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Scleroderma: Unique Fish Tail Radiographic Appearance- A Case Report

Dr. Akanksha Bhandari[1*], Dr. Chetan Bhadage[2], Dr. Ajay Bhoosreddy [3], Dr. Gayatri Mehrotra[4], Dr. Priyanka Vedpathak[5]
[1]PG student, Department of Oral Medicine & Radiology, MGV's KBH Dental College & Hospital, Nashik, Maharashtra, India
[2]Reader, Department of Oral Medicine & Radiology, MGV's KBH Dental College & Hospital, Nashik, Maharashtra, India
[3]Professor & Head, Department of Oral Medicine & Radiology, MGV's KBH Dental College & Hospital, Nashik, Maharashtra, India
[4]MDS, Oral Medicine & Radiology, MGV's KBH Dental College & Hospital, Nashik, Maharashtra, India
[5]MDS, Oral Medicine & Radiology, MGV's KBH Dental College & Hospital, Nashik, Maharashtra, India

Case Report

***Corresponding author**
Dr. Akanksha Bhandari

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Abstract: Progressive systemic sclerosis (Scleroderma) is a generalized auto-immune, connective tissue disorder which is characterized by fibrosis that involves skin, muscles, and internal organs like the GIT, lungs, heart, blood vessels and Kidneys. Systemic sclerosis not only affect the facial structures but also oral structures thus presenting a diagnostic dilemma. Oro-facial and radiographic manifestations include mask like facial appearance, microstomia, restricted mouth opening, xerostomia, periodontal diseases, malocclusion, widened periodontal ligament (PDL) space, pseudo ankyloses, pulpal calcifications, and osseous resorption. Early diagnosis and appropriate therapy help to manage this disorder which is treatable but not curable. Hereby, presenting a case of systemic sclerosis in a 43 year old female patient with mask like face appearance, digital ulceration, claw like deformity of hands, reduced mouth opening, and fibrosis of oral mucosa. OPG showed generalized PDL space widening and resorption at posterior border of ramus of mandible. The distinctive presentation enabled us to give the diagnosis of progressive systemic sclerosis. Treatment should be initiated at its earliest because of various complications reported. Therefore, an adequate knowledge of clinical characteristics, oral manifestations and therapeutic aspects is essential in order to provide an effective treatment to these patients.
Keywords: Scleroderma, Widened PDL space, Resorption, Connective tissue disorder

INTRODUCTION

Scleroderma is a connective tissue disorder and is characterized by fibrosis of skin, blood vessels and internal organs [1]. The term Scleroderma is derived from Greek word *skleros* which means hard and derma meaning skin. It can be both systemic as well as localized in involvement. The name progressive systemic sclerosis was proposed when the systemic nature of the disease became evident [2]. It is also known as "Hidebound disease" since hidebound skin is a characteristic feature of the disease.

Systemic sclerosis is a chronic inflammatory disorder of unknown etiology that cause microvascular damage and is characterized by excessive deposition of collagen in skin and internal organs [3,4]. The incidence is quite low with females being affected 3-4 times more frequently than males. People between the age range of 30 and 50 yrs are more commonly affected [5].

The exact mechanism of fibrotic changes is not yet known; however, hyperplastic changes of collagen have been reported. Moreover, inflammatory changes and globulin deposits have also been focused in blood vessel wall which explains the basis for altered collagen [6]. As per the pathological findings, it has been found that fibroblasts are activated to produce excessive amount of collagen and other components of cellular matrix [7]. It occurs in 2 main forms- that is localized and systemic. In localized form, there is involvement of skin and subcutaneous tissue whereas in generalized form, apart from skin, fibrosis is also present in internal organs such as blood vessels, GIT, heart, lung and kidney. Cutaneous manifestation includes thickening of skin, which initially starts as pitting edema and is thereafter replaced by tightening and hardening of skin [8].

Raynaud's phenomenon is usually the first symptom seen in these cases. Oral manifestations include microstomia, tongue rigidity and there is a classical skin hardening which gives face 'mask like appearance'. Radiographic features commonly seen are generalized widening of PDL space and bone resorption at angle of mandible [9]. Treatment of Scleroderma is itself quite challenging, thus should be diagnosed at its earliest. This article reports a case of clinical and radiographic manifestation of systemic sclerosis.

CASE REPORT

A 43 yrs old female patient reported to the department of Oral Medicine and Radiology with the chief complaint of carious teeth and decreased mouth opening. Further, patient revealed that since last 2 yrs, she is experiencing dryness of mouth especially during night time and difficulty in swallowing and mastication. Patient also reported that taste perception of food has been changed. Her medical history revealed that she had been diagnosed with polyarthritis, interstitial lung disease, anaemia and upper respiratory tract infection. Patient was on d-penicillamine and other calcium and iron supplements for the same. On general examination, it was found that patient had a thin built with pallor present and her weight was reduced to 35.6 kg. Her pulse rate was high with 112/min and low blood pressure 100/60 mm of Hg.

Extraoral examination showed that the skin of the face was stretched with loss of skin folds giving mask like appearance. Fingers of hand showed claw like deformity with bandage over the fingers since the patient was suffering from digital ulcers. Skin of both hands and feet appeared stretched and thickened. Patient was having microstomia with interincisal opening of 31 mm. Tenderness and clicking was present with both left and right TMJ. On intraoral examination, blanching was found with both left and right buccal mucosa and fibrous bands were palpable. De-papillation was seen on dorsal surface of tongue and was stiff on palpation with restricted tongue movements. Gingiva was pale pink and firm in consistency. Root piece was present with 32,38 and proximal caries with 12,13,22,23,31,37,41,42,43,44,45,46,47. Intra oral radiographs showed generalized and symmetrical widening of PDL space. Panoramic radiograph showed resorption at posterior border of ramus with both sides and generalized widening of pdl space. Hand wrist radiographs also revealed resorption at the interphalangeal joints. Patient was subjected to ANA test which was positive. Sm/RNP, Scl 70, Ro52 and SSA was also positive. Hematological reports revealed reduced hemoglobin 8.2 gm% and raised ESR 50mm. Thus, on the basis of clinical, radiographic and laboratory findings, patient was diagnosed with systemic sclerosis.

Patient was advised for oral prophylaxis followed by restoration of carious teeth and replacement of missing teeth. Artificial salivary substitutes were given to the patient to be used as rinse along with topical fluoride application as patient was having reduced salivary flow. Mouth opening exercises were advised for the improvement of mouth opening. Patient was recalled after every 1 month for regular follow up.

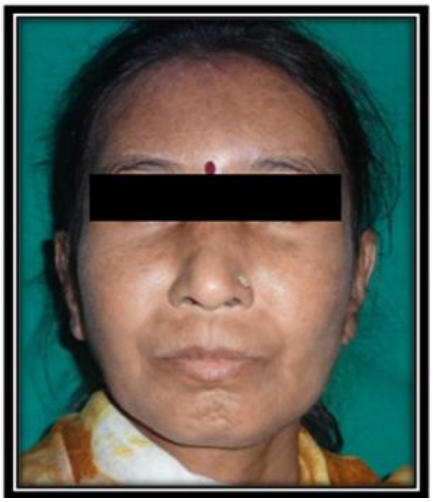

Fig-1: Face showing mask like appearance

Fig-2: Fingers showing claw like deformity

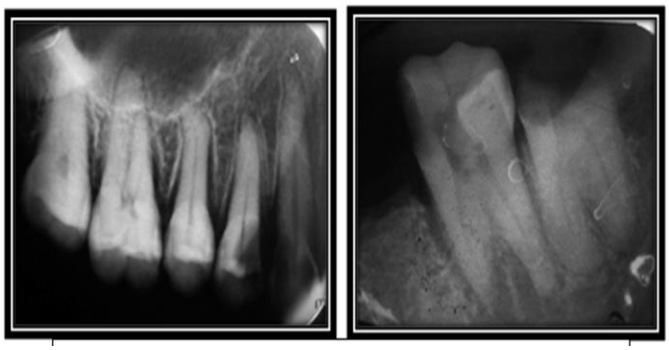

Fig-3: IOPA Showing Generalized and Symmetrical widening of PDL space

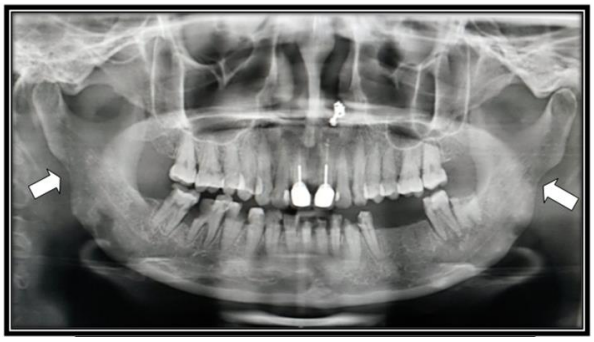

Fig-4: OPG showing resorption at posterior border of ramus giving a "fish tail appearance" and generalized widening of PDL space

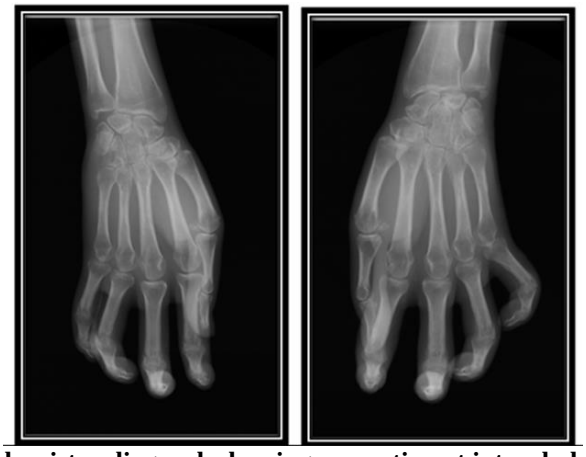

Fig-5: Hand wrist radiograph showing resorption at interphalangeal joint

DISCUSSION

Scleroderma or PSS is a quite rare condition [10]. It was first described by Curzio of Naples in 1752 [11]. The name PSS was proposed by Goetz in 1945, when systemic nature of the disease was proven [2]. The pathogenesis of SSc involves the following-

- Firstly, vascular dysfunction that manifests as damage to endothelial cells.
- Immunological activation of T cells, cytokines and inflammation.
- Fibrosis.
  Localized Scleroderma has been classified into 3 types by Tuffanelli and Winkelmann-
- Circumscribed sclerotic plaques (morphia)
- Streak on the skin (Linear Sc)
- Generalize morphia widespread skin involvement with multiple plaque and frequent muscle atrophy.
- Lepoy *et al.* classified systemic scleroderma as –
- Limited cutaneous SS which was formally called CREST Syndrome (Calcinosis, Raynauds phenomenon, Esophageal dysmotility, Sclerodactyly, Telangiectasia)
- Diffuse Cutaneous SS: Involves sclerosis of face, trunk, proximal extremities.
- SS sine Scleroderma- no skin thickening only organ fibrosis.

Clinical manifestation of SS depends on involvement of site. The earliest symptom which appears in these cases is Raynaud's Phenomenon which is characterized by paroxysmal vasospasm of fingers causing a change in color of fingertips as a reaction to cold or emotion [12]. Another most common symptom seen is symmetric thickening, tightening and induration of skin in distal portion of proximities [13]. It initially starts as pitting edema and is thereafter replaced by tightening and hardening of skin [14]. Claw like deformity is seen with the fingers because of functional atrophy or ischemic damage to tips of fingers. Patient suffers from cosmetic problem such as hyperpigmentation, telangiectasia and subcutaneous calcification.

Most common oral finding appears to be trigeminal neuropathy which is then followed by enlargement of PDL space [15]. Another oral finding of significance reported in these cases is the increased risk of oral cancer, particularly, SCC of tongue [16]. There is narrowing of eyes and loss of skin folds around the mouth giving mask like appearance known as Mona Lisa face. The lips become constricted or fissured showing fish mouth appearance. Due to the excess collagen deposition in peri-oral structures, the temporomandibular joint is also affected causing microstomia that is limited mouth opening leading to pseudoankylosis [17]. Due to fibrosis and atrophy of major and minor salivary glands and lacrimal glands, patient also suffers from xerostomia and keratoconjunctivitis sicca [18]. Tongue becomes stiff and rigid with restricted movement giving rise to feature called chicken tongue.

Most characteristic radiographic feature is widening of PDL space at expense of alveolar bone especially around posterior teeth [19]. Other radiographic features include resorption of condyle, coronoid process and angle of mandible [20] in areas where there is muscle attachment due to excessive pressure exerted. In some cases, complete destruction of lamina dura and resorption at apical end of root has been found.

Scleroderma Renal Crisis is a severe life threatening renal disease found more commonly in diffuse form of Systemic Sclerosis than limited form [21]. Apart from renal, GIT involvement also takes place which causes dysphagia and esophageal dysfunction. Pulmonary manifestation includes interstitial lung disease, pulmonary hypertension, pleuritis, pleural effusion and aspiration pneumonia [22]. Cardiac involvement includes pericarditis, congestive heart failure and conduction problems.

Treatment is quite difficult in such cases because of microstomia and tongue rigidity. Thus, prevention is essential in these cases by maintaining proper oral hygiene habits. Artificial salivary substitutes are given for patients with xerostomia. Oral exercises are advised to increase the mouth opening such as use of tongue blades. Moreover, patients with increased resorption of mandible are at increased risk of pathological fracture at time of extraction.

CONCLUSION

Scleroderma is a multisystem connective tissue disorder with oral and cutaneous manifestations. Treatment should be initiated at its earliest because of a number of complications reported. Therefore, an adequate knowledge of clinical characteristics, oral manifestations and therapeutic aspects is essential in order to provide an effective treatment to these patients.

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