Acta Scientific Dental Sciences (ISSN: 2581-4893)

Volume 6 Issue 7 July 2022

Case Report

Systemic Sclerosis: Wrinkle Free Mask

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DOI: 10.31080/ASDS.2022.06.1417

Received: June 20, 2022

Published: June 30, 2022

Abstract

Scleroderma is a chronic autoimmune inflammatory disease wherein the blood vessels and connective tissue is the target for the antibodies. Systemic sclerosis can range from mild to severe form in severity as well as in progression from localized to systemic sclerosis with/without diffuse scleroderma. The disease process has a wide range of clinical manifestations ranging from the CREST syndrome accompanied with restrictive pulmonary disease, pulmonary hypertension, arthralgia, myopathy, myocardiopathy and progressive renal insufficiency. Herein, we report a classic case of systemic sclerosis with oro-cutaneous manifestations which has progressed over time and its impact on the life of the individual.

Keywords: Sclerosis; Calcification; Collagen; Chronic; Inflammation; Skin; Xerostomia

Introduction

Scleroderma is derived from the greek words skleros meaning hard and derma representing skin which may be systemic as well localized in form. However, the term systemic sclerosis is rendered only after the systemic presentation of the disease process [1].

Systemic sclerosis is defined as a chronic inflammatory disease of unknown origin and autoimmune nature which shows excessive collagen and glycosaminoglycan's deposition in the connective tissue of the dermis as well as the internal organs [2,3].

They have a definite female predilection, especially in the third and fifth decade of life million [1,2,5]. The etiologic cause of the disease process remains a mystery till date with the previously hypothesized genetic, autoimmune, nervous system factors being ruled out and the focus on vascular and environmental factors being the current causative factors [5,6].

Systemic sclerosis can be easily differentiated from its localized form in the presence of Raynaud's phenomenon, acrosclerosis and internal organ involvement [1-3]. The most commonly deposited excess collagen types are I and III, because of an immunological

activity, in response to which there is excessive activation of fibroblasts [7].

Herein, we report a classic case of systemic sclerosis with orocutaneous manifestations which has progressed over time and its impact on the life of the individual.

Case Report

A 45-year-old female patient reported to the oral medicine and radiology department presented with chief complaint of pain in the lower right back tooth region since a week sudden, severe, aggravating on taking food, radiating to the floor of the mouth.

The patient revealed a history of diagnosed systemic sclerosis for a period of 20 years but was not undergoing any treatment or supportive care for the same. No other significant medical history was revealed.

Her skin biopsy report showed that the dermis had mild perivascular lymphocytic infiltration with normal collagen. The ANA test was found to be positive, suggestive of collagen vascular disease.

The barium swallow test revealed faucity of peristalsis with barium emptying only in erect posture, suggestive of motility disorder consistent with systemic sclerosis.

On extra-oral examination the patient looked pale and anaemic with taut and tense skin which was also smooth and glisteningly shiny giving it a stoned, mask like appearance, without any wrinkles or the ability to change facial expressions. The short atrophied pinched nose made it mimics a “mouse facies”.

Further, the lack of any flexibility of the skin of the face made it difficult for the patient as the maxillary anterior teeth had become evident because of lip thinning (leading to incompetency). Temporo-mandibular joint was stiff on palpation and the patient had decreased mouth opening of only 27 mm.

The patient also could not completely close her fingers to a fist because of the tautness of the skin.

On intra-oral examination, the oral mucosa was pale and blanched across the labial and the buccal mucosa with a shrunken uvula. The tongue was rigid with restricted movements. The mouth mirror was sticky against the mucosal surfaces, suggestive of decreased salivation.

Generalized inflammation of marginal gingiva was seen. Deep caries w.r.t 36 extending to the pulp with tenderness on percussion, decay was noted in 17. Maxillary and mandibular teeth showed grade I mobility.

Orthopantomogram (OPG) showed generalized PDL space widening, especially in the mandibular posterior teeth, along with interdental bone loss in maxillary and mandibular anterior as well as posterior teeth. Further, there was loss of angle of mandible bilaterally seen.

A final diagnosis of systemic sclerosis along with generalized periodontitis and deep dental caries w.r.t 36 was rendered.

The patient was given oral hygiene instructions followed by thorough scaling and root planning. The patient was treated with antibiotics and analgesics and was advised root canal treatment w.r.t 36 and restoration w.r.t 17. Further, mouth stretching exercises were communicated to the patient to improve the mouth opening.

Discussion

Scleroderma is an autoimmune disease wherein the blood vessels and connective tissue is the target for the antibodies [8].

Systemic sclerosis can range from mild to severe form in severity as well as in progression from localized to systemic sclerosis with/without diffuse scleroderma inhibiting/reducing the skin movement of the phalanges and on the face (stone-faced). Latter form is termed as limited cutaneous scleroderma or CREST syndrome (calcinosis cutis, Raynaud’s phenomenon, oesophageal dysfunction, sclerodactyly, and telangiectasia) [9].

The disease manifests in three forms

- **Morphea**: (Circumscribed scleroderma) Characterized by local thickening of the skin
- **Generalized or progressive scleroderma**: (Diffuse form) Characterized by tautness of the skin with distinctive involvement of the lungs, heart, kidneys, and gastrointestinal tract, as well as osteolytic changes of the skeleton
- **Acrosclerosis**: Amalgamation of scleroderma of the extremities and Raynaud’s disease is noted [10].

Crest Syndrome [11,12]

Crest Syndrome [11,12]	
C	Calcinosis cutis
R	Raynaud's phenomenon
E	Esophageal dysmotility with dysphagia
S	Sclerodactly
T	Telangiectases

Table 1

The disease process has a wide range of clinical manifestations ranging from the CREST syndrome complemented with restrictive pulmonary disease, pulmonary hypertension, arthralgia, myopathy, myocardiopathy and progressive renal insufficiency [1-3].

Oral and cutaneous manifestations of systemic sclerosis [12,13]

Oral and cutaneous manifestations of systemic sclerosis	
Cutaneous manifestations [12]	**Oral manifestations [13-15]**
Thickening of skin	Limited ability to open the mouth
Pitting edema	Xerostomia
Tightening and	Periodontal disease.
Hardening of skin	Increased periodontal ligament (PDL) width Osseous resorption of the mandible & mandibular angle
Thin and taut facial skin	Increased frequency of dental caries
Decrease in facial wrinkles	Dysphagia

Table 2

Our patient showed all the oral as well as cutaneous manifestations of the disease process and as it was a diagnosed case, it became more evident as to the progression of the disease process.

Radiographic observations includes widening of the periodontal space, mandibular resorption, "tail of the whale" appearance which represents the blunting of mandibular angles [1,5,16,17].

Radiological impressions of the oral cavity show an increase in the width of the PDL around the anterior as well as posterior teeth, with a more definite widening observed in the posterior teeth. However, the teeth remain immobile as there are no changes in the lamina dura or the gingival attachments [16-18].

We found generalized PDL space widening, especially in the mandibular posterior teeth, along with interdental bone loss in the Orthopantomogram.

Clinical manifestations and investigations seen across different systems involved in the body [19-21].

We did not carry out any investigations to identify the stage of the disease process for each system and only limited it to the oral cavity, however the changes observed in the physical examination of the disease process, made it evident that it was systemic sclerosis.

Diagnosis

The diagnosis of systemic sclerosis is based on the identification of features that distinguish it from other disease, and thus a detailed history and careful physical examination are required.

The American College of Rheumatology has proposed criteria to assist in identifying those affected with the condition [21]

- **Major criteria**: scleroderma proximal to the metacarpophalangeal joints
- **Minor criteria:** sclerodactyly, digital pitting scars, and bibasilar pulmonary fibrosis

To fulfill a diagnosis of systemic sclerosis, either 1 major or 2 minor criteria are needed

In about 90% of cases, antinuclear antibodies are seen.10 Anticentromere antibodies are more likely to be associated with limited systemic sclerosis, whereas autoantibodies to topoisomerase-I (anti-Scl-70) are more likely to be associated with diffuse systemic sclerosis [22].

S.I. No	System involved	Clinical manifestations/complications	Investigations
1	G.I tract	Heartburn, dysphagia or respiratory complaints eg. Dyspnea, Barrett's esophagus	Endoscopy
2	Pulmonary system	Interstitial lung disease, pulmonary hypertension, pleuritis and pleural effusion, and aspiration pneumonia.	Chest radiography, pulmonary function testing, or HRCT of the chest
3	Cardiac system	Pericarditis with effusion, constrictive pericardium, arrhythmias, and congestive heart failure	Electrocardiogram, echocardiogram, a stress test, or cardiac catheterization
4	Renal system	Progressive renal failure, normal or mildly elevated blood pressure to malignant hypertension, elevated plasma renin levels, elevated serum creatinine, proteinuria, micro-angiopathic hemolytic anemia	Serial echocardiograms, right heart catheterization

Table 3

More commonly, anaemia of chronic disease or iron deficiency anaemia is seen; the latter is associated with chronic bleeding in the gut from esophagitis or watermelon stomach or other telangiectasia.

There have been various management protocols for systemic sclerosis, but none of them are standardized or conclusive and are based on the organs affected i.e target specific, rather than generalized. Use of corticosteroids has no effect on the status of the progression of the disease process, whereas the use of pencillamine is advocated as it inhibits collagen deposition [5,6,14].

Localized skin lesions respond well to ultraviolet light therapy by decreasing the skin tautness. Other supportive measures for skin lesions are high-dose topical corticosteroids, topical antibiotic ointments, and oral analgesics. Treatment with oral cyclophosphamide for a period of 1 year which showed modest improvement in lung function, functional status, and health-related quality of life [23].

A recent study has shown that patients with scleroderma treated with 1 year of oral cyclophosphamide had modest improvement in lung function, reduced dyspnea and functional status of the organs [24].

Figure 1

Figure 2

Figure 3

Conclusion

Systemic sclerosis is a chronic disease process which significantly impacts the life of the individual with the progression of the disease process. Our patient was a classic case of systemic sclerosis that showed all the signs and symptoms of the disease process which had progressed to the later stage. With the limited options to treat and manage the disease process, it is difficult to provide a better quality of time for the individual. However, it becomes important for the physician to provide the maximum supportive care for the affected individual.

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