# Maxillofacial Manifestations of Systemic Sclerosis: Case Report

## Maksillofasiyal Belirtileriyle Sistemik Sklerozis: Olgu Sunumu

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## ABSTRACT

Systemic Sclerosis is a rarely seen autoimmune inflammatory disease. The main sign of systemic sclerosis is a combination of thickened and stretched skin. Alongside the symptoms in the skin, inflammation and scar formation are observed in many parts of the body such as lungs, kidneys, heart, the intestinal tract. Systemic sclerosis is a disorder affecting oral and facial tissues with skin hardening, thin lips, deep wrinkles, xerostomia, tongue rigidity, and microstomia. The oral manifestations of the illness are caused by deposits of collagen in the tissues or as a result of collagen deposition around nerves and vessels, and all the oral tissues are affected. The most commonly known oral manifestation is widened periodontal ligament spaces. The aim of this study was to present and discuss the oral findings of a 46-year-old male patient with systemic sclerosis. Oral and facial examinations were performed clinically and radiologically. Results: In this study, the oral symptoms of the patient were dysphagia, periodontal problems and mouth opening limitation. SSc patients should have regular follow-up with a yearly panoramic radiograph for other oral and maxillofacial manifestations.

**Key Words:** Systemic sclerosis, Oral manifestations, Radiogram

## ÖZ

Sistemik sklerozis nadir görülen otoimmün inflamatuvar bir hastalıktır. Sistemik skleroziste ana bulgu cildin kalınlaşması ve gerilmiş olmasıdır. Deri bulgularının yanı sıra akciğerler, böbrekler, kalp ve sindirim sistemi gibi vücudun birçok yerinde inflamasyon ve skar gelişimi izlenir. Bununla birlikte sistemik skleroz; ciltte sertleşme, ince çizgiler, derin kırışıklıklar, kserostomi, dilde sertlik ve mikrostomi sebebiyle oral ve fasiyal dokuları da ilgilendiren bir hastalıktır. Hastalığın oral bulguları, dokulardaki veya sinir ve damarlar etrafındaki kollajen birikiminden kaynaklanır ve tüm oral dokular etkilenir. En sık görülen oral bulgu, genişlemiş periodontal ligament alanlarıdır. Çalışmanın amacı sistemik sklerozu olan 46 yaşında erkek hastanın oral bulgularını sunmak ve tartışmaktır. Klinik ve radyolojik olarak oral ve fasiyal muayene yapıldı. Bu çalışmada yer alan hastanın oral semptomları; disfaji, periodontal problemler ve sınırlı ağız açıklığı idi. Sistemik sklerozis hastaları, diğer oral ve maksillofasiyal bulgular için yıllık panoramik radyografilerle düzenli izlem yaptırmalıdırlar.

**Anahtar Sözcükler:** Sistemik sklerozis, Oral belirtiler, Radyogram

## INTRODUCTION

Systemic Sclerosis (SSc) is a chronic autoimmune inflammatory connective tissue disorder. It causes progressive fibrosis of the skin and sometimes also of internal organs because of an increased deposition of collagen (1-3). Although rgw pathogenesis of the disease is not known, it is thought that vascular abnormalities that might be caused by genetic, immunological and environmental factors have an important role in the pathogenesis (1,2,4). There are two types as limited cutaneous SSc and diffuse cutaneous SSc according to the

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extent of skin fibrosis and the pattern of organ system involvement (1,2,4,5). SSc is more frequent in women than men and the peak onset is between 30 and 65 years (2,4,5).

Pulmonary, renal, gastrointestinal, musculoskeletal, cardiac and endocrine system and vascular involvement (especially Raynaud's phenomenon) are systemic manifestations (4). It may present with clinical features such as Raynaud phenomenon, which is the first sign of the disease, skin thickening, esophageal dysmotility, pulmonary hypertension, arthralgia, and renal insufficiency (2,6).

The oral and maxillofacial manifestations include skin and oral mucosa atrophy, reduced maximum opening of the mouth, mask-like appearance, trismus, muscular atrophy, thin atrophied lips, trigeminal neuralgia, bone resorption of the jaws, and periodontitis in the orofacial region (2,4). Fibrosis of the salivary glands may cause hyposalivation and xerostomia (5).The temperomandibular joint may also be affected and result in pseudo ankylosis (1,5). The uniform widening of the periodontal ligament space, especially around the posterior teeth, can be seen radiographically (5).

In this study, the oral symptoms of a patient admitted to our clinic with dysphagia, periodontal problems and mouth opening limitation ave been presented.

## CASE REPORT

The department of Oral and Maxillofacial Radiology, Faculty of Dentistry, Akdeniz University, referred a 46-year-old male patient who had clinical typical skin lesions and disorders of upper limb flexion. He was admitted to our clinic with complaints of tooth pain. The medical history revealed that he suffered from polyarthralgia and sclerodactyly complicated by digital ulcer. Hand arthritis associated with sclerodactyly led to flexion contracture and a claw-like deformity (Figure 1). The patient has been using Azathioprine 50 mg per day for fifteen years.

Extraoral examination revealed loss of expression lines on the face and mouth opening limitation (32 mm), giving a mask-like appearance. Intraoral findings included loss of papillae on the tongue, generalized pallor and blanching of the mucous membrane, and diffuse fibrosis of the buccal mucosa with loss of normal elasticity (Figure 2). Periodontal tissue destruction was determined in the teeth in both the examination and panoramic radiography. Gingival recession and tooth mobility were the periodontal findings (Figure 3).

## DISCUSSION

Vasculopathy, fibrosis, and inflammation/autoimmunity contribute to tissue damage, although the etiology of SSc is not fully understood (2,7,8). It affects patients in the third or fifth decade (7,8). SSc is nearly 8 times more common in women than in men (1,4,8). However, the patient presented in our case was a male and 46 years old.

Orofacial features are found in about 80% of SSc patients (2). The oral and maxillofacial region are mainly affected as a result of collagen deposition in the subcutaneous tissue of the skin, causing facial muscle atrophy and skin fixation to the underlying structures (2). Oral problems of patients with SSc include decreased opening of the mouth, decreased saliva production, microstomia, increased number of missing teeth, osseous resorption of the mandible, and widening of the periodontal ligament space (PLS) (8,9).

These patients have mask-like facies as a result of hardening and tightening of the skin inducing changes in the face, and reduced mouth opening (2,5,9). In our present study, the patient showed reduced mouth opening and had a

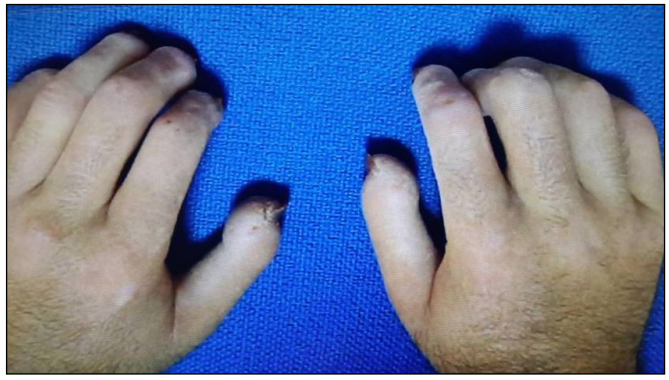

**Figure 1:** Flexion contracture of upper extremity and digital ulcers due to scleroderma.

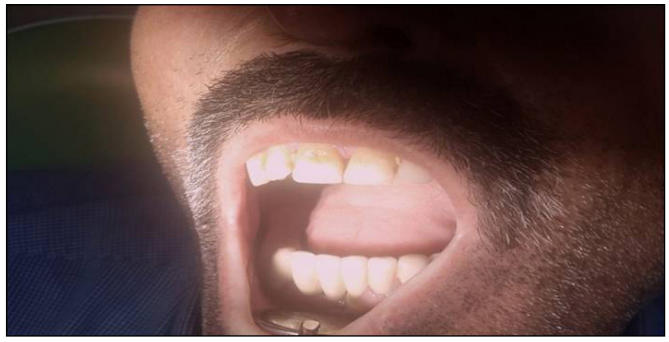

**Figure 2:** Mouth opening limitation.

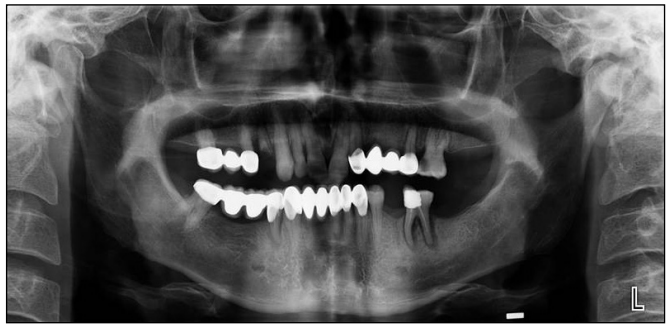

**Figure 3:** Patient's panoramic radiograph.

characteristic face, and the lack of mouth opening was among the first complaints. These patients also often have severe flexion deformities of their fingers such as claw-like fingers and decreased manual dexterity (4,9). Our patient also had such finger deformities. It may therefore be difficult to ensure oral hygiene in these patients (4). This may be the reason for the poor oral hygiene and increased number of missing teeth in our patient.

Dry mouth is another oral finding in scleroderma. This symptom may be related to salivary gland fibrosis (10,11). The unstimulated salivary flow rate was measured and a low flow rate detected in our patient.

The uniform widening of the periodontal ligament space is one of the earliest manifestations of the disease (9). The posterior teeth are involved more frequently than the anterior teeth (9). Enlarged periodontal ligament space may be caused by excess deposition of collagen or increase in masticatory occlusal forces (5,9). Dagenais et al. showed that PLS widening of at least one tooth was noted in 38% of the patients (8). Jagadish et al. reported that 66% of the patients had a widened periodontal ligament space (5). In our patient, widening of the periodontal ligament space was detected in the posterior teeth. Using the periodontal disease index (Ramfjord 1959), the periodontal attachment loss was measured as 3-6 mm.

Bone resorption of the mandibula is another clinical manifestation of SSc (5). Mandibular findings include resorption of the angle, condyle, coronoid process, ascending ramus, and antegonial notch (2,4). Resorption of the mandibula can be due to excessive pressure exerted by hardened overlying tissues and the vasculopathy associated with this disease (2,5). Jagadish et al. found that the angle of the mandible was involved in 83.33% of the patients (5). The mandibular resorption rate was reported to be lower, at 7% and 10% in other studies (12,13). We found no angular bone defects in our patient.

SSc is a multisystem connective tissue disease with numerous oral and maxillofacial manifestations. SSc can cause various oral problems, most commonly limited mouth opening. The best treatment option for this problem is probably mouth stretching exercises. Routine dental control should be recommended to patients with scleroderma for the maintenance of good oral health. All SSc patients should have regular follow-up with a yearly panoramic radiograph for other oral and maxillofacial manifestations.

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