Case Report

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Scleroderma with Typical Manifestations: A Rare Entity

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Abstract

Systemic sclerosis is a connective tissue disorder with a variety of oral manifestations. This case reports we make an attempt to correlate oral and systemic findings that enable the clinician for a better diagnosis and to make a treatment plan. After obtaining the patient's informed consent, relevant medical history and oral manifestations were recorded. The oral changes included restricted mouth opening and widening of periodontal ligament space radiographically and resorption of the mandible. Patients with systemic sclerosis often show wide range of oral manifestations, which is of major concern for the dentist.

Keywords: Scleroderma; Systemic; Calcinosis; Periodontal ligament; Telangiectasis.

Introduction

Systemic sclerosis is a multisystem disorder, clinically characterized by thickening of the skin caused by excessive accumulation of connective tissue.[1–3] The variants of disease are diffuse cutaneous scleroderma, which often shows involvement of proximal and distal extremities, face, trunk, kidney, and visceral organs and the second one limited cutaneous scleroderma, which is limited to distal extremities and face, and shows features of CREST syndrome.[2–4] This syndrome includes *c*alcinosis, *R*aynaud's phenomenon, *e*sophageal dysmotility, *s*clerodactyly, and *t*elangiectasia.[3–6] The involvement of visceral organs may also occur in the absence of skin involvement, which is referred to as systemic sclerosis sine scleroderma.[2,3,5–7] The incidence shows more predilections for female compared to male.[3–5,7,8] The exact etiology is unknown, but it is characterized by both vascular injury and excessive production of normal collagen due to stimulation of fibroblasts caused by the mediators produced by T-lymphocytes.[2,4–7,9] A case report of a female who presented to our department with features of scleroderma is presented here.

Case Report

A 60-year-old female patient was referred from the Departments of General Medicine and Dermatology for dental evaluation. Patient was under taking treatment for diabetes mellitus, hypertension and scleroderma. The rest of the history was non contributory.

Extraoral examination revealed generalized pallor, rigidity of hands, limited movements of fingers and restricted mouth opening (Figs. 1,2 and 3). Intraoral examination revealed rigidity, restricted tongue movements (Fig. 4). Multiple carious and periodontally involved teeth were detected. A provisional diagnosis of chronic generalized periodontitis was arrived. The differential diagnosis was arrived with anemic stomatitis and early OSMF. Panoramic radiograph revealed multiple teeth with periapical and periodontally involved teeth, generalized widening of periodontal ligament space, tooth floating in air appearance (Fig. 5). The final diagnosis of scleroderma was arrived.

Fig. 1: Extraoral photograph showing limited mouth opening.

Fig. 2: Photograph of hands.

Fig. 3: Photograph of hands.

Fig. 4: Intraoral photograph showing rigid and fissured tongue.

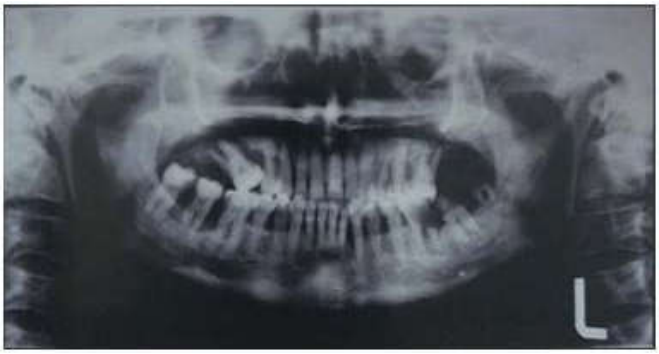

Fig. 5: Panoramic radiograph showing generalized widening of periodontal ligament space, generalized horizontal bone loss and tooth floating in air appearance.

Discussion

Systemic sclerosis is a generalized disorder of small arteries, micro vessels and connective tissue, characterized by fibrosis and vascular obliteration in the skin, gastrointestinal tract, lungs, heart, and kidneys.[1–5] The sclerosis of the perioral tissues is considered to be the reason for limited opening.[5–9] The dysphagia resulting from fibrosis of the tongue and smooth muscles of esophagus also occurs.

The skin develops a diffuse, hard texture which is difficult to pinch and its surface is usually smooth, and mask like facies.[2,3,5] The nasal alae become atrophied, resulting in a pinched appearance to the nose, called a mouse facies.[2,3,6,7] Skin over the extremities, faces, and trunk may become darkly pigmented and contrasting areas of hypo pigmentation may also develop.

The oral manifestations include microstomia, xerostomia and telangietasia.[3–7,10] The tongue can become rigid, making speech and swallowing difficult. The soft tissues around the temporomandibular joints were also affected, which results in pseudoankylosis. The radiographic findings include: uniform widening of the periodontal ligament space, especially around the posterior teeth.[3,8,10–15] Also the mandible shows varying degree of bone resorption.[3–5,8–15]

Maintenance of oral hygiene may be difficult due to the patient's inability to grip and manipulate a tooth brush or other oral hygiene aids. This particular problem of reduced manual dexterity may be attributed to the maintenance of poor oral hygiene by the patient.

Radiographically, the striking feature of this disease is the widening of the periodontal ligament space.[3,7–15] Enlarged periodontal ligament space may be caused by excess deposition of collagen and oxytalan fibers and subsequent resorption of the alveolar crest bone surrounding the roots.[3,7–15]

Systemic sclerosis can give rise to many oral problems, most commonly reduced mouth opening. Mouth stretching exercises are probably the best treatment for microstomia and have been shown to improve mouth opening and help with speaking, eating and oral hygiene.[2,3,7,10] Good oral hygiene is essential for patients helping them to keep the mouth free of dental caries and periodontal diseases.

Conclusion

The mouth, oral cavity and teeth can be affected in systemic sclerosis leading to difficulties with speech, feeding and reduced quality of life.

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