Case Report

JRAD Journal of Research and Advancement in Dentistry

J Res Adv Dent 2018;8:2:271-275.

Systemic Sclerosis with Intraoral Manifestation: A Case Report

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ABSTRACT

Background: Scleroderma is an autoimmune connective tissue disorder which is characterized by fibrosis of visceral organs, skin and blood vessels. This condition can be localized or systemic. It generally affects woman between 30 and 50 years of age and has a low prevalence (250 cases in a million). Dermal manifestations are stiff, tight (decrease number of wrinkles), and shiny skin usually of the hands and feet due to swelling and thickening of the connective tissue. Other symptoms include difficulty in swallowing, bloating, abdominal pain, tiredness, lack of energy, weight loss, pain in muscles, joints, and bones. Lungs, heart, and kidneys are the vital organs that may get involved. Resorption of the mandibular angle and coronoid process can be observed in patients with scleroderma due to the pressure of fibrous mucocutaneous tissues. The aim of this case report is to present a 60-year-old female patient of systemic sclerosis with intraoral manifestations.

Keywords: Autoimmune disorder, systemic sclerosis, scleroderma.

INTRODUCTION

Systemic sclerosis (SSC), also known as scleroderma, is a multisystem, autoimmune disease, characterized by functional and structural abnormalities of small blood vessels, fibrosis of skin and internal organs, and production of autoantibodies. The word scleroderma comes from two Greek words, “sclero” meaning hard and “derma” meaning skin.[1]. Since hidebound skin is the clinical hallmark of the disease, it is also called “hidebound disease.”[2] Skin thickening is considered as a hallmark of it, which distinguishes it from other connective tissue disorders. These are classified into two subgroups as systemic and localized. Systemic sclerosis is associated with Raynaud’s phenomenon, acrosclerosis and internal organ involvement .[3,4,5] Although the etiology of scleroderma is not clear, genetic and environmental factors have been held responsible for the onset of this condition. There is an excessive deposition of normal type I and III collagen at various tissues caused by immunologically over-activated fibroblasts. [1]

Auto-antibodies and perivascular lymphocytic (mainly CD4 positive T lymphocytes) infiltrate indicate activation of immune system. The CD4 positive T cells are activated by laminine and type IV collagen which are endothelial basement membrane components . Consequently, these cells secrete an endothelial cytotoxic factor named granzyme as well as tumor necrosis factor which activates endothelial cells and transforming growth factor - β (TGF - β) which activates fibroblasts to express TGF - β and platelet-derived growth factor (PDGF). PDGF activates fibroblasts causing increased amounts of collagen secretion. The serological specificity of the disease is due to the presence of antinuclear antibodies (ANAs) which are directed mainly against cell nuclear enzymes, like DNA anti-topoisomerase-I (TOPO-I) and RNA polymerases, as well as centromeric proteins, anticentromere antibodies (ACA).[6] SSc has a worldwide distribution and affects all races. Factors

Received: June. 3, 2018: **Accepted**: July. 12, 2018
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pISSN 2278-0076
eISSN 2321-9270

such as age, sex, genetic background, and environmental exposure may influence its susceptibility.[7]

Oral and facial tissues are often affected, presenting very characteristic clinical picture. Most clinical manifestations begin with tongue rigidity and facial skin hardening, which gives it a classic mask-like appearance.[8] This is a case report of Systemic sclerosis with orofacial manifestations of the disease.

CASE REPORT

A 60 yr old female patient reported to the department of oral medicine and radiology with the chief complain of reduced mouth opening since 1years. On taking detailed history we came to know that she was a known patient of systemic sclerosis since 1 ½ yrs and was undergoing treatment for same. Patient was apparently alright 2yrs back when she complained of numbness and pain in extremities followed by pain in joints especially small joints and muscles, weakness and tightening of skin. Patient complains of shortness of breath and blurred speech since 1 year also decreased sweating and thinning of hair and cough. She consulted physician for same and after undergoing investigation like chest x-ray, CT Scan brain, blood investigations, endoscopy a diagnosis of systemic sclerosis was made. Since 1 year patient complains of difficulty in opening mouth which gradually reduced with no tenderness on TMJ region with no pain or difficulty in deglutition. She also complains of dry mouth. The patient was poorly built and nourished. Her vital signs were within the normal limits. Extra oral examination revealed perioral stiffening causing reduced mouth opening, thinning and furrowing of lips, incompetent lips, protruded anteriors ,(fig1) thinning of hair, generalized smooth shiny skin , reduced or loss of appendages, thickening of skin, loss of elasticity of skin, (fig 2) swollen hands with bulbous and rounded ends along with tightening of skin leading to inability to flex, (Fig 3,4) areas of hypopigmentation , loss of restricted jaw movements due to stiff skin. Intraoral examination showed restricted mouth opening (Fig 5) multiple root stumps and carious tooth, areas of hypopigmentation on buccal mucosa (Fig 6) and restricted movement of the tongue. Palpable bands were present on right and left buccal mucosa. Depapillation of tongue and xerosis was noted.

Panoramic radiograph showed generalized bone resorption widening of periodontal ligament space, multiple root stumps and carious involvement.

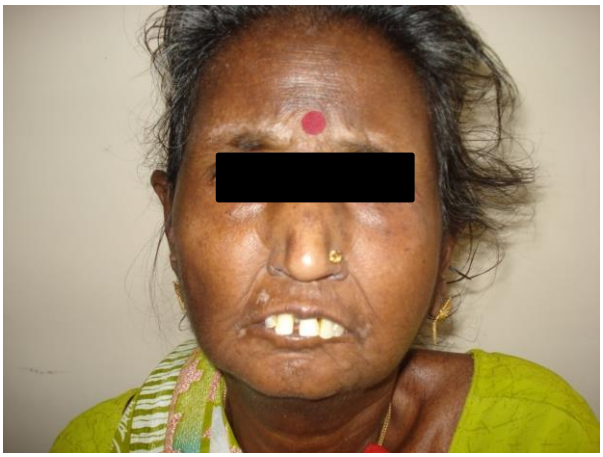

Fig 1: Perioral stiffening & incompetent lips

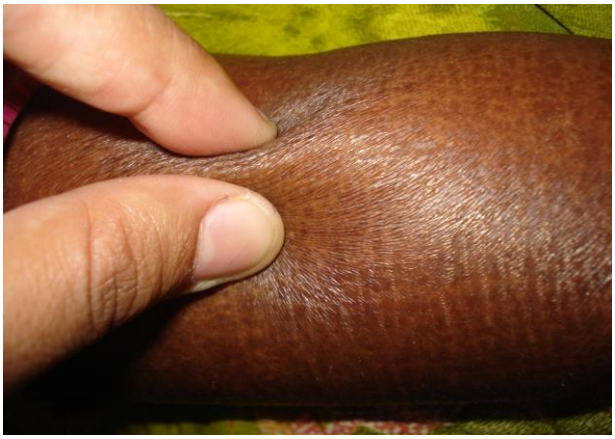

Fig 2: Shiny stretched skin (Hidebound)

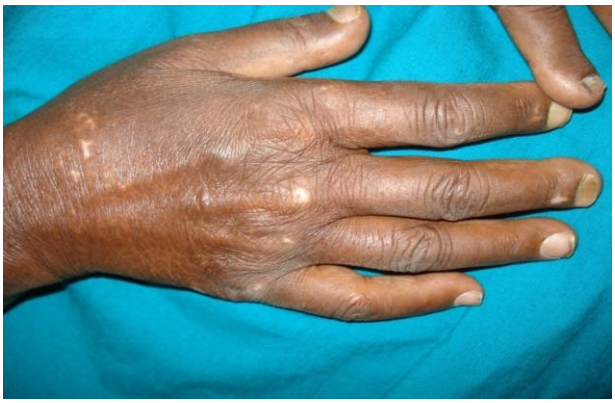

Fig 3 Areas of hypopigmentation

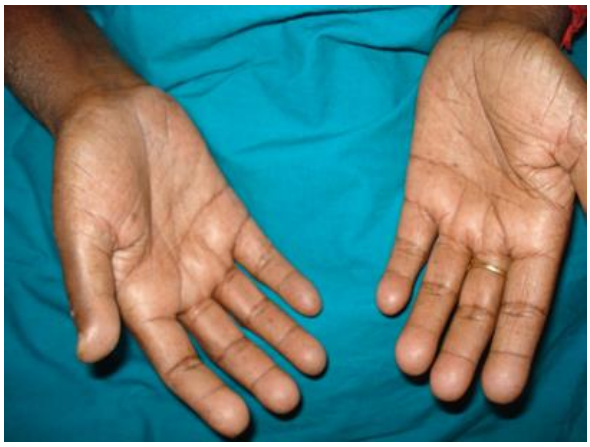

Fig 4 Swollen hands with bulbous and round end

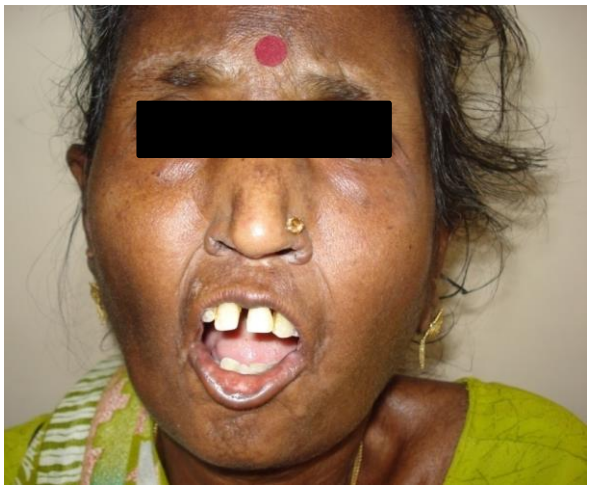

Fig 5 Restricted mouth opening (Fish mouth)

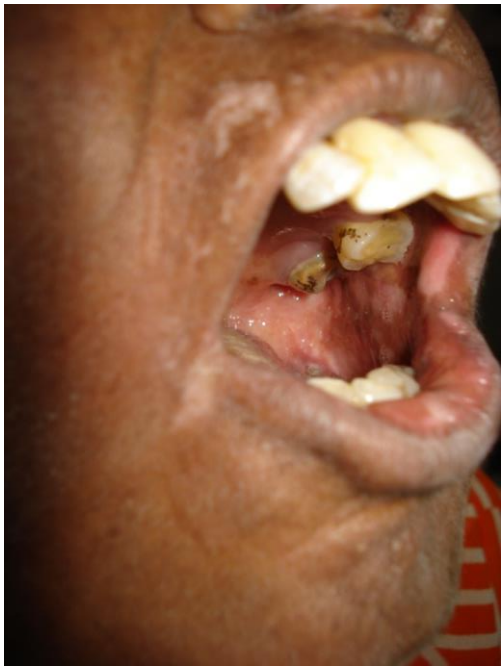

Fig 6 Areas of hypopigmentation

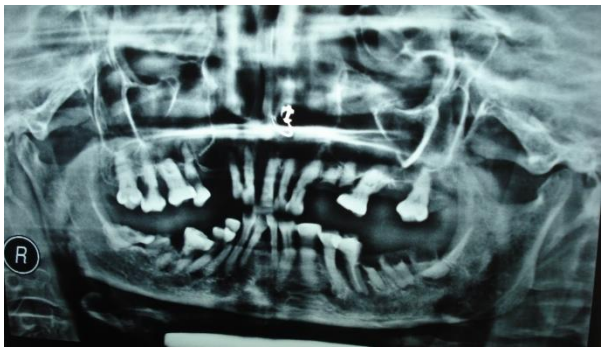

Fig 7 Generalized bone resorption & widening of periodontal ligament space Patient was advised jaw stretch exercise to improve the mouth opening.

DISCUSSION

Systemic Sclerosis is a rare multisystem disorder of connective tissue with an immunologically mediated pathogenesis characterized by extensive cutaneous and visceral fibrosis and small vessel vasculopathy, in which the antibodies are targeting blood vessels and connective tissues . [9] While the etiopathogenesis is unknown, pathological findings indicate that fibroblasts are activated to produce excessive amounts of collagen and other extracellular matrix components. Moreover, endothelial cell damage causing capillary loss and neo-intimal proliferation in small vessels plays a prominent role in tissue damage. Autoimmune mechanisms also are believed to be operative, because patients with Systemic Sclerosis spontaneously elaborate a variety of highly disease-specific circulating autoantibodies to nuclear and nucleolar antigens.

Collagen deposition in connective tissue leads to fibrosis and progressive limitation of mouth opening.[10] The skin develops a diffuse, hard texture, and its surface is usually smooth. Limited mouth opening is a frequent finding in patients with Systemic Sclerosis.[11]. Bilateral or unilateral commissurotomy has been described as a surgical means of correcting the limited mouth opening in Systemic Sclerosis patients.[12,13] Mouth stretching exercises and facial grimacing probably are the best non surgical treatment options.

Oral and radiographic examination reveals generalized enlargement of the periodontal ligament space which is caused by excess deposition of collagen and oxytalan fibres and subsequent

resorption of alveolar crest bone surrounding the roots.[14,15] .The oral mucosa may undergo changes in similar fashion as that found in the skin. Mild edema followed by gradual atrophy and induration of the mucosal tissues. Also there are areas of hypopigmentation.[14]

Due to severe flexion deformities of their fingers and other body joints and these pateints have reduced manual dexterity causing difficulty in the maintenance of oral hygiene. A larger-handled toothbrush or flossing aids may allow easier grasping and facilitate maintenance of good oral hygiene.[17]

Dental treatment is difficult in these patients due to restricted mouth opening and due to dry mouth they are more prone to dental caries. Taking good care of their oral health is essential for patients with systemic sclerosis helping them to keep the mouth free of dental caries and periodontal diseases. It is advised that patients with scleroderma should visit the dentist and dental hygienist at least once in three months to ensure the maintenance of good oral health care. Follow-up of these patients with periodic panoramic radiographs is essential to monitor and intercept potential consequences like pathological fractures and neuropathies. Dentists should be aware of the challenges associated with performing routine intraoral examination, periapical radiographs, and dental treatment for Systemic Sclerosis patients with limited mouth opening.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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