SYSTEMIC SCLEROSIS WITH ORO-FACIAL MANIFESTATIONS AND TREATMENT CHALLENGES: A CASE REPORT

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ABSTRACT

Systemic sclerosis is a multisystem connective tissue disease characterized by inflammation, vascular and fibrotic changes of the skin as well as various internal organs. Common oro-facial findings include xerostomia, oral mucosal and gingival fibrosis, mandibular bone resorption and limited mouth opening which complicates maintenance of oral health and dental treatment. This article reports a rare case of systemic sclerosis with oral manifestations diagnosed mainly based on the clinical signs and symptoms and confirmed by the elevated levels of antinuclear antibodies.

INTRODUCTION

Scleroderma is a connective tissue disorder of excess collagen production characterized by intense fibrosis of the skin, with internal organ involvement. The term Scleroderma is derived from the Greek, means hard skin (skleros=hard, derma=skin). Scleroderma or progressive systemic sclerosis (PSS) which was named by Goetz in 1945 is an autoimmune rheumatic condition affecting the connective tissue and has a profound impact on oral health1. The oral manifestations of the disease result from the deposits of collagen in the tissues or as a result of collagen deposition around nerves and vessels. The oral radiographic finding most commonly recognized is wide periodontal ligament spaces. Limited function, impaired healing and neurologic symptoms all may be present with varying degrees depending on the form and progression of the disease. Oral and facial manifestations lead to major functional discomfort, mainly due to decreased mouth opening that seems to be frequently associated to oesophageal involvement. Xerostomia is also frequent and is commonly observed in anticentromere antibodies positive cutaneous limited forms of systemic sclerosis (Vincent et al 2009)2.

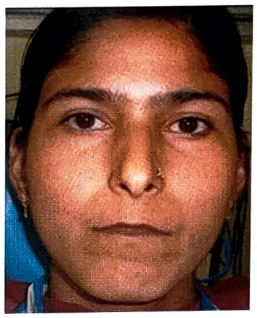
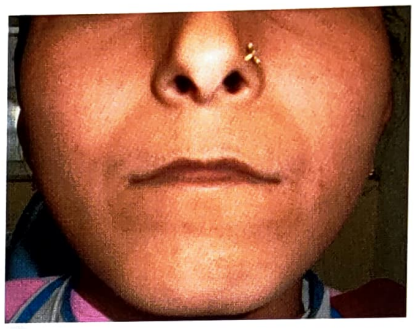


Fig 1



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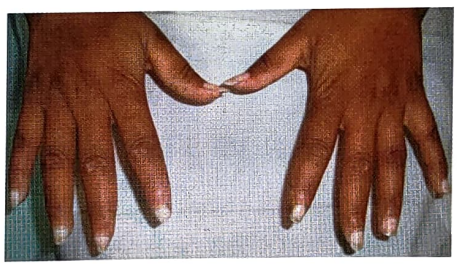

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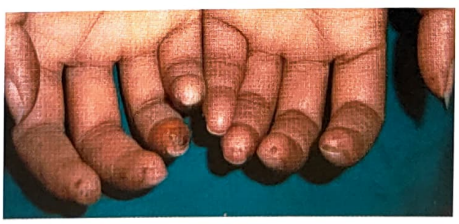

Fig 4

Females are known to be affected seven times higher than males3. This disease is broadly classified as localized scleroderma and systemic sclerosis. Localized scleroderma is further divided into linear scleroderma, localized and generalized morphea. Systemic sclerosis is subdivided into limited or diffuse cutaneous systemic sclerosis. In scleroderma, anti-nuclear antibodies against topoisomerase or centromeres result in fibroblasts producing excess collagen. A gene which codes for fibrillin-I may put some people at risk for scleroderma. The suspected trigers for scleroderma include viral infections, adhesive and coating materials, organic solvents such as vinyl chloride or trichloroethylene, silicone breast implants have been

blamed but evidence has not been found for this4. Here, we report a case of systemic sclerosis with oro-facial manifestations with diffuse cutaneous involvement and its management. This article emphasizes the importance of clinical diagnosis and treatment challenges in the management of systemic sclerosis patients with oro-facial findings.

CASE REPORT

A 21 years old female patient reported with a chief complaint of bleeding gums and bad breath. Extra-oral examination revealed tight and shiny skin with inability to wrinkle and express (mask like), and unable to be picked or pinched **(Fig. 1)**.

Lips appeared to be thin and characteristic microstomia with furrows radiating from the commissure bilaterally giving rise to purse string appearance **(Fig. 2)**.

There was stiffening at the tempero-mandibular joint bilaterally. Finger tips were blanched and narrow with bulbous and hard nail beds with difficulty to bend or stretch the fingers **(Fig. 3)**.

Crusted scars on the fingers were present which increased during the winter **(Fig. 4)**.

Patient also complained of exertional dyspnoea and palpitations.On intra-Oral examination, buccal mucosa and palate appeared to be dry and shiny. There was limited mouth opening **(Fig. 5)**

Gingiva was inflamed and enlarged in the maxillary and mandibular anterior teeth region. Oral hygiene was poor **(Fig. 6)**.

Patient complained of dry mouth and discomfort during chewing and swallowing. Radiographic examination revealed generalized widening of the periodontal ligament spaces specifically at the posterior teeth **(Fig. 7)**.

Hand wrist radiographs showed resorption at the terminal phalanges **(Fig. 8, Fig. 9)**.

Based on the history of bluish discoloration of skin, whole body and face (skin) tightening, difficulty to extend or flex the terminal phalangeal joints, bulbous and hard nail beds, microstomia, xerostomia, and discomfort during swallowing, occasional dyspnoea and palpitations, radiographic findings enabled us to diagnose this case of progressive systemic sclerosis with diffuse cutaneous involvement.Serological tests showed granulocytosis, ESR was 17mm/hr, SGOT 35 IU/L. Urine examination revealed only few pus cells and epithelial cells. Chest X-ray was normal. Echo cardiogram was normal.The antinuclear antibodies test was positive. Serum anti-centromere antibodies were elevated thus correlating the disease severity. Anti-nuclear antibody test showed the end point titre at 1:1280 which was of high clinical significance. ANA positivity of greater than or equal to 1:160 titre is of clinical significance in collagen vascular disorders.

TREATMENT

Patient was administered antibiotics, Calcium channel blockers for angina, ecosporin as a prophylactic drug against arterial occlusive events like stroke etc., angiotensin-2 receptors antagonist, antacids, topical steroids application for skin. Nutritional supplements like zincovit, calcium supplements were advised. Dental treatment consisted of oral hygiene instructions, scaling and root planing, mouth rinses were prescribed. Since the inflammation reduced by phase I therapy no further periodontal treatment was required. Patient was recalled after every 2 weeks for follow-up. Oral hygiene maintenance was improved **(Fig. 10)**.

Patient is still under follow-up for treatment of the skin lesions.

DISCUSSION

Systemic sclerosis is characterized by progressive fibrosis of skin and multiple organs. Progressive systemic sclerosis (PSS) is a systemic disease that affects many organ systems, including the skin, gastrointestinal tract, respiratory, renal, cardiovascular and genitourinary systems. The onset of the systemic sclerosis is usually insidious; its most dramatic effects are cutaneous changes. Earlier genetic, environmental, autoimmune, vascular, nervous factors were proposed to be involved in the etiology of the disease5-7. Progressive tissue fibrosis ensues as normal type I and III collagen are deposited in extraordinary amounts,

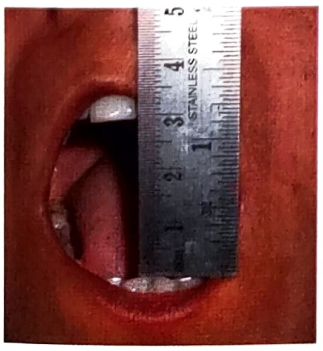

Fig 5

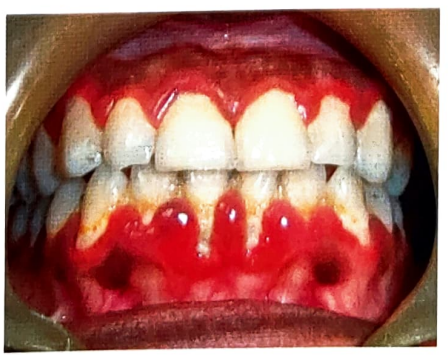

Fig 6

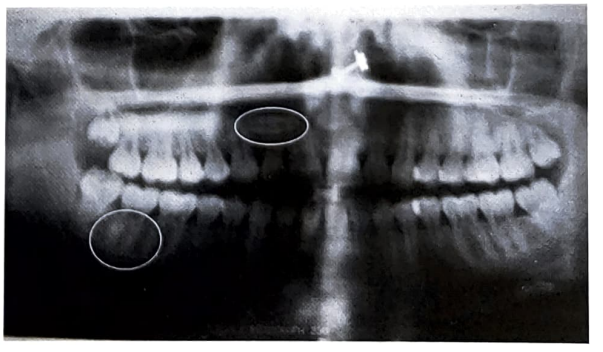

Fig 7

likely because of immunologically over activated fibroblasts in various locations. There is a quantitative increase in the amorphous ground substance, glycosaminoglycans and fibronectin, within the connective tissues. Vascular alterations affect the small arterioles, resulting in thickening of the vessel walls and reduction of the diameter of the lumen8.

Subcutaneous collagen deposition in facial skin results in a characteristic smooth, taut, mask-like facies. Other important orofacial manifestations include fibrosis of the salivary and lacrimal glands, and symptoms consistent with dry mouth or xerostomia. Some patients may develop dry eyes with keratoconjunctivitis sicca or xerophthalmia. This is particularly problematic because scarring of the eyelids results in a chronic widening of the palpebral fissures and inadequate closure of the eyelid, which causes further drying of eyes. Inadequate salivary flow compromises buffering within the oral cavity and allows the acidity produced by bacterial metabolism and GERD (Gastroesophageal reflux disease) to erode the dentition. Further, facial and mucosal fibrosis compromises oral access because of microstomia, which limits mouth opening in 70% of these patients9. As a consequence, oral hygiene and fabrication of removable dentures, other dental procedures are difficult because of limited access and the obliteration or shallowing of the mucobuccal folds.

Management of the systemic effects of this disease is not well established, although some large uncontrolled series suggest that D-penicillamine has beneficial effects10. Interferon-gamma is effective, but its use is limited because of inflammatory sequelae. GERD is managed with antacids, H2 blockers, proton-pump inhibitors, prokinetic agents, smaller meals and laxatives11.

Control of operatory temperature and patient comfort are crucial for preventing cold- or stress-induced vasospasm in the dental setting. When pulmonary function is compromised, supplemental oxygenation is indicated during dental procedures. Renal crises are managed with aggressive use of angiotensin-converting enzyme inhibitors at the earliest signs of hypertension12. Removal of severely painful, draining or infected calcinotic deposits is occasionally required. Large dietary doses of vitamin C (> 1 g/day) should be avoided by patients with PSS since vitamin C is integral to collagen formation and deposition. Patients with limited range of motion and mouth opening can benefit from regular physical and occupational therapy to maintain range of motion and to minimize or delay contractures.

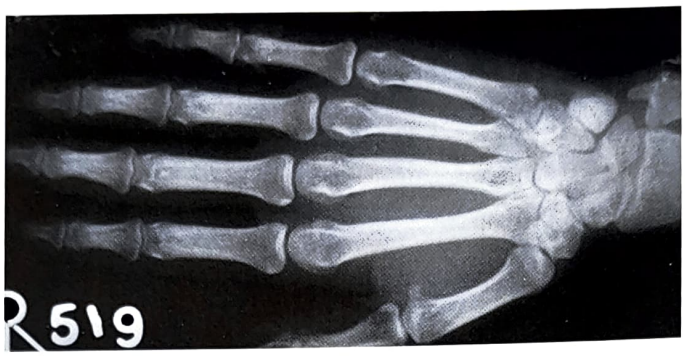

Fig 8

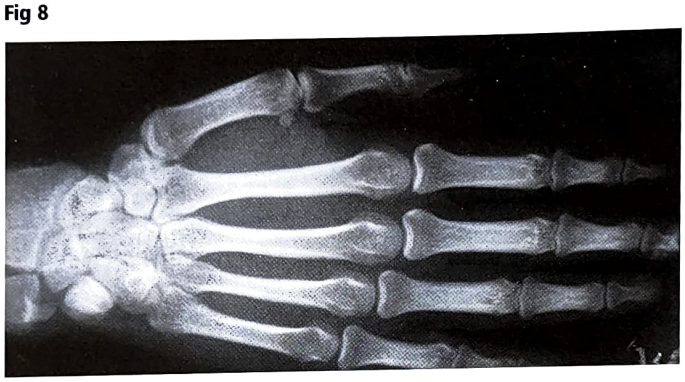
Fig 9

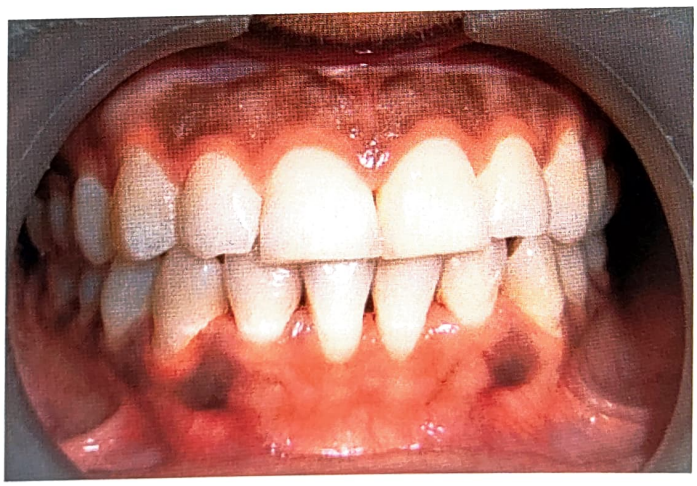
Fig 10

CONCLUSION

Dental practitioners may be the first to note some of the signs of progressive systemic sclerosis. The concomitant presence of GERD, xerostomia and limited mouth opening may alert practitioners to the possibility of scleroderma. Special care and attention must be paid to keeping the surrounding environment in the dental operatory warm for patients who have vasospasms to help avoid a vascular crisis. Patients with severe restrictive lung disease may require supplemental oxygen. Instructions and reinforcement of oral hygiene, along with frequent dental assessment and management by the dental practitioner are essential measures to preserve the oral health of those affected with PSS.

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