called as scleroderma is a collagen disorder characterized by fibrosis that involves skin, mucosa, muscles, and internal organs such as the gastrointestinal tract, lungs, blood vessels, and kidneys. This rare disorder involves oral and paraoral structures that form the distinct domain of clinical manifestation of this disorder.[1-5] These mainly include mask-like facial appearance, microstomia, periodontal diseases, widened periodontal ligament (PDL) space, xerostomia, and osseous resorption.[3-8] The present article reviews the orofacial considerations in scleroderma.

CASE DESCRIPTIONS

3 patients, 26-year-old female (Case 1), 28-year-male (Case 2), and 46-year-old female (Case 3) reported to the dental hospital with a chief complaint of reduced mouth opening or narrowing of the mouth. The third case was the diagnosed case of scleroderma since 4 years with orofacial involvement.

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On an average, all the patients mentioned the discomfort in food intake and brushing of the teeth, tightened feeling of the extremities, and the weakness. The summary of the analysis of clinical features in 3 cases is presented in Table 1. Case 1 and Case 2 had reduced mouth opening to 2 cm and 4 cm, respectively, with blanching; they were treated as the cases of submucous fibrosis. For the third case, although there was feeling of tightening, there was no obvious reduction of the mouth opening. For all the 3 patients, the physiotherapy in the form of mouth stretching exercises and tongue blade therapy was advised to improve their mouth opening. Case 1 also had a history of frequent oral ulcerations. She was given supplements as Vitamin B complex, folic acid, and lycopene. The second case also had generalized periodontal involvement with pockets, and mobility of teeth, scaling, and oral hygiene maintenance therapy were instituted for the patient.

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Table 1: Summary of clinical features seen in 3 cases reported

Clinical features	Case 1 [Figure 1]	Case 2 [Figure 2]	Case 3 [Figure 3]
Taut, shiny facial skin	Yes	Yes	Yes
Skin cannot be pinched off	Yes	Yes	Yes
Hypo or hyperpigmented patches on skin	No	No	Yes, on face, morphea on both cheeks [Figure 4]
Mask-like or mouse-facies	Yes	Yes	Yes
Obliteration of infraorbital fold	Yes	Yes	Yes
Pinched nose	Yes	Yes	Yes
Lip incompetency with thinning			
Microstomia with puckering	Yes	Yes	Yes
Temperomandibular joint hypomobility	Yes	Yes	No
Restricted mouth opening	Yes	Yes [Figure 5]	Yes
Restricted tongue movement	No	Yes [Figure 6]	No
Xerostomia	Yes	Yes	Yes
Blanching of oral mucosa	Yes	Yes	No
Gingival involvement	No	Yes	Yes
Periodontal involvement	No	Yes	No
Oral ulcerations	Yes	No	Yes, infrequent
Pulpal calcifications	No	No	No
Any other oral involvement	No	No	Oral candidiasis
Features on extraoral radiograph	Periodontal ligament widening in posterior regions of jaws [Figure 7]	Genetic interdental bone loss, bilateral condylar resorption, reduced lower third facial height [Figure 8]	No
Shortened distal phalanges of hands	Yes	Yes [Figure 9]	No
Involvement of great toes	No	Yes	No
Raynaud's phenomenon	Yes	No	No
Telangiectasia	No	No	No

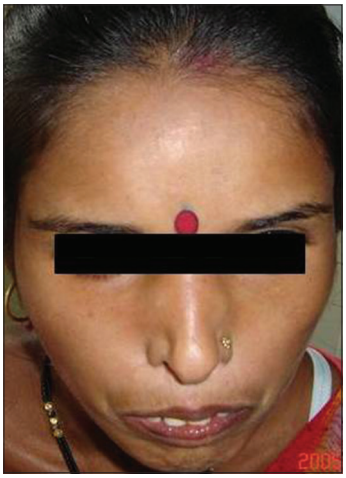

Figure 1: Case 1, showing orofacial appearance with mask-like or mouse-facies as taut facial skin, loss of wrinkles and skin folds, pinched nose, thin lips, puckered mouth, and proclinated anteriors

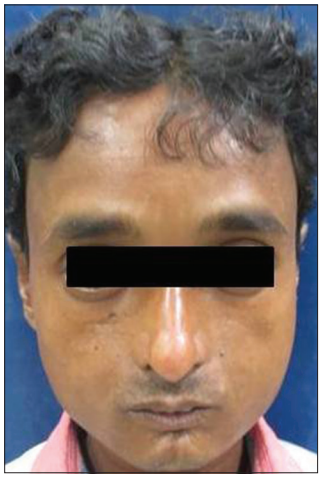

Figure 2: Case 2, showing orofacial appearance with mask-like or mouse-facies as taut facial skin, loss of wrinkles and skin folds, pinched nose, thin lips, puckered mouth, and proclinated anteriors

All the patients were sent to the hospital for the thorough physical and dermatologic evaluation. The clinical, radiographic, and laboratory findings were consistent with a diagnosis of scleroderma.

REVIEW OF LITERATURE

PSS is a connective tissue disorder that involves the skin, mucosa, blood vessels, and the viscera. To the best of our knowledge, it was first mentioned by Curzio of Naples in 1752 as scleroderma, the term is splitted as "*sklero*'' (hard) and ''*derma*'' (skin).[9,10] Since hidebound skin is the clinical hallmark of the disease, it is also called "hidebound disease."[1] Although mainly thought of autoimmune origin, the other etiological factors such as genetic, environmental, nervous, and vascular are postulated to be the causes of the disease.[1,8,11]

Classification

The disease can occur in three forms:

- Circumscribed or localized scleroderma, it is manifested in two forms as morphea and linear scleroderma. Morphea is characterized by localized thickening of the skin. It starts as violaceous or

Figure 3: Case 3, showing orofacial appearance with Masklike appearance and lip puckering

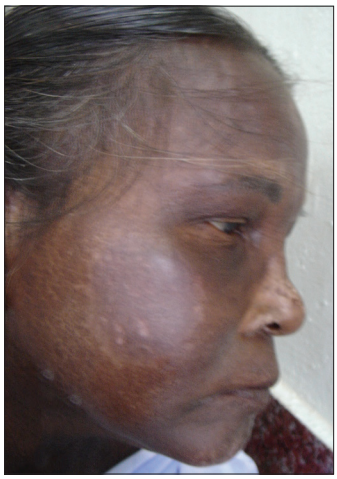

Figure 4: Case 3, Morphea as oval violaceous patch on right cheek and hypopigmented patches on face

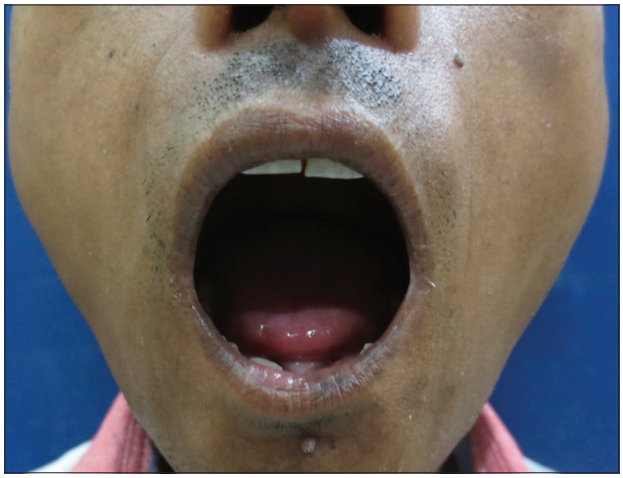

Figure 5: Case 2 showing restricted mouth opening

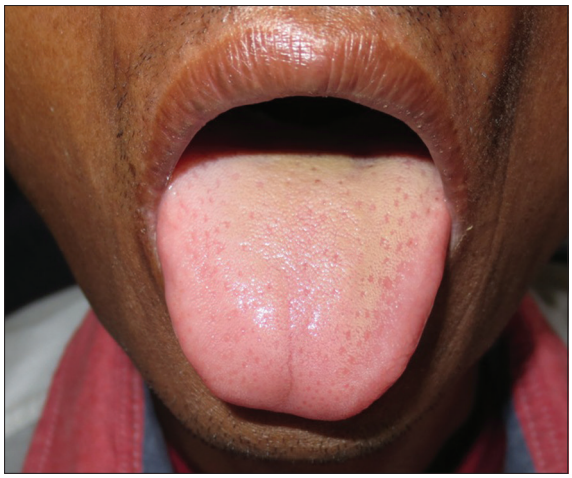

Figure 6: Case 2 showing reduced tongue mobility

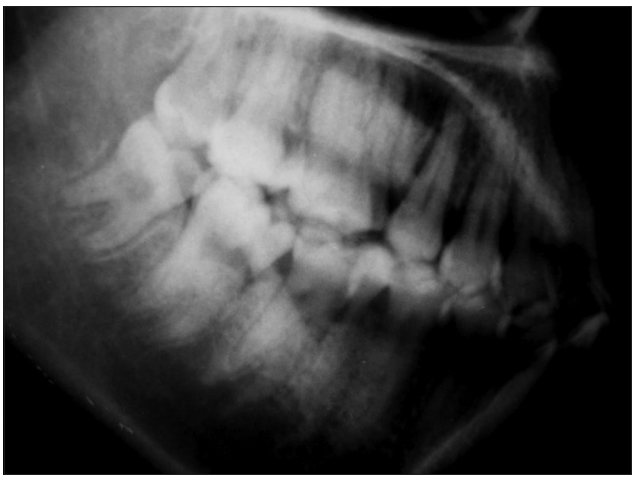

Figure 7: Case 1, lateral view showing periodontal ligament widening with posterior region of jaw bones

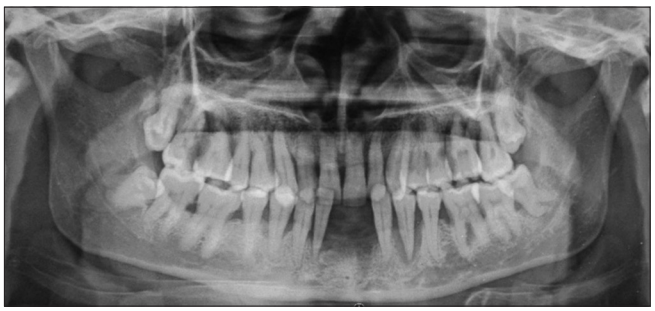

Figure 8: Case 2, showing Orthopantomogram with generalized periodontitis, missing lower central incisors and flattening of condyle on right side

purplish-brown oval skin patches, that enlarge, become indurated, and eventually lose hair and the ability to sweat. Linear scleroderma is a form of the localized disease and develops as a thin band of sclerosis that may run through the entire length of an extremity, involving underlying muscle, bones, and joints.[1,5,6] The lesion of linear localized scleroderma of the head and face is called *en coup de sabre*, and these lesions may result in hemiatrophy of the face[5]

- Generalized or progressive scleroderma (diffuse form-PSS), characterized by tautness of the

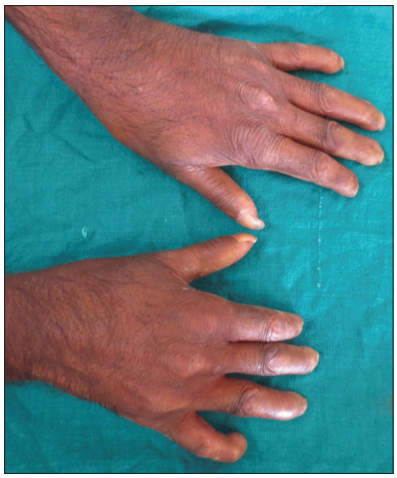

Figure 9: Case 2 showing shortened distal phalanges of hands

skin with distinctive involvement of the lungs, heart, kidneys, and gastrointestinal tract, and osteolytic changes in the skeleton, Raynaud's phenomenon, acrosclerosis, and oral and perioral tissues involvement.[5,12-14]

- Acrosclerosis, a combination of scleroderma of the extremities and Raynaud's disease[14,15]
- A variant of this disease is known as the "crest syndrome," which is an abbreviation for calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility with dysphagia, sclerodactyly, and telangiectases.[15]

Clinical features

PSS is a disease of low incidence, with 4–253 cases per million or an average of 4–19 new cases per million. The blacks or African-Asians are involved more than whites or Caucasians.[1,2,8,13,16] The disorder primarily affects the females in the age group of 30–50 years.[16,17] Although disease manifests variably, the tightening of skin and Raynaud's phenomenon would be the early manifestation. The cutaneous involvement usually begins as the pitting edema followed by thickening and tightening of the skin to include the distal portions of the extremities most commonly. Depending of the involvement, it may lead progressively to pulmonary disease, renal disease, esophageal dysmotility, dysphagia, polyarthralgia with the stiffness of joints, myopathy, myocardiopathy. Subcutaneous calcifications, telangiectasia, and hyperpigmentation may be the features of the disease. The extent of involvement leads to corresponding discomfort, disability, and cosmetic problems.[1,2,8,12-17]

Oral manifestations

The head and neck region is involved in approximately 70% cases of PSS.[6,11,18,19] The characteristic orofacial features are the mask-like appearance or the mouse-facies due to the pinched nose, atrophy of the ala of the nose, thin and rigid lips, and loss of skin fold on the face.[1,6,10,20] Other features are inability to open the mouth wide, reduced interincisal opening, tongue rigidity, xerostomia, pigmentations, mouth ulcers, telangiectasia, periodontal disease, pulpal calcifications, pseudo ankylosis and trigeminal neuropathy,[21] and the secondary manifestations such as mobility of teeth, dental caries, candidiasis, subluxation, and altered mandibular guidance. The esophageal dysmotility may cause acidity induced by gastroesophageal reflux that lead to dental erosion and dental caries.[6] The radiographic features include PDL widening in approximately two-third cases with a predilection for the posterior region of the jaws as compared to the anterior region.[10,20] The pressure resorption of the jaw bone is prominently seen at angle, condyle, coronoid process, gonial angle, posterior border of ascending ramus, and at the muscle insertion sites.[6,15,19,22] The exact etiology of the osteolysis is unknown, but there are three proposed theories: (1) Tightening of the facial skin may exert excessive pressure on the mandible and induce the bone loss; (2) the vasculopathy associated with this disease may diminish the blood supply to the mandible resulting in bone ischemia and necrosis; and (3) atrophy of the muscles of mastication may lead to bone necrosis.[8,15] Calcinosis of the soft tissues around the jaws may be evident.[23]

Diagnosis

The clinical sign of stiffened skin texture and a skin biopsy may confirm the diagnosis.[7] The biopsy reveals the features of atrophy of skin or mucosa, thickening and hyalinization of collagen fibers, and sclerosis of the blood vessel walls. Several serological tests are also useful for confirmation of the diagnosis.[3] Antinuclear antibodies may be seen in approximately 90% of scleroderma patients and are characteristically antinucleolar or anticentromere antibodies or anti-ribonucleic acid polymerase III antibodies.[3,8]

The microscopic changes in the PDL consist of a widening as well as an appearance of hyalinization with diminution in the number of connective tissue cells than usually found.[24]

Orofacial considerations

The narrowing of the oral cavity, submucosal fibrosis, limited oral access, and tongue rigidity make the speaking, eating, and brushing difficult, also the limitation makes the endodontics, prosthetics, surgical, and restorative procedures complicated.[21,25-27] Self-care education using long or extended handled brush or powered toothbrushes or flosses may be of use for daily care. Due to the debilitating nature of the disease, the patient may not be able to sit

in a chair for long time, so that it brings limitation on working time of dental professionals. The mild analgesic taken 1 h before the sitting may add comfort to the patient.[17] The salivary hypofunction due to fibrosis or medications affect the oral health, speech, swallowing, and denture retention.[6,11] Pilocarpine or oral lubricators like carboxymethylcellulose may be of use. Some of the patients may need daily fluoride therapy. The physiotherapy in the form of oral and facial stretching exercises may help to maintain the opening and the oral hygiene maintenance. Use of increasing number of tongue blades between the posterior teeth, Kabat's technique, and mechanical devices like TheraBite to stretch the facial tissues is an effective technique to increase mouth opening.[1,6,8,11,28] The oral opening may improve up to 5 mm by stretching exercises. In case of advanced cases, a bilateral commissurotomy may be necessary.[11,27] In case of gingival and periodontal involvement, maintenance therapy should be adapted in place of surgery as progressive vascular fibrosis and deficient wound healing makes any surgical procedure difficult and harmful in such patients. The mucosal erosions and ulcerations need frequent attention. The antibiotics may need to be prescribed. The 5% tranexamic acid solution rinses are required for patients receiving oral anticoagulants, and the international normalized ratio must be checked on the morning of the day of oral surgery, the adrenaline should be avoided as it may exacerbate the existing micro-angiotherapy. Patients with Raynaud's phenomenon also need the warming aids in the room.[1,11] Patients with extensive resorption of the angle of the mandible are at a risk of developing pathological fractures from minor trauma, including dental extractions.[1,4,5,8]

The treating physicians should be consulted before orofacial management considerations. The early treatment interventions are crucial to avoid the systemic complications out of oral infections. The treatment modalities of the disease can be complex as is the nature of the disease, also the drug therapy of the condition include the calcium channel blockers and immunosuppressive agents which is the another matter of concern.

CONCLUSION

The management of scleroderma patients depends on the extent and severity of skin and organ involvement. Although the disease is not curable, the early diagnosis and personalized therapy as per the case helps to treat this disorder to some extent. The periodic dental evaluation is mandatory, maintenance of existing dentition is imperative as the microstomia and tongue rigidity can make the therapeutic interventions difficult. Self-care education and counseling to refrain from harmful habits such as tobacco, alcohol, betel nut, smoking, and caffeine may be of greater help in such patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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