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Dual trouble: coexisting oral submucous fibrosis (OSMF) and scleroderma - A rare case report

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Abstract

Background: OSMF and Scleroderma are two distinct and potentially debilitating conditions that affect different organ systems, yet rarely do they occur simultaneously in the same individual. Oral Submucous Fibrosis is a chronic, progressive, and potentially malignant disorder that primarily affects the oral cavity, characterized by the deposition of fibrous tissue leading to restricted mouth opening, mucosal rigidity, and difficulty in eating and speaking. On the other hand, Scleroderma is a chronic connective tissue disorder characterized by excessive deposition of extracellular matrix in the connective tissues resulting in vascular disturbances. The aetiology of the disease is unknown and the major consequence is tissue hypoxia. Mostly, manifestations include atrophy of the skin, and/or mucosa, subcutaneous tissue, muscles and even internal organs.

Case scenario: A 39-year-old female of Dravidian origin presented to our out-patient department with a chief presenting complaint of reduced mouth opening and increased sensitivity to hot and spicy food for 3 months. Her medical and dental history was noncontributory. On examination, salt and pepper

pigmentations were noted to be present in the mid-forehead region, chin, nose and periorbital regions in addition to trunk and back areas.

A thorough oral examination showed limited mouth opening (2 fingers), pigmented and blanched buccal mucosa, palate with decreased elasticity was appreciated. Areas of erosions on B/L buccal mucosa and tobacco associated keratosis were noted in the vestibular regions. Physical examination revealed stunted phalanges and tightness in both right and left hands.

Conclusion: This case report not only contributes to the existing medical literature on these two disorders individually but also provides valuable insights into their coexistence, which can aid clinicians in recognizing and managing such complex cases more effectively. As the prevalence of autoimmune and chronic connective tissue disorders continues to rise, the documentation of unique cases like this one is essential for enhancing medical knowledge and improving patient care in the field of rheumatology, dentistry, and beyond.

Keywords: OSMF, Sclerosis, Systemic scleroderma, CREST syndrome

OSMF and Scleroderma are two distinct and potentially debilitating conditions that affect different organ systems, yet rarely do they occur simultaneously in the same individual. Oral Submucous Fibrosis is a chronic, progressive, and potentially malignant disorder that primarily affects the oral cavity, characterized by the deposition of fibrous tissue leading to restricted mouth opening, mucosal rigidity, and difficulty in eating and speaking. On the other hand, Scleroderma, also known as systemic sclerosis, is a connective tissue disorder that can involve multiple organ systems, resulting in skin thickening, vascular abnormalities, and internal organ dysfunction.

OSMF is a chronic, progressive fibrotic disorder affecting the oral cavity, primarily associated with the habitual use of betel nut and tobacco. On the other hand, Scleroderma, or systemic sclerosis, is an autoimmune connective tissue disorder characterized by excessive collagen deposition leading to skin thickening and involvement of multiple organ systems. Both OSMF and Scleroderma are distinct entities with unique clinical features and underlying pathophysiology. However, their coexistence in the same individual is exceptionally rare and presents a diagnostic and therapeutic challenge for clinicians.

The coexistence of these two conditions in a single patient presents a diagnostic and management challenge, as their clinical manifestations may overlap, leading to diagnostic confusion and delayed treatment. Additionally, the pathophysiological mechanisms underlying the development of both OSMF and Scleroderma are complex and not fully understood, making the understanding of their coexistence even more intriguing and significant for clinical practice.

In this case report, we present a rare and fascinating clinical scenario of a patient diagnosed with both Oral Submucous Fibrosis and Scleroderma simultaneously. Through a comprehensive evaluation of the patient's clinical presentation, diagnostic investigations, and treatment approach, we aim to shed light on the distinctive challenges faced in diagnosing and managing this coexistence. Furthermore, this report emphasizes the importance of multidisciplinary collaboration and highlights the potential implications for the prognosis and quality of life of patients with these dual conditions.

Case presentation

A 39-year-old female patient presented to the outpatient section of Department of Oral Medicine and Radiology of the institute with complaints of progressive difficulty

in mouth opening, facial skin tightness, and dysphagia for the past two years. The patient reported a history of tobacco chewing for more than 20 years, with a habit of placing quid in the buccal vestibule. She had no significant medical or family history of autoimmune diseases or connective tissue disorders. On general examination, she was found to be conscious and oriented with well awareness of the surroundings. Her vital signs were also within normal limits.

Upon oral examination, the patient exhibited limited mouth opening (interincisal distance of approximately 20 mm) due to marked fibrosis of the buccal mucosa and inability to protrude his tongue. An oral examination also revealed microstomia due to rigid perioral skin, xerostomia (positive tongue blade test), and an erythematous patch on her B/L buccal mucosae The oral mucosa appeared pale, with vertical fibrous bands palpable on palpation. The presence of a blanched marble-like appearance of the oral mucosa, especially on the cheeks and retromolar areas, raised suspicion of Oral Submucous Fibrosis (OSMF). An intraoral examination revealed shrunken and rigid left side of his tongue otherwise his mucosa appeared to be normally moist. A hard tissue examination offered no relevant findings. Biopsies were taken from the affected areas for histopathological confirmation.

The patient was also subjected to dermatological evaluation and in addition to oral findings, the patient exhibited characteristic cutaneous changes associated with Scleroderma. Presence of diffuse salt and pepper pigmentations were noted in mid forehead, chin, nose and periorbital regions in addition to trunk and back areas.

A visually evident loss of fat, muscles, and subcutaneous tissue resulted in a shrunken appearance her face. The skin over his face, particularly around the mouth, appeared thickened and tight, with a reduced ability to form facial expressions. On palpation, the skin of her perioral and neck regions was sclerotic. The skin on her fingers also appeared to be pale on inspection and indurated when palpated. According to our patient, this paleness spreads symmetrically to all ten fingers when exposed to cold, suggestive of Raynaud's phenomenon. Additionally, his fingers showed evidence of sclerodactyly, with tightening and thickening of the skin, resulting in limited finger mobility elaborated in the distal pharyngeal phalanges. Skin on the affected side appeared to be sclerotic. An ocular examination disclosed no abnormalities. The clinical findings were suggestive of progressive systemic sclerosis; hence, we recommended radiological and serological investigations.

Presence of oedematous, pitted scarring and ulcerations noted over the digital tips. Positive Ingram sign and presence of rhagades in conjunction with Reynold's phenomenon was noted in the patient. On systemic evaluation patient gave history of difficulty in breathing, orthopnoea and instances of paroxysmal nocturnal haemoglobinuria, dysphagia and reflux esophagitis suggestive of CREST as a differential. Nailfold capillaroscopy was performed, revealing dilated and irregularly-shaped capillaries, supporting the diagnosis of Scleroderma.

Radiological examinations show presence of generalised and widened PDL space with intact lamina dura as well as areas of mild maxillofacial bone resorptions along right mandibular angle, condyle, and coronoid process as noted from panoramic imaging. Advanced imaging findings using CBCT were concurrent with those of conventional images.

Discussion

In India the overall incidence is 0.2%–0.5%. Ranganathan (2004) found the risk of developing OSMF was double in younger patients less than 21 yrs and the younger patients develop OSMF in 3.5 yrs, while the duration for those above 21 years was more than 6.5 years. OSF is a chronic, debilitating disease of the oral cavity characterized by inflammation and progressive fibrosis of the submucosal tissues (lamina propria and deeper connective tissues). It results in marked rigidity and an eventual inability to open the mouth. The buccal mucosa is the most commonly involved site, but any part of the oral cavity can be involved, even the pharynx. Several factors contributing to submucous fibrosis include general nutritional or vitamin deficiencies and hypersensitivity to various dietary constituents. The primary factor appears to be habitual chewing of the areca (betel) nut. It appears that the condition is due to impaired degradation of normal collagen by fibroblasts rather than excess production.(1)

The prevalence of scleroderma, though much lesser than arthritis affects nearly two of 10,000 persons. The predilection for female gender in the population in the ratio 5:1and a general onset between 30 and 50 years is similar to rheumatic diseases.

Scleroderma, a chronic sclerosing disease of the connective tissues, is characterized by an excess production and accumulation of collagen and results in chronic hardening and thickening of the skin.(2,3)

Inflammation manifests initially as a perivascular macrophage infiltrate. The most serious complications namely pulmonary arterial hypertension and renal crisis arises from prominent thickening of vessels due to injury to endothelial cells. Increased deposition of fibroblasts results in destruction of normal tissues that leads to tissue and organ dysfunction in skin, lungs, kidneys and GI tract.(4)

There are different autoantibodies which are extremely helpful in forecasting different subtypes of scleroderma. However, autoantibody absence does not preclude the diagnosis of this disease as 20 % of the patients with different subtypes of scleroderma do not show these antibodies. Highly specific antinuclear antibodies (ANA) for scleroderma include anti-single-stranded, anti-histone, and anti-topoisomerase antibodies. Some other ANA that commonly present in scleroderma include anticentromere, anti-U3-RNP, anti-Th, anti-fibrillin, anti- phospholipid, and antimitochondrial antibodies. Both of our cases were positive for a few of these sero-logical markers and the markers were extremely helpful in yielding a final diagnosis.

The coexistence of Oral Submucous Fibrosis and Scleroderma in our patient represents an exceedingly rare clinical scenario. The relationship between the two conditions remains poorly understood, and their simultaneous occurrence raises intriguing questions about potential shared pathogenic mechanisms.

Tobacco chewing has long been recognized as a significant risk factor for the development of Oral Submucous Fibrosis. It is interesting to note that our patient's history of tobacco chewing may have played a contributory role in the pathogenesis of both OSMF and Scleroderma. It is plausible that the chronic inflammation induced by tobacco components may have triggered an autoimmune response, leading to the development of Scleroderma.(5–7)

The diagnosis of this dual condition posed a challenge due to the overlapping clinical features of OSMF and Scleroderma. The clinical suspicion for Scleroderma was further supported by positive ANA and ACA blood tests. This highlights the importance of a thorough evaluation

and collaboration between Rheumatology and Oral Medicine specialists to ensure accurate diagnosis and appropriate management.

Although the patient showed some improvement with the implemented treatment strategies, the chronic nature of both conditions necessitates long-term follow-up and continuous management. Further research is warranted to explore the underlying pathophysiological links between OSMF and Scleroderma, which could potentially pave the way for targeted therapeutic interventions.

Conclusion

This case report presents a unique and challenging clinical scenario of coexisting Oral Submucous Fibrosis and Scleroderma. The simultaneous occurrence of these two conditions requires a multidisciplinary approach involving Rheumatology and Oral Medicine specialists for accurate diagnosis and effective management. While the precise mechanisms underlying their coexistence remain unclear, this case emphasizes the importance of recognizing rare presentations and facilitating further research to enhance our understanding of complex autoimmune and connective tissue disorders. Early diagnosis and appropriate management can lead to improved patient outcomes and enhance their quality of life.

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Legends Figures and Tables

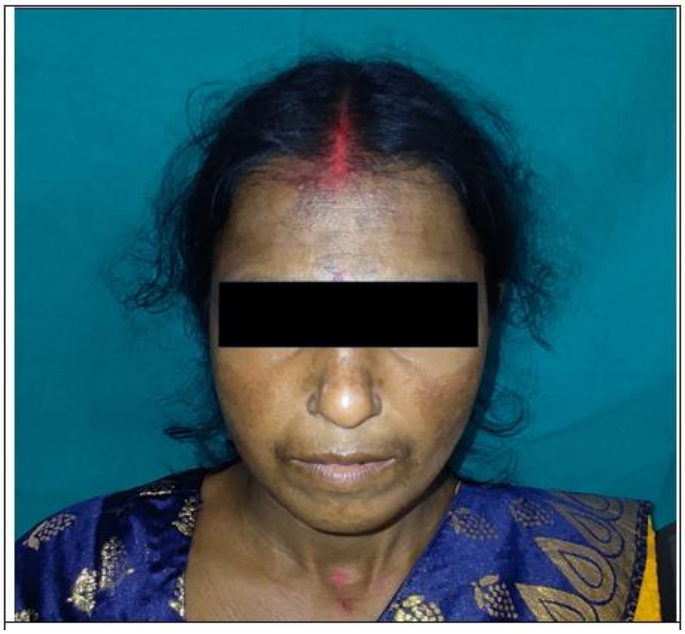

Figure 1: Frontal profile view showing diffuse salt and pepper pigmentations, malar melasma

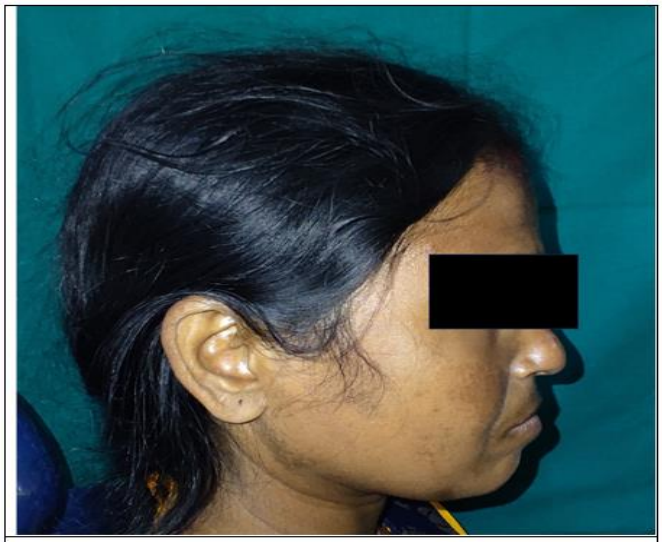

Figure 2: right profile view showing marked sclerosis of the orofacial muscles and skin.

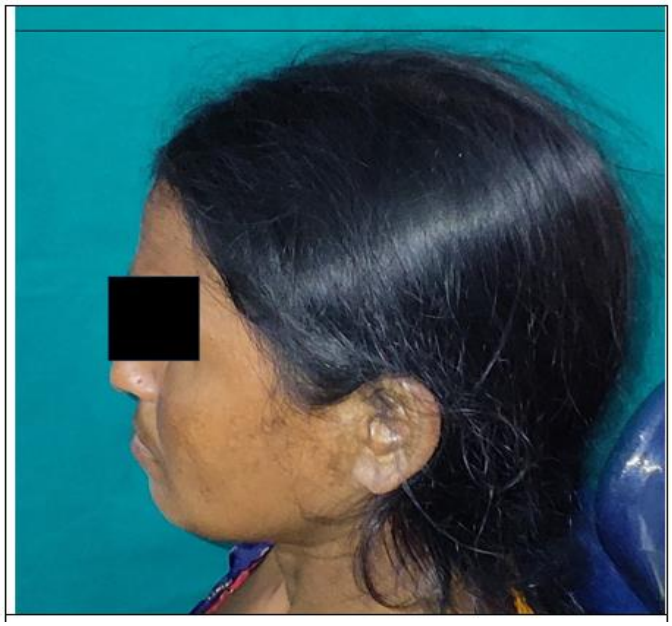

Figure 3: Left profile view showing marked sclerosis of the orofacial muscles and skin

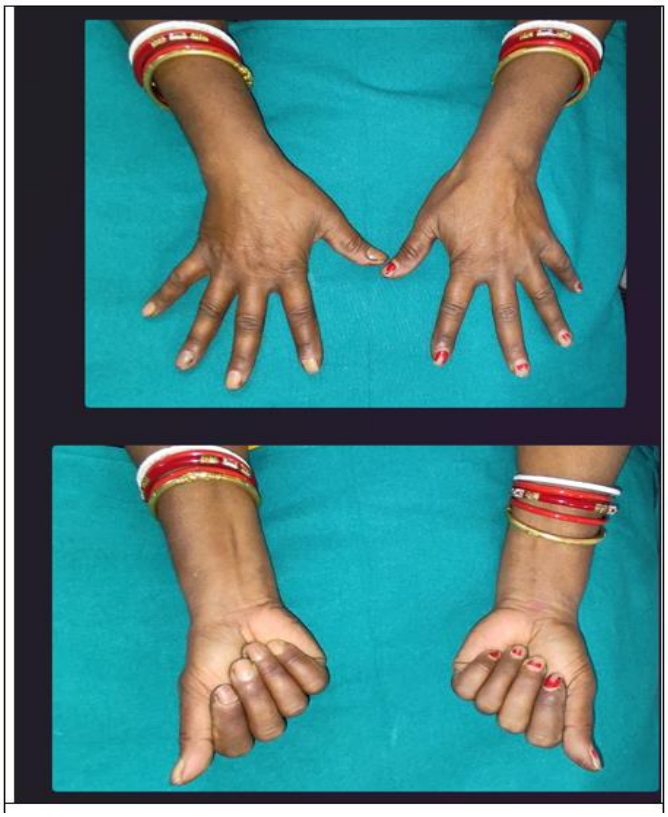

Figure: 4 Terminal phalanges

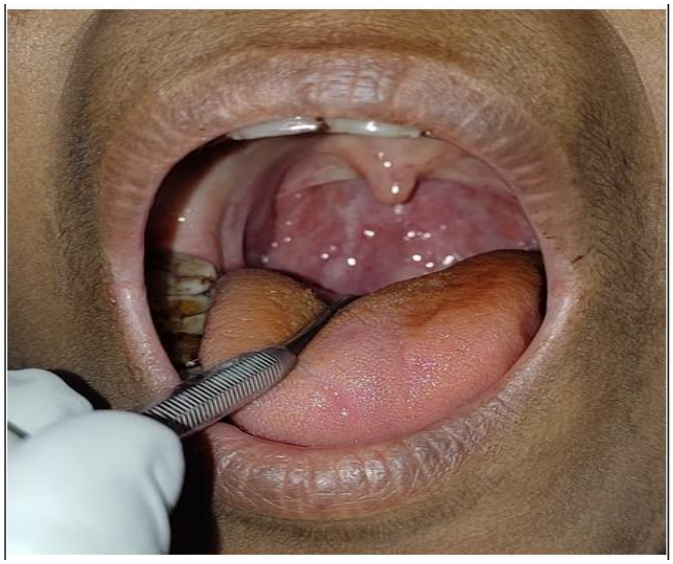

Figure 5: intraoral examination- shrinking uvula

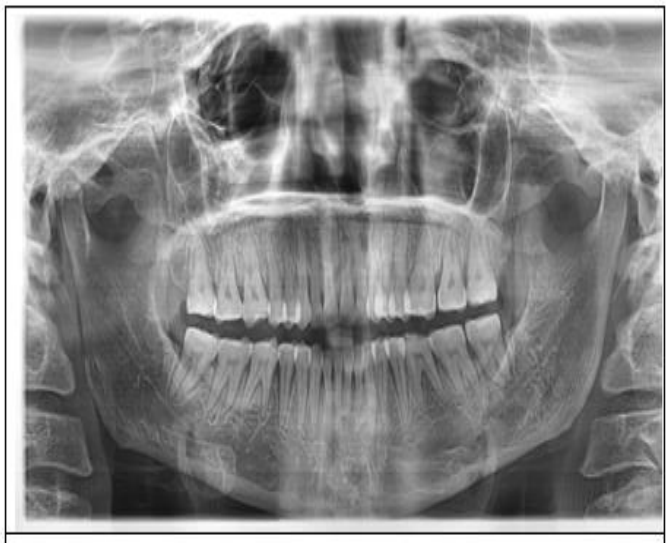

Figure 6: Panoramic view showing generalized widened PDL space and intact lamina dura. possible resorption of right mandibular angle region

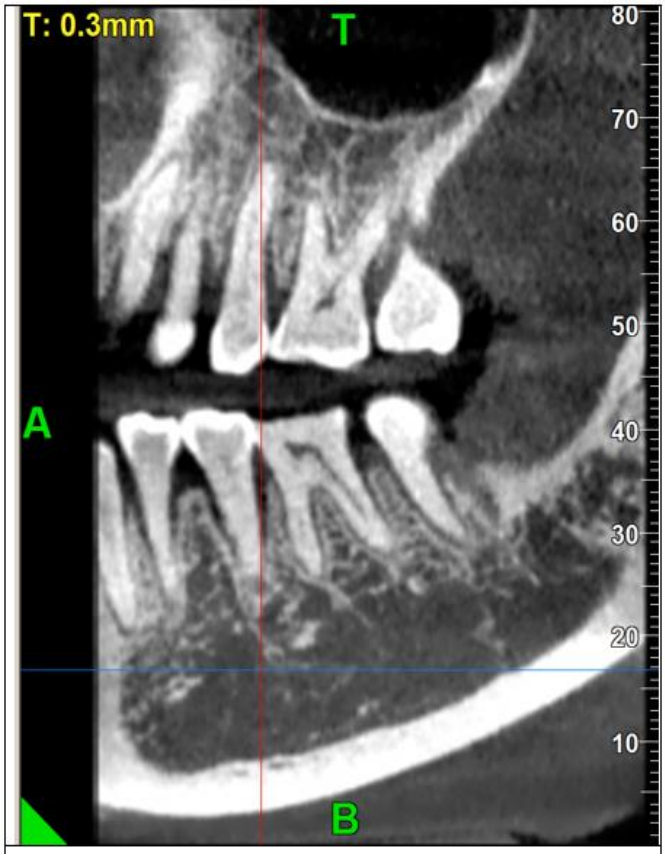

Figure 7: Sagittal sections showing widened PDL space and intact lamina dura (left side)

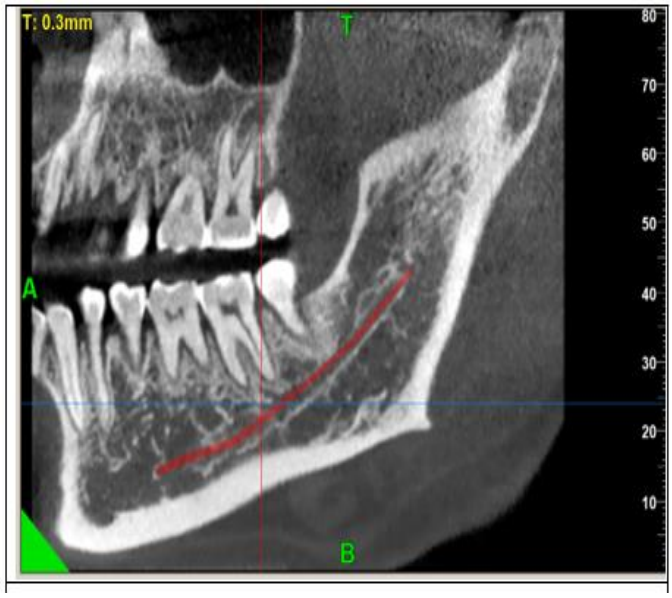

Figure 8: Sagittal sections showing widened PDL space and intact lamina dura (Right side)