INTEGRATIVE JOURNAL OF MEDICAL SCIENCES
ISSN: 2658-8218
MBMJ

2025, Volume 12, ID 746
DOI: 10.15342/ijms.2025.746

CASE REPORT

Systemic Scleroderma: Clinical Features and Management in Oral Surgery – A Case Report

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ABSTRACT
Introduction: Systemic scleroderma is a rare autoimmune disease characterized by progressive fibrosis of connective tissue and vascular damage, frequently involving the skin, internal organs, and oral cavity. Oral findings, including microstomia, xerostomia, telangiectasias, gingival alterations, and mandibular bone resorption, are common and significantly impact masticatory function, nutrition, oral hygiene, and dental care. **Case Report:** We report the case of a 45-year-old female patient diagnosed with systemic scleroderma in 2010, who sought consultation for the rehabilitation of her oral cavity. Extraoral examination revealed a typical scleroderma facial appearance and microstomia. Intraoral findings included poor oral hygiene, buccal telangiectasias, an atrophic and depapillated tongue, and gingival inflammation with signs of periodontitis. Panoramic radiography demonstrated bone resorption and widening of the periodontal ligament spaces. The patient's follow-up regarding etiological therapy was carried out rigorously; however, the prosthetic treatment could not be completed due to her lack of motivation. **Conclusion:** This article highlights the oral and extraoral clinical signs of systemic scleroderma and discusses the precautions necessary for managing patients with this condition in oral surgery. Effective treatment requires a multidisciplinary approach, including physiotherapy for microstomia, preventive strategies for hyposalivation, and careful selection surgical techniques. Prosthetic rehabilitation must be tailored to accommodate limited mouth opening, while implant placement remains a complex but viable option in select cases. Clinicians should adopt individualized treatment plans to optimize oral function, minimize complications, and improve the patient's quality of life.

KEYWORDS: systemic scleroderma; systemic sclerosis; oral surgery; case report.

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INTRODUCTION
Systemic scleroderma, or systemic sclerosis, is a chronic connective tissue disease characterized by skin fibrosis and potentially severe vascular and visceral lesions that may be life-threatening [1].
The disease involves excessive collagen production and connective tissue fibrosis, alongside specific microvascular pathologies and inflammatory infiltrates. The etiology remains poorly understood but is considered multifactorial, involving environmental factors and immune dysregulation on a genetically predisposed background [2].
Clinically, the disease presents with severe skin hardening, particularly on the extremities and face. Raynaud's phenomenon, marked by painful digital ischemia, is an early symptom, often progressing to digital ulcers and necrosis. Systemic scleroderma may affect the lungs, heart, kidneys, gastrointestinal tract, and bones [3,4]
Orofacial manifestations such as limited mouth opening, tongue rigidity, and mandibular bone resorption are reported. This case illustrates the spectrum of oral manifestations and highlights therapeutic nuances [3].

CASE REPORT
The patient, a 45-year-old woman, was referred to the oral surgery department at the Dental Consultation and Treatment Center in Rabat, for dental extractions to restore oral health. During the anamnesis, she reported being diagnosed with systemic sclerosis in 2010 and has been, since, on Methotrexate and corticosteroid therapy.
Facial examination shows characteristic tight, mask-like features with pale skin, absent expression lines, retracted eyelids, pinched nose with narrow alae, and thin, pale lips with perioral radial wrinkles (Figure 1).
Inspection of both hands shows mild curvature of all fingers (Figure 2), along with signs of sclerodactyly and ulcerations at the fingertips (Figure 3).
Examination of lymph nodes and the temporomandibular joint is normal, but mouth opening is severely restricted (microstomia, two-finger breadths) (Figure 4).
The oral mucosa is pale, dry, and exhibited multiple buccal telangiectasias (Figure 5). The tongue is dry, depapillated, with reduced volume and mobility; with a thickened, short lingual frenulum making tongue protrusion difficult (Figure 6).

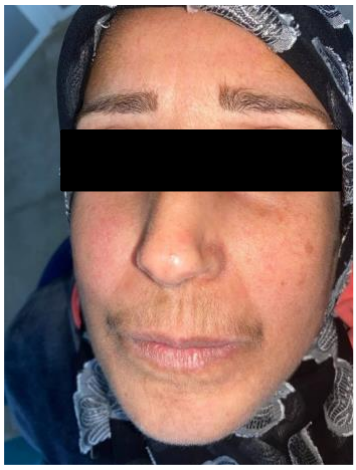

Figure 1: Exobuccal view showing a typical sclerodermic face.

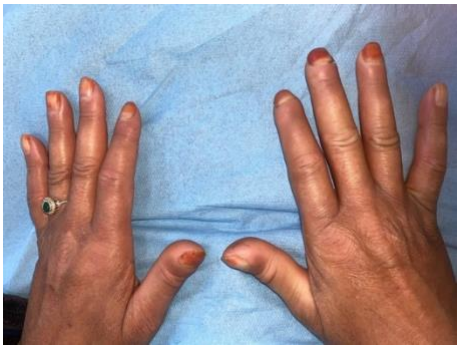

Figure 2: Photograph of the hands showing sclerodactyly.

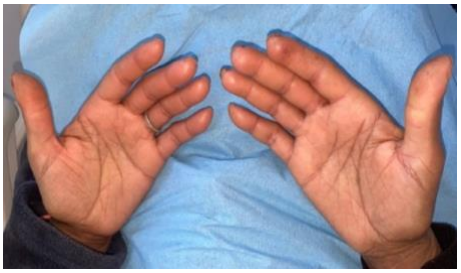

Figure 1: Onset of digital ulceration.

Figure 2: Extraoral photograph showing a Microstomia with a two-finger opening.

Gingival examination reveal generalized moderate marginal and papillary inflammation, pronounced papillary hypertrophy in the anterior maxillary and mandibular regions, these findings were associated with supragingival and subgingival calculus deposits, indicative of poor oral hygiene (Figure 7).

Dental examination notes two upper right premolar roots, provisional restoration on the upper right lateral incisor, and a horizontally impacted lower right wisdom tooth. Periodontal probing identifies periodontal pockets. Panoramic radiography reveals absence of interradicular septa, periodontal ligament widening, and primarily horizontal bone resorption in the coronal third of all teeth (Figure 8).

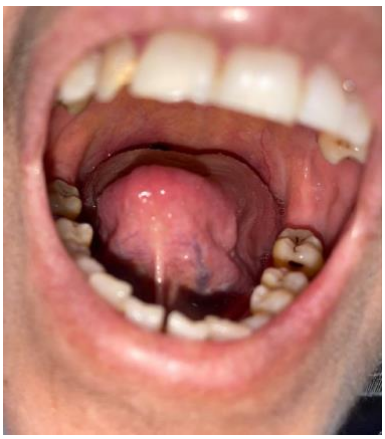

Figure 3: intraoral photograph showing short lingual frenulum restricting tongue protrusion.

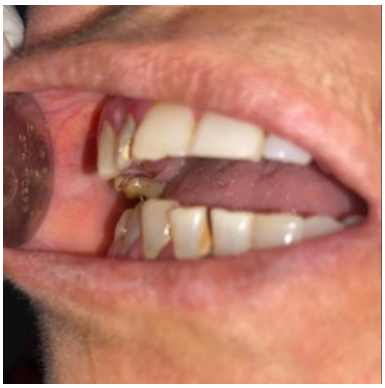

Figure 4 : intraoral photograph showing buccal telangiectasias.

Figure 5 : intraoral photograph showing hypertrophy of gingival papillae.

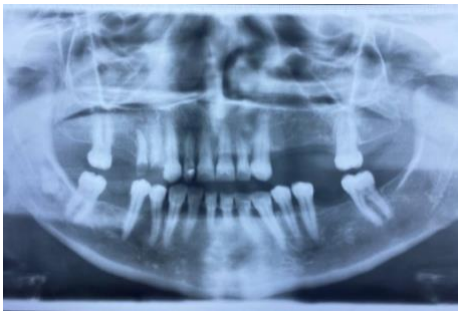

Figure 6 : Panoramic X-ray.

The therapeutic approach includes oral hygiene motivation, supra- and subgingival scaling with root debridement, extraction of the roots of the two upper right premolars under antibiotic prophylaxis (amoxicillin 2 g, one hour before the procedure), endodontic treatment of the upper right lateral incisor, and prosthetic rehabilitation. Regarding the horizontally positioned impacted wisdom tooth, therapeutic abstention is chosen due to the limited mouth opening.

We explained to the patient that she must maintain proper oral hygiene to avoid invasive dental treatments, which are difficult in her case due to limited mouth opening.

Note that the patient's complete blood count (CBC) was normal.

The patient's follow-up regarding etiological therapy was carried out rigorously; gingival inflammation decreased after periodontal debridement, and there are no periodontal pockets remaining. However, the prosthetic treatment could not be completed due to her lack of motivation.

Discussion

Systemic scleroderma is a rare connective tissue disorder of unknown origin, characterized by three main pathological abnormalities: widespread fibrosis, vasculopathy, and immunological dysfunctions. This condition leads to hardening and thickening of the skin and internal organs, potentially affecting multiple body systems, including the cardiovascular, pulmonary, and digestive systems [5].

Orofacial manifestations of systemic sclerosis are present in 80% of affected patients, representing most clinical symptoms. These are caused by excessive collagen deposition in subcutaneous tissues, facial muscle atrophy, and the adhesion of the skin to underlying structures [6,7]. Skin thickening and tightening result in facial changes leading to a typical scleroderma face that appears smooth and taut, lacking facial wrinkles, often described as "mask-like" or resembling an "Egyptian mummy". Lip fibrosis, resulting in microcheilia, causes reduced mouth opening, known as microstomia, which significantly hampers nutrition, oral hygiene, dental care, prosthetic rehabilitation, and intraoral radiography. The nose appears pinched, with atrophied alae due to nasal cartilage atrophy [1,8,9,10].

All these manifestations were observed in our patient.

Other common cutaneous manifestations include hypo- and hyperpigmentation of certain skin areas, hypohidrosis, and changes in lip coloration [5].

Intraorally, the oral mucosa, often presenting telangiectasia, appears pale, tense, atrophic, and inelastic. Another frequent oral manifestation is salivary gland fibrosis, leading to hyposalivation and/or xerostomia, contributing to secondary dry syndrome. The oral mucosa and dental tissues may show lesions and ulcers caused by gastroesophageal reflux, frequently observed in systemic sclerosis patients due to esophageal dysmotility or side effects of treatments [9,11].

Fibrosis in the orofacial region may extend to the oropharyngeal and esophageal mucosa, the tongue, as well as the hard and soft palate, leading to swallowing and speech difficulties, as well as voice changes. These alterations, combined with reduced facial expression, impair communication abilities and social interactions for patients with systemic sclerosis [12].

The oral mucosa and dental tissues may exhibit lesions and ulcerations caused by gastroesophageal reflux, often associated with esophageal dysmotility or side effects of treatments. In cases of gastroesophageal reflux, mucogingival paresthesia may occur, with an increased risk of tongue or pharyngeal carcinoma [11].

On the osseous level, Rout et al. (1995) and Haer et al. (1996) reported mandibular resorption incidence in systemic sclerosis ranging from 10% and 20 to 33%, respectively. The mandibular angles are most frequently affected, often bilaterally, followed by the condyle, coronoid process, and the posterior edge of the ascending rami. Early detection of condylar resorption is crucial, as it can limit mouth opening and lead to temporomandibular joint ankylosis [3,13].

These resorptions may result from excessive pressure exerted by hardened overlying tissues. In systemic sclerosis, increased muscle fibrosis makes muscles bulkier. Simultaneously, reduced muscle vascularization due to arterial wall fibrosis leads to muscle atrophy, exerting pressure on the bone at attachment sites. Mandibular resorption may also be attributed to skin tension exerting pressure on the bone, causing pressure resorption. This pressure intensifies during jaw movements, as stiff muscles restrict mandibular motion [13,14].

Periodontally, Weisman et al. (1978) and Eversole et al. (1984) observed that systemic sclerosis patients with severe oral fibrosis and limited mouth opening suffered mucogingival issues, such as loss of attached gingiva and multiple gingival recession sites [1].

Widening of periodontal ligaments, particularly around molars and premolars, as well as dental root resorption, may also occur. Although rarely reported, these manifestations might be caused by excessive collagen and oxytalan fiber deposition, resulting in subsequent alveolar crest resorption [11,15,16].

In our case, there was no root resorption, but we noted a pronounced periodontal ligament widening around the molars.

The American Academy of Periodontology (AAP) and the European Federation of Periodontology (EFP) have included periodontal manifestations such as bone resorption and periodontal ligament widening as direct manifestations of certain systemic conditions, including systemic scleroderma, even in the presence of poor oral hygiene that may interfere with the local etiology of the disease.

Nonetheless, in our case, it is not possible to conclude whether this periodontitis is a manifestation of systemic sclerosis or a periodontitis related to bacterial plaque in the context of systemic sclerosis.

Currently, management recommendations for systemic sclerosis-related oral complications are well-established. These were developed by a multidisciplinary group of French experts, validated by a review committee, and endorsed by the American College of Rheumatology (ACR) (2018) [11].

Microstomia can be managed through physiotherapy, which involves a series of exercises to stretch the cheeks and mouth. Additionally, intense pulsed light therapy has been tested, although its efficacy remains uncertain. For more severe cases, commissuroplasty combined with an autologous fat graft may be considered. These therapeutic options should be tailored to the severity of the condition and the specific needs of the patient, in collaboration with a multidisciplinary team [17].

In patients with systemic scleroderma, oral mucosal ulcers or atrophy may occur due to nutritional deficiencies or as adverse effects of medications, particularly methotrexate, azathioprine, or cyclophosphamide. Folic acid supplementation is recommended to reduce the risk of toxicity and methotrexate-associated stomatitis [17].

If hyposalivation occurs, management strategies include frequent oral hydration, the use of sugar-free chewing gum

to stimulate salivary flow, and the use of artificial saliva substitutes; Regular dental visits for preventive measures, such as the application of topical fluoride, are strongly advised [17].

Routine dental care can be safely performed in systemic sclerosis patients, with local anesthesia using 3% lidocaine if necessary. Adrenaline should be avoided as it may worsen microangiopathy [9].

Endodontic treatments, prosthetic preparations, and dental surgeries are best scheduled in the morning. Common dental materials such as composite resins, glass ionomer cement, and amalgam can be used safely [11].

Mucogingival surgery is not recommended due to limited efficacy and increased postoperative complications [11].

For patients with severe microstomia, prosthetic treatments may be challenging. Split impressions can be considered to facilitate prosthetic tolerance. For partial or complete denture wearers, antiseptic gels can be applied to the denture's internal surface to alleviate dryness-related pain. For ulcers, the denture fit must be adjusted, and topical anesthetics or hyaluronic acid gels can be used for pain relief. Fungal infections require antifungal treatment, such as oral amphotericin B.

Dental implants in patients with systemic scleroderma are poorly documented. Implant-prosthetic treatment is complex and requires an individualized evaluation of risks and benefits, combined with interdisciplinary consultation. Specific challenges include limited mouth opening, increased susceptibility to periodontal diseases due to hyposalivation, microstomia, and ankyloglossia, as well as reduced manual dexterity resulting from systemic scleroderma [18].

However, if the periodontal condition is favorable, dental implants may be considered in two main situations: to stabilize a removable prosthesis in the anterior mandibular region, improving comfort and reducing the risk of ulcers, or to restore aesthetics in the maxillary incisor and canine region. The use of overdentures can be favored to promote salivary flow and facilitate maintenance. Implants are contraindicated in patients with marked gingival fibrosis and severely compromised microvascularization [1].

In cases of oral or dental infections, antibiotic therapy is recommended. The combination of amoxicillin and metronidazole is the first-line treatment, particularly in the presence of generalized aggressive periodontitis. In cases of allergy or concurrent methotrexate use, macrolides may be prescribed to reduce the risk of methotrexate toxicity when used alongside penicillin. For patients on anticoagulants, 5% tranexamic acid mouthwash is required [17].

Dental care should be delivered quadrant by quadrant to avoid lengthy sessions, often poorly tolerated due to limited mouth opening [11].

CONCLUSION

Systemic sclerosis is a complex autoimmune disease with diverse manifestations, including significant oral complications that affect patients' quality of life. Issues such as microstomia, xerostomia, bone resorption, and periodontal anomalies require tailored, multidisciplinary management.

Well-defined current recommendations provide clear guidelines for optimizing care and preventing complications. Increased practitioner awareness and specific therapeutic protocol adaptations are crucial to improving the prognosis and comfort of systemic sclerosis patients.

AUTHORS' CONTRIBUTIONS

The participation of each author corresponds to the criteria of authorship and contributorship emphasized in the Recommendations for the Conduct, Reporting, Editing, and Publication of Scholarly Work in Medical Journals of the International Committee of Medical Journal Editors. Indeed, all the authors have actively participated in the redaction and revision of the manuscript and provided approval for this final revised version.

ACKNOWLEDGEMENT:

None

PATIENTCONSENT:

Written informed consent was obtained from the patientfor thepublicationofthis casereport.

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