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CASE REPORT

Clinicoradiological Findings in Progressive Systemic Sclerosis: A Case Report

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ABSTRACT

Scleroderma is a multisystem connective tissue disease affecting small arteries, microvessels and fibroblasts result in vascular obliteration, collagen accumulation and scarring of skin and internal organs. This condition can be localized or systemic. Its estimated prevalence is 250 cases in a million and it is more common in women than in men. It is presented with wide variety of clinical as well as radiological oral and maxillofacial findings such as resorption of the mandibular angle and coronoid process. Decreased number of wrinkles due to sclerosis and distinct facial features because of the atrophy of ala nasi. The aim of this case report is to present a 43-year-old female patient with Progressive systemic sclerosis who presented with various general physical, extraoral and intraoral manifestations.

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INTRODUCTION

Scleroderma is a multisystem connective tissue disease affecting small arteries, microvessels and fibroblasts result in vascular obliteration, collagen accumulation and scarring of skin and internal organs. It is a disease of immune system, blood vessels and connective tissue.The word scleroderma originates from Greek words "scleros" meaning hard and "derma" which means skin. It is characterized by thickening and fibrosis of skin. Since hidebound skin is the clinical hallmark of the disease. It is called as "hidebound disease". The exact etiology is unknown but it is characterized by both vascular injury and excessive production of normal collagendue to stimulation of fibroblast caused by mediators produced by T-lymphocytes.[1,2]

It is of two types: progressive systemic sclerosis which includes CREST syndrome and diffuse cutaneous scleroderma and localized scleroderma which includes linear scleroderma and morphea.[3]

One of the first signs of the disease is Raynaud's phenomena. Scleroderma causes various changes at the oral and facial tissues both clinically and radiographically. Clinically includes purse string appearance, microstomia, mouse like faces and radiographically includes widening of periodontal space, loss of lamina dura, and resorption of the mandibular angle.[4] The aim of this case report is to present some clinical and radiographic characteristics of a patient diagnosed as having Progressive systemic sclerosis.

CASE REPORT

A female patient aged 43 years presented to the Department of Oral Medicine and Radiology with the chief complain of pain in upper and lower gums on chewing for 20 days. Patient gave history of pain in maxillary and mandibular gingiva on chewing along with xerostomia since past 20 days. Patient also presented with history of difficulty in chewing and

swallowing since 1 year. Along with history of bluish discoloration of fingers of hands and feet on exposure to cold since past 20 years. Patient is hypertensive and is under medication for the same for 20 years. No relevant dental history was reported.

General physical examination of the patient revealed sclerodactyly (shiny and pale hidebound skin over fingers of hand and feet), along with claw like hands, calcinosis cutis and hardening and swollen nail beds. Raynaud's phenomenonwas positive. (Fig 1)

Extra orally skin over the face showed generalized hyperpigmentation along with contrast hypopigmented areas along with multiple erythematous macules over nasal bridge as well as bilateral malar region. There were telangiectasias over nasal bridge. There was thinning and rigidity of the upper lip along with microstomia. There was characteristic "purse string appearance" around the corners of mouth along with thinning of ala of nose fiving characteristic "Mouse like appearance". There was tightening over bilateral masseter muscle along with microstomia. (Fig 2 and 3)

Intraorally,mouth opening was reduced i.e 23mm (Fig 4) and the gingiva was pale and abnormally firm along with generalized gingival recession. (Fig 5) There were tan brown pigmented macules present over the tongue. (Fig 6) Tongue movements were not restricted. Tightening of the mucosa over soft palate and uvula. Xerostomia was present and reduced salivary flow was observedon manipulation of the parotid and submandibular glands. There was positive "lipstick", "tongue blade" and "thread" sign along with generalized gingival recession.

On the basis of above-mentioned clinical findings, a provisional diagnosis of Generalized periodontitis secondary of Progressive systemic sclerosis was made.

Patient was advised radiographic examinations i.e., orthopantomogram, ultrasonography of bilateral submandibular glands and Sialography for bilateral submandibular glands. Along with routine blood investigations, thyroid function tests, renal, liver and pulmonary function tests along with serum electrolytes and cholesterol analysis, CT chest, Anti-nuclear antibody (ANA) testing and coomb's test, which revealed patient's haemoglobin was low 9gm/dl, along with reduced MCV (Mean corpuscular volume) and MCH (Mean corpuscular Haemoglobin). TFT, RFT, LFT and PFT were within normal limits. Serum chloride level was increased and serum HDL and LDL levels were also deranged. ANA was strongly positive. OPG revealed generalized horizontal alveolar bone loss, along with radiographically missing right maxillary first molar and restored left mandibular 1st molar. There was generalized widening of periodontal ligament in maxillary teeth. CT chest revealed Interlobular septal thickening with subpleural honeycombing in bilateral lower lobes and in the anterior segments of bilateral upper lobes suggestive of Interstitial Lung disease. Ultrasonography of bilateral submandibular gland revealed minimal inhomogeneity with hypoechoic area in right submandibular gland giving score of 1 in SGUS (Salivary gland Ultrasonography). Sialography of the Right Submandibular gland revealed dense uniform radio-opacity along the course of Wharton's duct bifurcating at the level of right mandibular third molar with maximum width of 3-2 mm at the middle of the primary duct. The main duct tapers towards the periphery of the gland showing "bush in winter appearance" with multiple branches extending laterally from the main duct suggestive of Mild dilatation with respect to right Wharton's duct and grossly normal study for right submandibular gland. (Fig 7)

On the basis of clinical examination, history taking and various above mentioned investigations final diagnosis of Progressive systemic Sclerosis was made. Patient is managed via combined approach of Rheumatologist, Internal Medicine specialist, pulmonologist, and Oral Medicine specialist and was prescribed with Calcium Channel blockers for Raynaud's phenomena, D-penicillamine for skin tightening, cyclophosphamide for interstitial lung disease, carboxymethyl cellulose for xerostomia. Patient was advised for oral prophylaxis along with root planing and curettage along with mouth opening exercises.

Patient is on periodic follow up since then.

FIG 1

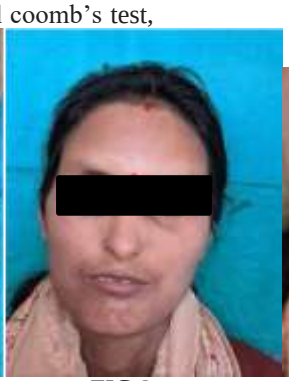

FIG 2

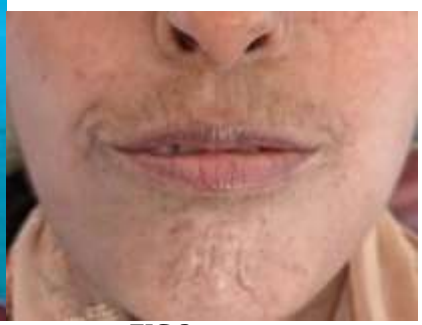

FIG 3

FIG 4

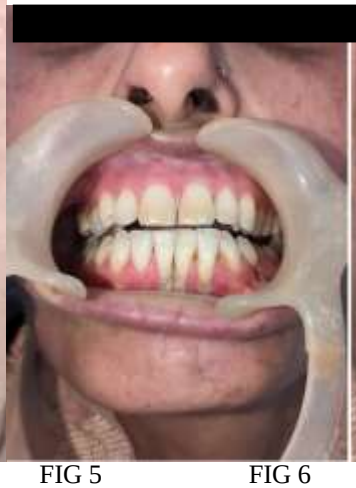

FIG 5

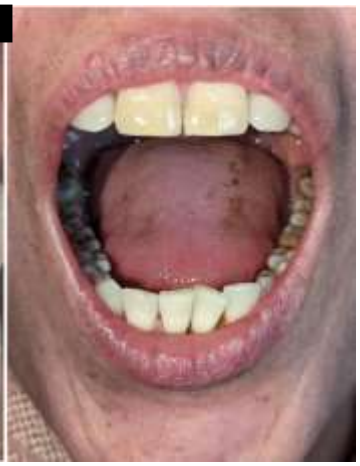

FIG 6

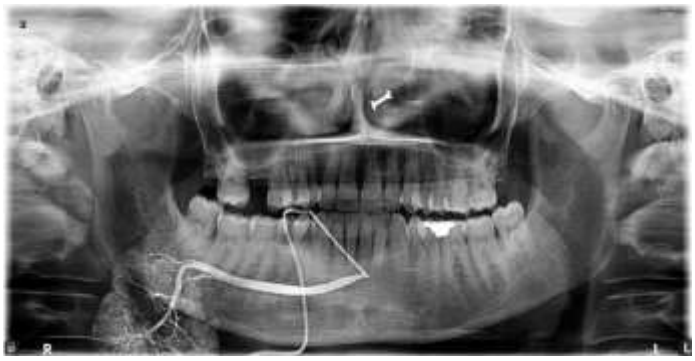

FIG 7

DISCUSSION

Scleroderma is an autoimmune disease in which the antibodies target blood vessels and connective tissues.[5] There is no particular treatment protocol.[5] Although being controversial, penicillamine has been used to inhibit collagen deposition.[6] Otherwise it is treated symptomatically and treatment for scleroderma is selected and organized according to the affected organs.[7]Raynaud's phenomenon, resorption of the terminal phalanxes, claw-like fingers, ulcers on finger tips are common in patients with scleroderma.About 80% of them present with symptoms in the head and neck region.[8] Dysphagia and gastroesophageal reflux are frequently reported complaints in this group of patients. Trismus, widening of the periodontal space (PDL), decrease in facial wrinkles owing to fibrosis of skin, orofacial telangiectasia, resorption of mandibular angle are some of the changes which may occur in the oral and maxillofacial region.[1] Rarely, mandibular resorption can lead to pathologic fractures.[2] Similarly our case presented positive Raynaud's phenomena, claw like fingers, dysphagia, widening of PDL space, fibrosis of skin, telangiectasias showing features of CREST syndrome(Calcinosis cutis, Raynaud's phenomena, oesophageal dysmotility, Sclerodactyly, and Telangiectasia).

Excessive collagen deposition in the cutaneous tissues around the mouth causes microstomia which prevents the patient from opening and closing the mouth. Fibrosis of salivary glands leads to xerostomia, dysphagia, and subsequently periodontal infections.[2,5]Xerostomia, microstomia and severe periodontitis were present in our case. Along with above findings,interstitial lung disease was observed in our case suggestive towards the diagnosis of Progressive Systemic Sclerosis.

CONCLUSION

Scleroderma is a systemic disease which causes significant radiographic and clinical changes in the oral and maxillofacial structures. Dental professionals must be aware of these changes since xerostomia and limited mouth opening are especially important for the right diagnosis and treatment. Multidisciplinary approach is of utmost importance for the scleroderma patients because of the complications of disease.

Conflict of Interests

There is no conflict of interests regarding this paper.

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