CASE REPORT

Systemic Sclerosis: A Case Report and Review of Literature

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ABSTRACT

Systemic sclerosis (scleroderma) is a rare generalized disorder of connective tissue origin. It is an auto-immune, collagen disorder and is characterized by diffuse fibrosis that involves skin, muscles, and other internal organs like the GIT, lungs, blood vessels and kidneys. Systemic sclerosis can also affect the facial and oral structures. This condition is predominantly diagnosed by clinical signs and symptoms. Early diagnosis and individually tailored therapy help to manage this disorder which is treatable but not curable. This case report describes the presentation of systemic sclerosis in a 45 year old female patient.

Keywords: Connective tissue disorders, Scleroderma, Restricted mouth opening, Progressive systemic sclerosis.

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INTRODUCTION

Systemic sclerosis is a chronic connective tissue disease of unknown etiology that causes widespread microvascular damage and excessive deposition of collagen in the skin and internal organs. Connoting the pathognomonic clinical appearance of the disorder which is characterised by hard skin, the disease derives its name from Greek language wherein sclerosis means hard and derma means skin. It is also referred to as hidebound disease due to the fact that hidebound skin is the key feature of the disease.[1]

Though the etiology of systemic sclerosis is very unclear it has been hypothesized that injury to vascular system and auto immunity may play a very significant role and also the relevance of multiple risk factors such as age, sex, genetic background and environmental exposure is to be considered.[2] In the initial stage of the disease the skin is infiltrated by T lymphocytes which causes abnormality in activation of fibroblasts which leads to excessive production of collagen.[3] There is an associated proliferation of intima resulting in narrowing of artery and arteriole and inflammation of vessel wall.[4]

Females are affected three to four times more frequently than males. It most commonly occurs between 30 and 50 years of age.[5] Based on the extent of involvement the disease is categorized as localised and diffuse variety. In the diffuse form, there is generalized involvement of skin with quick progression of involvement of internal organs. On the contrary, there is minimal involvement of skin in localised form which is particularly confined to distal aspect of fingers and the face. CREST syndrome is a variant of it associated with calcinosis cutis, Reynaud's phenomenon, esophageal dysmotility with dysphagia, sclerodactyly and telengiectases.[6]

CASE REPORT

A 45-year-old female patient reported to the oral medicine and radiology department, with the chief complaint of stains on her teeth. Her past medical history revealed that she had undergone treatment for skin disease.

On extra oral examination, facial skin was tense, smooth and shiny, taut and mask like (Fig. 1). It was firm and could not be picked up. Nasal alae was atrophied giving a pinched appearance to the nose, resulting in a 'mouse facies'. Stiffening of phalangeal joints in both fingers and toes was observed. Temperomandibular joint was stiff on palpation.

Oral manifestations showed microstomia with characteristic furrows radiating from the mouth, which led 'Purse String Appearance.' The mouth opening was restricted up to 30 mm (Fig. 2). On palpation circum oral fibrotic bands are palpable. Lip thinning and incompetency with prominent anterior teeth was seen (Fig. 3). Oral mucosa showed blanching and paleness, more prominent on the buccal mucosa. There was diffuse fibrosis of buccal mucosa with loss of normal elasticity of mucosa. Tongue was rigid and movements are limited.

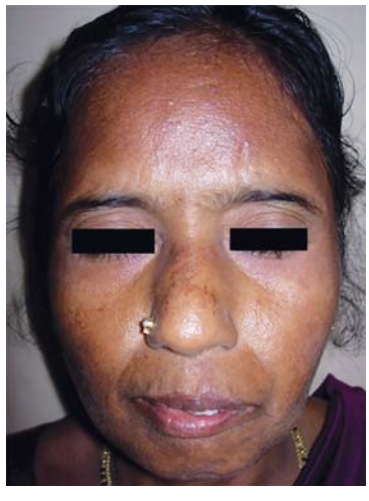

Fig. 1: Facial features in systemic sclerosis

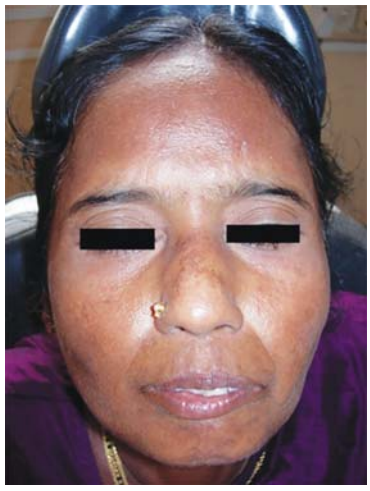

Fig. 2: Restricted mouth opening in systemic sclerosis

Patient was then subjected to radiographic examinations and Intra oral periapical radiograph showed widening of the periodontal ligament space (Fig. 4). Initial therapy of scaling was done. Then the patient was referred to dermatology department for cutaneous problem.

DISCUSSION

Systemic sclerosis (scleroderma) is a multisystem chronic connective tissue disease of unknown etiology that involves hardening of skin and mucosa with smooth muscle atrophy and fibrosis of internal organs.

The historical highlight of scleroderma dates back to 1752 when Carlo Curzio of Naples wrote about it in his monography.[7] It was in 1847 that Gintrac named it as Sclerodermie. Later in 1945 after eliciting the systemic nature of the disease Goetz[8] gave the name as progressive systemic sclerosis.

Unlike any auto immune pathologies there is a female predilection and the disease is common between 3rd and 5th decade of life.[9] The incipient or warning symptom which appears first is a special phenomenon called as Raynaud's phenomenon which is characterized by paroxysmal vasospasm of fingers resulting in colour change of fingertips as a response to cold or emotion and resorption of terminal phalanges.[10]

Also symmetric thickening, tightening and induration of the skin in the distal portion of extremities is the most common symptom.[11] Pitting oedema usually is replaced by tightening and hardening of skin.[12] Functionally atrophy or ischemic damage to the tips of the fingers and contractures preventing which prevents straightening of fingers lead to claw like fingers. Clubbed with morphological deformity cosmetic problems include hyperpigmentation, telengiectasias and subcutaneous calcifications.[13]

Based on the extent of involvement the disease is categorised as localised and diffuse variety. Localized scleroderma can be categorised as circumscribed and linear form. The circumscribed form also referred to as Morphea starts with violaceous patches on the skin burns out as it progresses appearing as hypo- or hyperpigmented area depressed below the level of skin. However in linear localized form, the face is frequently involved and the area of fibrosis resembles the scar from a sabre cut, which may result in hemiatrophy of the face. Scleroderma is referred to as acrosclerosis when it is localized to hands.[14]

The most frequent oral finding to precede systemic involvement appears to be trigeminal neuropathy followed by enlargement of the periodontal ligament (PDL) space.[15] The trigeminal neuropathy is characterized by slow and gradual facial muscle inactivity followed by pain, sometimes severe, and paresthesia. Another oral finding of significance is the increased risk of oral cancer. In particular, a recent study found an increased risk of squamous cell carcinoma of the tongue.[16]

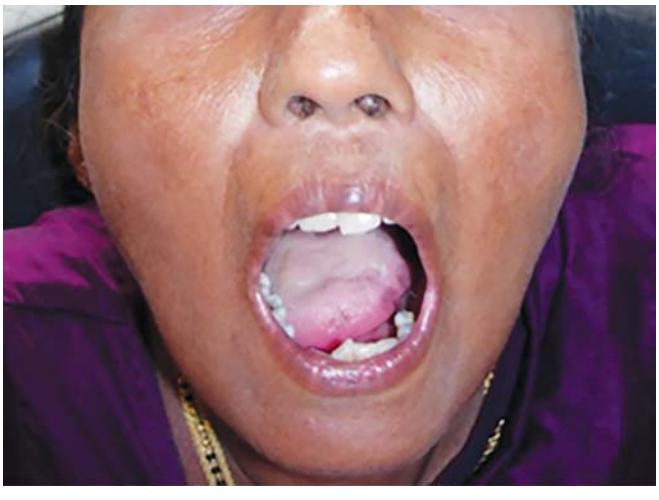

Fig. 3: Incompetent, thin lips with prominent anterior teeth

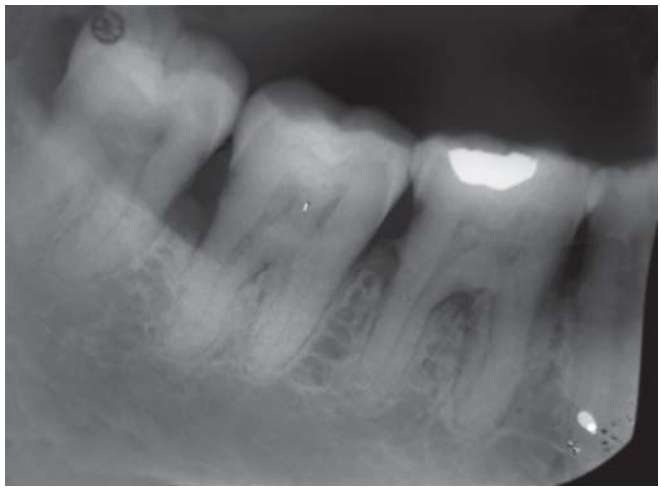

Fig. 4: Intraoral periapical radiograph showing widening of the periodontal ligament space

It includes narrowing of eyes, loss of skin folds around the mouth giving a Mona Lisa face (mask-like appearance). The lips might be constricted or pursed with radiating furrows giving a fish mouth appearance. Involvement of periarticular tissues of temperomandibular joint along with microstomia may greatly limit opening of mouth, causing a pseudoankylosis.[17]

Involvement of oral submucosa may cause the tongue to become stiff and narrowed giving rise to feature called as chicken tongue. Rarely few patients had salivary hypofunction, keratoconjunctivitis sicca or both.[18]

Radiographically, significant 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.[19] Other radiographic features include resorption of the condyle, coronoid process and the angle of the mandible[20] which are apparently related to pressure atrophy or ischemia. Occasionally resorption in the apical part of the roots and complete destruction of the lamina dura has been reported.[21]

Renal involvement is common in systemic sclerosis. Scleroderma renal crises (SRC), a severe and life-threatening renal disease, develops in approximately 10% to 15% of patients with the diffuse cutaneous form of systemic sclerosis, and arises much less frequently in limited cutaneous systemic sclerosis.[22] SRC is an early complication of systemic sclerosis that almost invariably occurs within the first year of onset of the disease.

There is no specific treatment that has demonstrated efficacy in arresting or improving skin changes. Conservative management includes maintenance of core body temperature and avoiding peripheral cold exposure. Ultimately management of systemic sclerosis is focused specifically at the organ/system involved. Localized dermatological lesions have been found to improve with ultraviolet light therapy showing good prognosis in skin tautness.[23] Antibiotics are useful in treating infections of the ulcerated skin.

Maintenance of existing dentition is important because microstomia and tongue rigidity can create difficulty in prosthetic rehabilitation. As part of prevention, proper oral hygiene habits and the importance for good permanent restorative dentistry should be stressed to these patients since progressive constriction of the oral cavity eventually limits access to perform adequate dental treatment. 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. As part of complications, patients with extensive resorption of the mandible are at risk for pathologic fractures from dental extractions.

CONCLUSION

It is imperative to provide appropriate dental hygiene care of these patients which requires knowledge of clinical characteristics, the identification of oral facial manifestations, treatment aspects and pharmacological interventions so as to provide safe, compassionate and effective dental hygiene management and care to these patients.

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