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Orofacial Manifestations of Systemic Sclerosis: A Case Report

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Abstract: Systemic sclerosis is a rare connective tissue disorder that affects skin and viscera. It is characterized by severe and progressive fibrosis with vascular obliteration. The face and the oral cavity are frequently involved. The aim of our paper is to present the orofacial manifestations of a middle-aged female patient with systemic sclerosis.
Keywords: Systemic scleroderma, Fibrosis, Bone resorption, Xerostomia, Mouth diseases

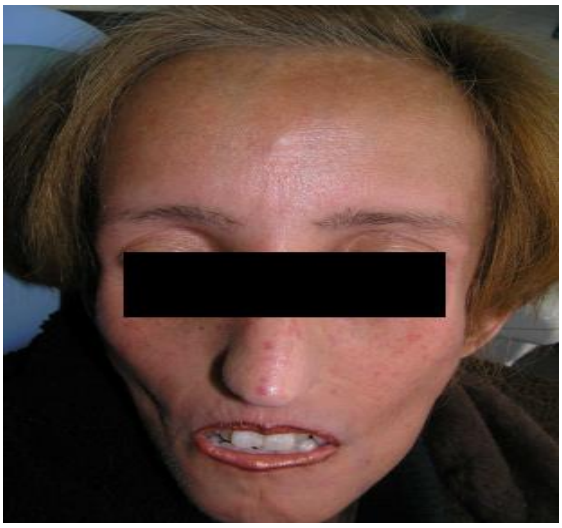
INTRODUCTION

Systemic sclerosis (SSc), also known as scleroderma, is a rare connective tissue disorder that affects skin and viscera, particularly the lung, heart and digestive tract. It is characterized by severe and progressive fibrosis as a consequence to excessive accumulation of collagen along with small arteries and arterioles disease due to thickening and proliferation of endothelial cells [1]. Although the pathogenesis of SSc remains incompletely understood, it involves complex interactions between vascular and immune system [2]. Women are predominantly affected (F/M sex ratio around 4:1) [1]. SSc is a heterogeneous disease in which clinical manifestations and sites involvement vary considerably. There are three main subsets of SSc; the limited cutaneous SSc, the diffuse cutaneous SSc and SSc without skin involvement [3]. The face and the oral cavity are frequently involved in SSc. The main clinical manifestations are thickening of the skin with mask-like facial expression, facial and oral telangiectasia, limitation of the oral aperture, dryness of the mouth, predisposition to dental caries and periodontal diseases [4].

The dentist must work in concert with the medical and paramedical team to improve the quality of life of patients with SSc. His role aims to screen, monitor and treat the orofacial complications of this disease. The aim of our paper is to present the orofacial manifestations of a middle-aged female patient with SSc.

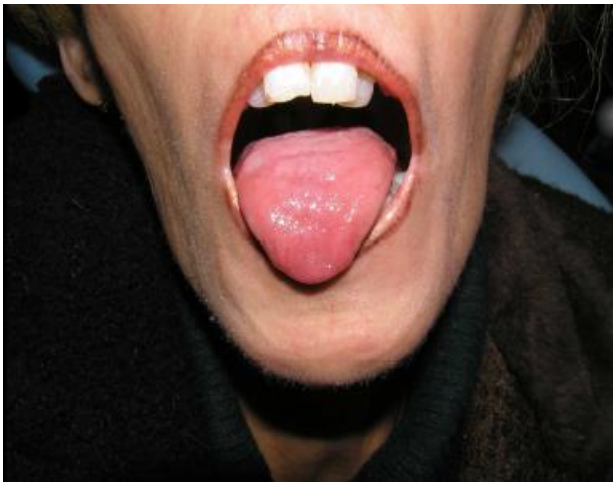
CASE REPORT

A 36-year-old woman was referred to our department with chief complaint of dental pain. She has been followed-up in the internal medicine department for a SSc for five years. The patient had an associated Sjogren's syndrome, a pulmonary hypertension and esophagitis with dysphagia. Physical examination showed classic orofacial signs and symptoms of SSc including tightening of the skin with a beak-like appearance. Her nose was pinched and deviated to the right side. There were several telangiectasias on the malar and the nasal region (fig−1).



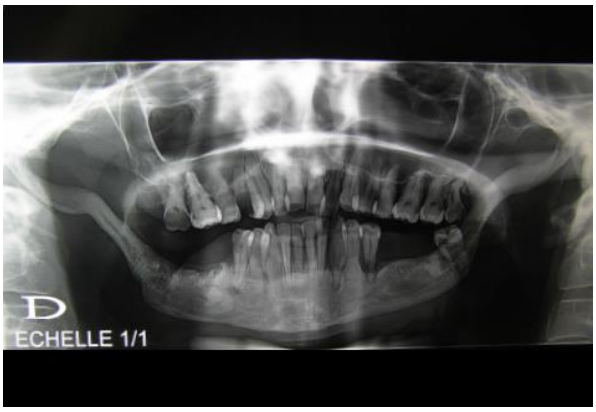
Fig−1: Facial view: A beak-like appearance with pinched and deviated nose associate with several telangiectasia's on the malar and the nasal region.

The oral aperture was severely reduced due to the perioral skin fibrosis. Intra oral examination showed fibrosis of the tongue with reduced mobility and loss of papillae, a poor oral hygiene and generalized periodontitis (fig−2).



Fig−2: Intraoral view: Reduced oral aperture, fibrosis of the tongue with reduced mobility and loss of papillae.

Examination of the temporomandibular joints detected no abnormalities. Panoramic radiograph showed severe resorption of the mandibular angle, posterior and inferior border, the coronoid and the zygomatic arch. We also noticed a generalized widening of the periodontal space, several retained teeth and dental caries (fig− 3).



Fig−3 : Panoramic radiograph : Severe resorption of the mandibular angle, posterior and inferior border, the coronoid and the zygomatic arch, a generalized widening of the periodontal space, several retained teeth and dental caries.

The patient received oral hygiene education, a dental scaling, root planning and extraction of the dental roots. The patient is still under regular follow-up.

DISCUSSION

Scleroderma originates from two Greek words “sclerosis” meaning hard and “derma” meaning skin. It is a potentially serious and debilitating connective tissue disease characterized by fibrosis of the skin and internal organs, a microcirculatory disease and production of autoantibodies [1].

SSc often affects the facial and oral structures and this involvement may even be the presenting symptom of this disease. Widening of the periodontal space is the most common radiological finding in patients with systemic sclerosis. This phenomenon remains unclear. Auluck *et al.* suggest that the widening of the periodontal space is attributed to primary occlusal trauma. In fact, increasing collagen deposit leads to tautness of the masticatory muscles which become bulky, resulting in the augmentation of the occlusal forces [5]. However, Mehra *et al*. suppose that systemic sclerosis is rather associated with vascular ischemia and atrophy of the masticatory muscle, therefore excluded the occlusal trauma hypothesis. They attribute the widening of the periodontal space to the increase of collagen synthesis in the periodontal ligament [6].

Osteolytic is a well-recognized skeletal feature of systemic sclerosis. This process most occur on the terminal phalanges, rib, cervical spine and the mandible [7]. Many hypothesis are proposed to explain this process. Seifert et al. speculate that pressure ischemia secondary to facial skin sclerosis and muscle atrophy is implicated in the pathogenesis of mandibular resorption [8]. However, Ruprecht *et al.* when investigating an extensive osteolytic of the coronoid process and the mandibular angle with MRI, find no abnormalities of the masticatory muscle or increase of fibrous tissue [9]. Ramon et al. assume that mandibular resorption is due to vascular ischemia. Considering the fact that vascular obliteration is common in systemic sclerosis and that mandibular resorption occurred in areas that are supplied by small muscular arteries, i.e., mandibular angle, coronoid process and condyle, this may explain the occurrence of the mandibular resorption [10].

Macrosomia and limitations of mouth opening are the result of cheek and labial skin sclerosis with gradual installation without joint participation. It is often associated with major functional impairment. Pizzo et al. have proven the efficiency of non-surgical management of microstomia by stretching exercises. All patients reported a subjective improvement in eating, speaking and in executing oral hygiene measures after 18 weeks of daily exercise [11]. Bilateral commissurotomy may be indicated in selected patients with severe mouth opening limitations [12].

Patients with systemic sclerosis seem to exhibit a poor oral hygiene and periodontal inflammation [12]. In fact, Sundqvist et al. noticed a reduced average of manual dexterity of 68%-80% in women with systemic sclerosis compared with healthy person [13]. This may explain the difficulty of ensuring an adequate tooth brushing. The orofacial complications constitute also a risks factors for poor oral hygiene particularly macrosomia and xerostomia [14]. Sicca syndrome is a common symptom in patients with systemic sclerosis. It is related to the sclerosis of the connective tissue of the salivary gland. Less frequently, it is incriminated to the

inflammatory infiltrate of the salivary gland related to an associate Sjogren's syndrome [15]. In both instance, mouth hydration with water, sialagogue treatment and salivary substitutes may be proposed [16].

The American college of rheumatology has established a set of recommendations for the care of oral involvement for patients with systemic sclerosis. It is recommended to screen the patient biannually for scaling and root planning with hygiene education. Conventional dental care can be performed safely in SSc, including the use of resin materials, amalgam, glass ionomer or dental crowns. These treatments must be performed quadrant by quadrant to avoid long sessions often poorly tolerated due to the limitation of mouth opening [16]. Prosthetic treatment may be difficult to perform for patients with severe limitations of mouth opening. Moreover, deceased salivary flow can cause a reduced tolerance to pressure generated by the prostheses. Dentures may be sectional or foldable to allow insertion and positioning into the mouth [17].

Few cases of implant rehabilitations for patients with systemic sclerosis are reported with short-term follow-up [18, 19]. Considering these single cases and in the absence of well-designed study dealing with success and survival rate of implant in systemic sclerosis patients, the decision to perform dental implant surgery must take into consideration the disease severity and its comorbidities particularly the macrosomia, the presence of sicca syndrome and the patient ability's to perform a good oral hygiene. It is assumed that dental implant may be useful in the anterior portion of the jaws to stabilize the dentures, thus improving comfort and decreasing the risks of traumatic ulcer [16].

CONCLUSION

The orofacial complications of SSc are often associated with aesthetic and functional concerns and may be the presenting symptoms of the disease. Systematic evaluation by the dentist and regular follow-up are required for early detection of dental caries and periodontal diseases to avoid invasive procedures often difficult to perform due to the limitation of mouth opening.

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