

Oral Complications of Systemic Sclerosis; A Case Report

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ABSTRACT

Objective: Systemic Sclerosis is a connective tissue disorder with multi-system involvement. It is an autoimmune disease with multiple aetiological factors including; genetic, birth disorder, infectious and environmental factors. Sclerosis of the skin and systemic fibrotic manifestations are the hallmark of this condition. We report a case of a 30-year-old female, who presented to the Periodontology clinic of the University of Benin, Teaching Hospital, Benin City with complaints of breakage of her teeth and previous multiple extractions. Clinical examination revealed multiple carious lesions, retained root, mobile teeth with varying degrees of mobility, tooth wear lesions and gingival recession.

Case Description: She also presented with extra-oral features which include sparse and fluffy hair, reduced lacrimation, progressively reducing mouth opening and thickening of skin of the forehead, cheeks, and extensor surfaces of the upper and lower limbs

Conclusion: A multidisciplinary management approach, involving the periodontologist, oral surgeons, oral physicians, prosthodontist, rheumatologists, chest physicians and dermatologists, was instituted.

Keywords: Systemic sclerosis, oral complication, case report

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INTRODUCTION

Systemic scleroderma also known as systemic sclerosis (SSc) is a multisystem autoimmune connective tissue disease (CTD) which, in its classical form, shows sclerosis of the skin as well as systemic involvement characterized by fibrosis, vascular (mostly microvascular) pathology and immunological abnormalities.¹ Fibrosis occurs in skin, the gastrointestinal (GI) tract, heart, lungs and other internal organs. Major organ involvement leads to decreased survival in SSc. Pulmonary fibrosis [interstitial lung disease (ILD)] and pulmonary arterial hypertension (PAH) cause more than half of all SSc-related deaths.²

Reported prevalence and incidence vary widely depending on geographic location. The prevalence varies from 30 cases/million in 1979 in New Zealand Wigley and Borman, 1980³ to 580 cases/million in Alberta, Canada between 1994 and 2007.⁴ Annual Incidence rate is reported to vary from 46 cases/million in the United States between 2003 and 2008⁵ (t and 1.96 cases/million from 1950 to 1973 in New Zealand (Eason RJ. 1981)⁶.

Mayes et al, (2003)⁷ reported higher prevalence in African- Americans compared to Americans of European decent. Many authors have reported a female predilection. A prevalence study done in a larger US population reported a female to male ratio of 4.6: 1⁸. A female predilection as high as 12:1 has

also been documented (Richard MK, 2018)⁹. Systemic sclerosis is rare in children and seen more in adults within 5th to 8th decades of life.⁸

The aetiology is largely unknown. However, genetic, gender, age, race, birth order, infectious and environmental factors all appear to play a role.¹⁰ Factors which have been implicated include; genetic predisposition and infectious agents, including cytomegalovirus. Parvovirus B19, *Helicobacter pylori*, hepatitis B virus, Epstein-Barr virus, *Toxoplasma gondii* and chlamydia have been implicated as possible triggers. Chemicals (such as polyvinyl trichloroethylene, some pesticides, organic solvents, hair dyes and silica) as well as drugs such as cocaine, pentazocine, bleomycin, penicillamine and vitamin K, have also been implicated. Other factors include radiation therapy, physical trauma and vitamin D deficiency.¹¹

Clinically, two types of systemic sclerosis are recognized. The two types, limited and diffuse cutaneous scleroderma, are differentiated by the extent of skin affected.¹² The limited disease is defined as skin thickening that only affects the extremities below the elbows and/or below the knees. Diffuse cutaneous disease is defined as skin thickening proximal to the elbows/or knees in addition to distal extremity involvement¹³). Truncal skin may also be involved in diffuse cutaneous systemic sclerosis. The face can be involved in both forms and has no bearing on subset designation. The clinical manifestations of SSc may be considered the result of these pathological processes, small vessel non-inflammatory obliterative vasculopathy, fibrosis of the skin and other organs and autoimmunity.¹⁴

The obliterative small vessel vasculopathy is responsible for Raynaud's phenomenon, scleroderma renal crisis, and pulmonary artery hypertension.¹⁵ Other pathological events in SSc may include impaired communication between endothelial cells, epithelial cells and fibroblasts; lymphocyte activation; autoantibody production; inflammation; and connective tissue fibrosis. These events result in an accumulation of constituents of the extracellular matrix, which replaces the normal tissue architecture, which in turn can culminate in organ failure.¹⁶

The orofacial manifestations of scleroderma include fibrosis and rigidity of facial skin (mask-like appearance of face), tongue, soft palate, larynx, salivary glands, and buccal mucous membrane leading to microstomia.¹⁷

Patients also complain of xerostomia, and frequently present with complications of prolonged dry mouth, typically multiple dental caries, oral thrush and halitosis. The tongue can also become fibrotic and rigid, making speech and swallowing difficult. The tongue becomes depapillated and there could be presence of ulcers, erosions and telangiectasia in and around the oral cavity. There is blanching of the buccal mucosa and ventral surface of the tongue.¹⁸

Periodontal manifestations are not uncommon. There is loss of attached gingiva and gingival recession may also occur. The radiographic findings include; uniform widening of the periodontal ligament space, especially around the posterior teeth. This is caused by increase in the collagen synthesis in the periodontal ligament, which is the hallmark of this disease. Collagen synthesis causes an increase in the thickness and space occupied by the periodontal ligament. Patients with systemic sclerosis have been reported to have increased susceptibility to chronic periodontitis. Marmar¹⁹, observed a significantly increased incidence of periodontal disease in patients with systemic sclerosis as compared to controls.

Temporomandibular joint can also be affected in SSc. Pseudoankylosis and osteolysis of the mandibular angle and condyles have been reported.²⁰ (Taveras, 1959). Resorption of the condyle, zygomatic arch, angle, and coronoid process were also reported in another report.²¹ Mechanism of this osteolysis is unknown. However, it has been explained that vasculopathy associated with this disease may diminish the blood supply to the mandible resulting in bone ischemia and necrosis.²²

CASE REPORT OF SYSTEMIC SCLEROSIS

A 30-year-old female patient presented to the Periodontology Clinic of University of Benin Teaching Hospital, Benin City Edo State, with a complaint of her teeth chipping off for over two years. There was associated pain and difficulty in mastication which had warranted several extractions. Her medical history revealed stiffness and shortening of the fingers and toes bilaterally with associated fibrosis of the skin and the forearm and the legs bilaterally. Patient had been diagnosed by the physicians with systemic sclerosis with interstitial fibrosis of the lungs in 2011 and was being managed using local and systemic drugs, notably steroids and immune-modulating drugs for the lung fibrosis

Past Dental history revealed that Patient has had several extractions (6) on account of tooth decay and mobile teeth. Patient was unable to carry out mechanical plaque control optimally because of her shortened and stiff fingers.

On general examination, patient appeared small for age, hair was fluffy and sparse, face was symmetrical but the skin over the cheeks and forehead had a leathery feel. The fingers and toes were rigid, shortened with marked reduction in range of mobility. The skin over the extensor surfaces of the arms, forearms, thighs and legs were fibrotic.

Intraoral examination revealed diminished mouth opening (3.5cm). Oral hygiene was poor. The saliva

on the oral mucosa was chord-like and saliva pool in the floor of the mouth was absent halitosis was perceived, mucosa lining was generally pale, blanched and fibrotic. There was mild ankyloglossia. There were carious lesions on 33, 32, 31, with retained root of 21. The mobile teeth present were; 41 (Miller's grade II), 42 and 43 (Miller's grade I). Notably was, gingival recession on several teeth; 11(miller's class II), 12(miller's class II), 13(miller's class II), 14(miller's class III), 41(miller's class III), 42(miller's class IV), and 43(miller's class II). Teeth 24, 25, 26 and 35 were abraded.



The following diagnoses were made; generalized chronic periodontitis, dental caries of 33, 32, 31, and Retained root of 21 in a background of systemic sclerosis. Comprehensive dental treatments commenced with scaling and polishing by the Periodontologist, thereafter extractions of the grossly carious teeth and retained roots were carried out by the oral surgeons. Prosthetic rehabilitation with removable partial denture has been carried out. Immediate assessment and six (6) months recall were satisfactory.

DISCUSSION

Systemic sclerosis is an autoimmune disease that is characterized by the distinctive pathogenetic triad of microvascular damage, dysregulation of innate and adaptive immunity, and generalized fibrosis in multiple organs. Although skin fibrosis is the distinguishing hallmark, the pathological changes in the lungs, gastro-intestinal tract, kidneys and heart determine the clinical outcome. In general, the extent of skin involvement and its rate of progression reflect the severity of visceral organ complications.^{23,24}

The microrstomia and mask-like appearance facial appearance are mainly caused by skin and muscular atrophy. Fibrosis of salivary glands led to hyposalivation and complaints of Xerostomia. Chronic periodontitis and rampant caries were consequent manifestations of hyposalivation.²⁵⁻²⁸

There is no consensus management protocol adopted locally for diagnosis and multidisciplinary management of systemic sclerosis. This patient's first presentation to the dental clinic was six years after the initial diagnosis by the physicians and referral was made on patient's immediate dental complaints. The overall treatment outcome and quality of life of the patient is markedly improved with the contributory inputs from the dental specialities. The multidisciplinary management protocol is hence strongly recommended for patients with systemic sclerosis and other connective tissue disorders.

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