Periodontal Manifestations in Systemic Sclerosis

A Review

Authored by Anshul Mehra, BDS, MDS and Suneet Kumar, MBBS, MD

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**Periodontal Manifestations in Systemic Sclerosis**

**A Review**

**LEARNING OBJECTIVES:**

After reading this article, the individual will learn:

- the etiology, pathogenesis, clinical features, oral manifestations, and treatment of scleroderma, and
- the periodontal manifestations seen in patients with systemic sclerosis.

**ABOUT THE AUTHORS**

**Dr. Mehra** is senior lecturer, Department of Oral Medicine and Radiology, U.P. Dental College and Research Centre, Lucknow, Uttar Pradesh, India. He can be reached at mehraanshul@yahoo.co.in.

**Dr. Kumar** is assistant professor, Department of Pathology, Kasturba Medical College, Mangalore, Karnataka, India. He can be reached at suneet302@yahoo.com.

**INTRODUCTION**

Scleroderma is a connective tissue disorder characterized by tissue fibrosis, obliterator microangiopathy, and immune abnormalities. The term scleroderma is derived from the Greek word scleros (hard) and derma (skin), hence the meaning hard skin. This term describes the pathognomonic clinical appearance of the skin seen in this disease. The exact etiology and pathogenesis of this disease are not clear. Most commonly implicated etiological factors are environmental in nature (silica dust, vinyl chloride, benzene, and tryptophan) or of viral etiology (cytomegalovirus, human parvovirus B19). The most important predisposing factors associated with this disease are B cell abnormalities and genetic susceptibility1,2 (Figure 1).

**CLASSIFICATION**

Scleroderma is classified as localized scleroderma and systemic sclerosis (Figure 2). Three types of localized scleroderma are recognized: morphea, generalized morphea, and linear scleroderma (en coup de sabre). Morphea is characterized by circumscribed sclerotic plaques on the skin with ivory-colored centers and violaceous borders that eventually lose hair and the ability to sweat (Figure 3). The plaques are indurated but not...
bound to the deeper structures. Most commonly, the lesions are single or few in number, but they may be multiple. In generalized morphea, there is widespread involvement of skin with multiple indurated plaques and hyperpigmentation. Linear scleroderma is characterized by a thin band of sclerosis that may run the entire length of an extremity, and involves superficial and deeper layers of the skin, with fixation to underlying structures. The lower extremities are most often involved, followed by the upper extremities, frontal areas of the head, and anterior thorax. The lesion of linear scleroderma of the head and face is called en coup de sabre, and these lesions may result in hemiatrophy of the face.

Two types of systemic sclerosis are recognized, defined by the extent of skin affected: limited cutaneous scleroderma and diffuse cutaneous scleroderma. In limited cutaneous scleroderma, there is skin thickening in the areas solely distal to the elbows and knees, with or without facial effects. It is generally a milder form of disease, and the patients frequently have problems with digital ulcers and esophageal dysmotility. Diffuse cutaneous scleroderma has a more acute onset and is defined by the presence of skin thickening that is proximal, as well as distal, to the elbows and knees, with or without facial or truncal effects. There is widespread internal organ involvement with potentially life-threatening cardiac and renal problems.¹⁻⁶ (Figure 4).

**CLINICAL FEATURES, TREATMENT AND PROGNOSIS**

Systemic sclerosis occurs mainly in the third to fifth decade of life. Women are affected 3 to 4 times as often as men. The initial symptoms of systemic sclerosis are non-specific and commonly include Raynaud’s phenomenon (Figure 5); swelling or puffiness of the skin (usually on the fingers, hands, and face) that is replaced by thickening and tightening of the skin (Figure 6); ulcerations and gangrene of the fingers (Figures 7 and 8); migratory polyarthritis, flexion contractures, and sclerodactyly with digital tuft resorption, subcutaneous calcifications, joint space narrowing, and focal erosions of periarticular bone on radiographs (Figure 9); and less commonly, gastrointestinal problems such as gastroesophageal reflux, dysphagia, and constipation. The clinical course of systemic sclerosis

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**LIMITED CUTANEOUS SSC (ISSC)**
- Previously known as CREST syndrome - Calcinosis cutis, Raynaud’s phenomenon, Esophageal dysmotility, Sclerodactyly, Telangiectasia
- Raynaud’s phenomenon for years (occasionally decades)
- Skin involvement limited to hands, face, feet, and forearms (acral)
- A significant (10-15%) late incidence of pulmonary hypertension, with or without interstitial lung disease, skin calcification, telangiectasia, and gastrointestinal involvement
- High prevalence of anticentomere antibody (70-80%)

**DIFFUSE CUTANEOUS SSC (dSSC)**
- Onset of skin changes (puffy or hidebound) within 1 year of onset of Raynaud’s phenomenon
- Truncal and acral skin involvement
- Tendon friction rubs
- Early and significant interstitial lung disease, renal failure
- Diffuse gastrointestinal disease
- Myocardial involvement
- Antitopoisomerase-1 (Scl-70) antibodies (30% of patients)
varies and depends upon whether the patient develops limited or diffuse cutaneous systemic sclerosis (Figure 4). Scleroderma does not have a cure, and treatment is focused on relief of symptoms (Figure 10). The lifespan of the patient with systemic sclerosis depends upon the rapidity, severity, and extent of internal organ involvement and the age of the patient at the time of presentation.1,2,4-6

**ORAL MANIFESTATIONS**

The oral and perioral tissues are also commonly involved. Most common findings include rigid lips, narrow oral aperture, loss of skin folds around the mouth (mask-like appearance), blanching of the oral mucosa due to fibrosis (Figures 11 and 12), sclerosis of the tongue (Figure 13), oral telangiectasia, and pseudoankylosis.7-9 Oral and perioral effects are mainly caused by skin and muscular atrophy seen in these patients. Xerostomia is also commonly seen in patients with systemic sclerosis and is either caused by glandular fibrosis or is found to be associated with Sjögren’s syndrome.1,4,7-11 Resorption of the mandible in areas of masticatory muscle attachments (angle, condyle, coronoid, and digastric) has also been reported.10,12-14

**PERIODONTAL MANIFESTATIONS**

The most common radiographic finding in patients with systemic sclerosis is widening of the periodontal ligament space (Figure 14). This finding was first reported by Stafne and Austin15 and was later confirmed by others.7-9,16-19 The exact mechanism for increase in the width of the periodontal ligament is not entirely clear. The most likely explanation seems to be an increase in the collagen synthesis in the periodontal ligament, which is the hallmark of this disease. Increased collagen synthesis causes an increase in the thickness and space occupied by the periodontal ligament. Microscopic examination also reveals excessive collagen deposition. This increase in the volume of the periodontal ligament exerts additional pressure on both tooth and alveolar bone. Wood and Lee19 did not observe any difference in the widths of the roots of the involved teeth between patients with systemic sclerosis and controls. Further, cementum is more resistant to resorption

**Figure 6.** Absence of wrinkles on the forehead when looking upward because of tightening of the skin.

**Figure 7.** Swelling and puffiness of the nailbeds.

**Figure 8.** Gangrene of the finger in systemic sclerosis.

**Figure 9.** Hand and wrist radiograph showing digital tuft resorption and subcutaneous calcifications.
than bone, as bone is richly vascularized and cementum is avascular. Therefore, the degenerative changes caused by altered blood flow, which is related to an increase in the pressure exerted by the periodontal ligament, affect bone more readily than cementum. Thus this increased volume of periodontal ligament is accommodated at the expense of alveolar bone without affecting tooth mobility.

The extent of periodontal ligament widening was found to be related to the severity of the disease. Recently, trauma from occlusion has also been reported as one of the probable causes for periodontal ligament space widening. It appears to be an unlikely cause, as the muscles of mastication undergo atrophy and thus mastication forces are reduced, thereby reducing the chances of trauma from occlusion.

Patients with systemic sclerosis have been reported to have increased susceptibility to periodontal disease; however, studies have shown conflicting results. Marmary, et al observed a significantly increased incidence of periodontal disease in patients with systemic sclerosis as compared to controls, whereas Nagy, et al and Eversole, et al did not find any significant difference between cases and controls. These differences in the findings were attributed to the differences in the control groups. The increased susceptibility to periodontal diseases can be explained by both pressure ischemia and obliterative vasculopathy, which causes atrophy of the periodontal ligament and, at a later stage, mobility of the involved teeth. Recent evidence also suggests that the increased prevalence of xerostomia in these patients may be one reason for an increased prevalence of periodontitis.

Although the mean periodontal ligament space width was higher in all the teeth of patients with systemic sclerosis, the increase in the widths were found to be more pronounced in the posterior teeth than in the anterior teeth. This can be explained by the fact that the teeth bearing more masticatory force (i.e., posterior teeth) have wider

### Table: Symptomatic Treatment for Scleroderma

<table>
<thead>
<tr>
<th>SYMPTOMS</th>
<th>TREATMENT</th>
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<tr>
<td>RAYNAUD'S PHENOMENON</td>
<td>Avoidance of cold exposure and smoking cessation, warm clothing, calcium channel blockers, aspirin and dipyridamole, Pentoxifylline, Fluoxetine, Topical nitroglycerine paste over digits, intravenous Alprostadil and prostaglandin</td>
</tr>
<tr>
<td>SKIN FIBROSIS</td>
<td>d-Penicillamine, Colchicine, Paraaminobenzoate, Gamma interferon, Cyclosporine, Extracorporeal phototherapy</td>
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<td>GI SYMPTOMS</td>
<td>Antacids, H2-blockers, proton-pump inhibitors, Prokinetic agents. Reflux precautions: frequent small meals, elevation of head at end of the bed, not lying down after meals, avoidance of tea, coffee, alcohol, spicy and fatty foods</td>
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<tr>
<td>PULMONARY DISEASE</td>
<td>Systemic steroid and cyclophosphamide for pulmonary fibrosis; intravenous or aerosolized prostacyclin for severe pulmonary hypertension</td>
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<td>RENAL DISEASE</td>
<td>Early management of hypertension with calcium channel blocker Management of renal failure, dialysis</td>
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<tr>
<td>CARDIOVASCULAR DISEASE</td>
<td>Treatment of heart failure Pericarditis may respond to systemic steroids</td>
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<tr>
<td>MUSCULOSKELETAL SYMPTOMS</td>
<td>NSAIDs and paracetamol for arthralgia</td>
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<tr>
<td>INFECTIONS</td>
<td>Proper antibiotic therapy should be instituted</td>
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Figure 10. Symptomatic treatment for scleroderma.

Figure 11. Blanching of the oral mucosa due to fibrosis.

Figure 12. Blanching of the ventral surface of the tongue with thickening of the lingual frenum.

Figure 13. Depapillation of the tongue due to fibrosis.
Periodontal ligament spaces than those bearing less force (ie, anterior teeth). Therefore, the increase in the bulk of the periodontal ligament in patients with systemic sclerosis is reflected more in posterior teeth as compared to the anterior teeth.

As noted, the widening of periodontal ligament space was typically seen in posterior teeth. Also, the incidence of periodontal ligament space widening was found to be variable, ranging from 10% to 37%. However, when the periodontal ligament space was meticulously measured under magnification and the mean periodontal ligament width was calculated, it was found that patients with systemic sclerosis had a higher mean width as compared to controls, and the mandibular lateral incisor was found to be the best tooth for predicting patients with systemic sclerosis.

CONCLUSION

Patients with systemic sclerosis exhibit a generalized pattern of widening of the periodontal ligament space, with the average periodontal ligament width being greater around the posterior teeth as compared to the anterior teeth. This finding is of limited importance clinically, as the affected teeth are not mobile initially. Also, the significance of this finding in the diagnosis of systemic sclerosis is questionable because prominent widening tends to occur in the later stages of the disease.
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REFERENCES


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POST EXAMINATION QUESTIONS

3. En coup de sabre is the linear scleroderma involving the ____.  
   a. upper extremities  
   b. lower extremities  
   c. thorax  
   d. head and face

4. Raynaud’s phenomenon is ____.  
   a. an abnormal spasm of blood vessels  
   b. an abnormal spasm of muscles  
   c. damage of the nerves due to fibrosis  
   d. none of the above

5. The sequence of change in skin color seen in Raynaud’s phenomenon is ____.  
   a. white, blue, red  
   b. red, blue, white  
   c. white, blue, pink  
   d. blue, white, red

6. The characteristic appearance of the face seen in patients with scleroderma is ____.  
   a. bird-like appearance  
   b. fish-like appearance  
   c. mask-like appearance  
   d. simian-like appearance

7. The most common periodontal finding seen in patients with scleroderma is ____.  
   a. widening of periodontal ligament space involving the entire root, with mobility of the tooth  
   b. widening of periodontal ligament space involving the entire root, without mobility of tooth  
   c. widening of periodontal ligament space involving the apex only  
   d. generalized loss of lamina dura

8. Widening of periodontal ligament space occurs at the expense of bone and not the tooth because ____.  
   a. cementum is avascular  
   b. cementum is more resistant to resorption than bone  
   c. bone is richly vascularized, so the degenerative changes caused by interference in the blood circulation affect bone more easily than cementum  
   d. all of the above
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