International Journal of Community Medicine and Public Health
Kaaki M. Int J Community Med Public Health. 2024 Nov;11(11):4508-4511
http://www.ijcmph.com
pISSN 2394-6032 | eISSN 2394-6040

DOI: https://dx.doi.org/10.18203/2394-6040.ijcmph20243317

Review Article

Oral and radiological features associated with systemic sclerosis: a literature review

Muhannad Kaaki*

Department of Oral and Maxillofacial Diagnostic Sciences, College of Dentistry, Taibah University, Medina, Saudi Arabia

Received: 07 October 2024
Accepted: 22 October 2024

***Correspondence:**
Dr. Muhannad Kaaki,
E-mail: mkaaki@taibahu.edu.sa

ABSTRACT

This review aimed to determine the orofacial radiological features associated with systemic sclerosis (SSc) characterises the oral health complications experienced by SSc patients and identifies their potential oral health care needs. SSc has several orofacial manifestations, including fibrosis of the overlying skin, limited mouth opening, a dry mouth, and swallowing difficulties. SSc patients also have an increased risk of periodontal disease and tooth decay. By outlining the orofacial radiological features and related complications, this review hopes to enhance the dental services provided to this population by enabling the prediction of their oral healthcare needs while calling for increased awareness among dental healthcare providers.

Keywords: Scleroderma, SSc, Oral health, Radiological features

INTRODUCTION

Radiological findings regarding the maxillofacial area arise mainly from the musculoskeletal structures in the pathological sclerosis of scleroderma and/or vascular ischemia, which may be caused by abnormal collagen deposition or the pressure effect from the tightening of the associated soft tissue. The review by Haers and Sailer, who analysed 22 publications with a total sample of 52 patients, reported an overall mandibular resorption prevalence of 50%, with an incidence of 20-30%. The study found that the resorption affected the mandibular angle in 37.6% of the cases, the coronoid process in 20%, the condylar head in 20.8%, the ascending ramus in 14.4%, and the bilateral condyles in 13.7% of cases.[1-5] Nonetheless, suggestion has been made that more severe sclerosis involves more severe mandibular erosion.[6]

ORAL RADIOGRAPHIC FEATURES

The most common oral radiographic feature is a widened periodontal ligament (PDL). A study involving 159 SSc patients found that 14.5% exhibited mandibular erosion, which is within the range mentioned in other reports (6.6-46.7%) (Table 1); moreover, 60.9% of the patients had multiple erosion sites in the mouth.[7]

Table 1: Radiographic oral findings of scleroderma.

Author and year	Sample size	Mandibular erosions	PDL widening
Dagenais, MacDonald (2015)[7]	159	14.47%	37.96%
Leung, Chu (2011)[8]	36	NR	Mean width of PDL was greater in SSc cases but not reported
Vincent, Agard (2010)[9]	30	6.67%	33.3%
Marcucci and Abdala (2009)[10]	15	46.7%	NR

Continued.

Author and year	Sample size	Mandibular erosions	PDL widening
Rout, Hamburger and Potts (1996)[11]	21	9.5%	33%
Wood and Lee (1988)[12]	24	29%	Mean width of PDL was greater in SSc cases but not reported
Janssens, Herman ([13]	47	NR	59%
Alexandridis and White (1984)[14]	26	NR	65%
Butts, Goetz (1977)[15]	35	8.6%	37%
Rowell and Hopper (1977)[16]	30	NR	70%
Marmary, Glaiss and Pisanty (1981)[17]	21	19%	100%
Seifert, Steigerwald and Cliff (1975)[1]	16	31%	NR

The resorption of zygomatic arches was found in SSc patients in association to the resorption of other bone parts, e.g., the mandibular angles and ascending ramus.[12,18] Other mandibular erosion patterns have also been reported, including the osseous resorption of the digastric region (2.53%) and the resorption of the posterior ramus (5.06%).[7] However, except for the rheumatoid factor, which was found to be positive in around 33% of SSc cases, the overall resorption pattern of the maxillofacial bones in SSc patients, particularly the bilateral bone resorption in mandibular angles, digastric region and both coronoids and condyles processes, might be due to the long-term (5-7 years) increase in the physical pressure from the overlying tight skin. Another reason may lie in the attachment sites of the masticatory muscles, such as the masseter, temporal, and lateral pterygoid muscles, as well as the anterior belly of the digastric muscles, which may impact the blood supply to the bone. Another explanation involves the disease's pathological process, which can cause ischemia due to vasculitis and perivascular fibrosis. This may be the case with the blood supply vessels of related bone structures, such as the maxillary artery to the condyle, the coronoid process, the mandibular angle, and the masseteric and pterygoid muscles.[5, 6, 19-21]

MRI FINDINGS ON MANDIBULAR OSTEOLYSIS

A previous study performed an MRI on 15 SSc patients to measure the masseter musculature in relation to mandibular osteolysis. It found a high ratio of fat replacement and muscular tissue atrophy linked to the disease.[4] A 20% incidence was reported for mandibular erosion in SSc, with an overall prevalence of 50%; meanwhile, 28.6% is considered mild degree of osteolysis. Notably, there is a suggestion that mandibular erosion is more severe when the sclerosis is severe.[6]

TEMPORO-MANDIBULAR JOINT INVOLVEMENT

Examining the temporomandibular joint (TMJ) involvement in SSc patients, MRI investigations revealed that 51.8% of patients (27; 12 diffuse and 15 limited) had scleroderma without displacement and 81.5% had disk displacement with reduction. The reported symptoms were TMJ clicking (63%), crepitation (70.4%), reduced mouth opening (66.7%), and TMJ and muscle tenderness (55.6%). Meanwhile, 55% reported pain lasting for at least 6 months.[22]

DEGENERATIVE BONE CHANGES

Degenerative bone changes were reported in 66.6% of SSc patients. The most reported change was a flattening of the condyle anterior surface (66.7%), followed by joint surface erosion and irregularities (55.6%), a flattening of the temporal eminence functional surface (48.1%), osteophytes (44.4%), synovitis (37%), subchondral cysts (11.1%) and idiopathic condyle resorption (11.1%). There was a significant correlation between the frequency of bone changes and the disease duration ($p<0.05$). However, re-adsorption theory may occur at different sites, including the mandibular angle, condyle and/or coronoid process due to atrophic ischemia; this may increase the risk of pathological fractures at the affected sites.[22]

ULTRASONOGRAPHIC FINDINGS

Ultrasonographic scanning was performed on 10 SSc patients to evaluate the extra-oral and intraoral changes under consideration of certain criteria. These were reduced facial expression range, impaired tongue movement, changes in lip redness, telangiectasia in the oral mucosa and facial skin, perioral folds, a flattened palate, a shortened uvula, myosclerosis in the mouth vestibule, decreased inter-incisal distance, and gingival inflammation. Two patients were found to have increased fibrotic deposition and sclerosis. Ultrasonography can be considered non-invasive investigating tool for monitoring disease activity and the effectiveness of the therapy. Colour Doppler ultrasound has also been reported to offer benefits in measuring dermal blood flow, which would provide information on underlying active disease.[4,23]

PDL WIDENING

Several recent studies have shown that the most common feature identified in oral radiography is PDL widening,

found in 38% of cases. A strong association was found between the number of teeth involved and disease severity. Notably, in SSc patients generalised collagen overproduction may cause PDL widening, although there is no clear correlation with other periodontal diseases as there is no significant loss of gingival attachment. Regardless, a significant relationship has been established between smoking, SSc severity, and PDL widening.[24]

Jung et al. found that PDL widening in SSc patients usually presents in both anterior and posterior teeth, with a greater tendency to occur in the posterior teeth.[25] Furthermore, PDL widening has been reported as a uniform widening in up to 10% of SSc patients[4], while periapical and panoramic radiographic investigations located PDL widening adjacent to the coronal and periapical areas of the roots in a study involving 163 SSc patients by using either periapical or panoramic radiograph.[7]

CALCIFICATION AND ROOT CANAL OBLITERATION

As previously indicated, radiography can reveal generalized PDL widening, and multiple maxillary teeth have been shown to highlight calcifications in the related PDL spaces. Additionally, any teeth that are impacted, specifically the premolars and incisors, have pulp stones and are vulnerable to root canal obliteration. However, such calcification has been associated with areas of PDL widening and the lamina dura remains intact. Such a clinical scenario may be derived from the disease's sclerotic features.[25]

A rare calcification of both a widened PDL and pulp canals was reported following cone-beam computerized tomography and panoramic radiography; this may be attributed to the disease's dystrophic calcinosis pattern. Another case report included a 13-year diagnosis of diffuse scleroderma with a history of persistent maxillary pain lasting a number of weeks; this case revealed the involvement of systemic organs, including lung, gastrointestinal and myocardial, and digital ulcers. A clinical examination found facial skin tightness, telangiectasia, thin and sclerotic lips and limited mouth opening. Generalized PDL widening was identified via radiological evaluation, and cone beam computed tomography (CBCT) showed that multiple maxillary teeth had calcifications in the related PDL spaces. Moreover, affected teeth such as the incisors and premolars presented pulp stones and were at risk of root canal obliteration. Here, calcification was correlated with widened PDL areas and an intact lamina dura. It should be noted, however, that sclerotic features of scleroderma may have contributed to this clinical scenario.[25]

EXTERNAL ROOT RESORPTION

There are infrequent cases involving external dental root abnormalities, such as resorption, dilaceration, and underdevelopment, linked to the affected area, especially in childhood cases (27.3%).[26] According to one study, a patient presented with intraoral features of SSc, including substantially limited mouth opening (20 mm) and telangiectasia on the hard palate and lateral tongue border. The radiological findings of left-side mandibular erosion coincided with the external distal root resorption of the lower left third molar. Nonetheless, the resorption of the apical third of the root was adjacent to the area of the mandible bone resorption. Once other causes had been excluded, it was proposed to be a result of the disease's erosive pathological process as well as the high external pressure from the adjacent muscular structure.[27] However, recent research has found that SSc patients may present with an external root resorption pattern located cervically rather than apically.[28]

CEPHALOMETRIC ANALYSIS AND SKELETAL MALOCCLUSION IN JUVENILE SSC

In a study of 16 patients with juvenile localised SSc, cephalometric analysis revealed a rare presentation in 9 patients (81.8%), namely an overgrowth of the lower third of the face with increased anterior face height. The authors also found skeletal malocclusion in 81.8% and TMJ asymmetry in 18.7%.[26]

CONCLUSION

SSc is a life-long debilitating disease with a wide range of systemic involvement, including changes to the oral and maxillofacial area. Understanding and identifying these changes can ensure the correct treatment, thereby alleviating the patient's discomfort. To this end, the changes to this region can be evaluated using a number of diagnostic and radiographic imaging modalities. Among the most striking radiographic features reported in SSc patients is the widening of the PDL, which is generally uniform and continuous with an intact lamina dura. Secondly, various bones in the oral and maxilla facial area can be subject to erosion. Often, the angle of the mandible produces an exaggerated antegonial notch. Other radiographic features reported via panoramic radiography, CBCT and cephalometric imaging include dystrophic calcification within the PDL and the presence of pulp stones, skeletal malocclusion, the resorption of the roots, jaw asymmetry, and sclerotic changes to the condyle.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Seifert MH, Steigerwald JC, Cliff MM. Bone resorption of the mandible in progressive systemic sclerosis. Arthritis Rheum. 1975;18(5):507-12.
2. Haers PE, Sailer HF. Mandibular resorption due to systemic sclerosis. Case report of surgical correction

of a secondary open bite deformity. Int J Oral Maxillofac Surg. 1995;24(4):261-7.
3. Doucet JC, Morrison AD. Bilateral mandibular condylysis from systemic sclerosis: case report of surgical correction with bilateral total temporomandibular joint replacement. Craniomaxillofac Trauma Reconstr. 2011;4(1):11-8.
4. Chapin R, Hant FN. Imaging of scleroderma. Rheum Dis Clin North Am. 2013;39(3):515-46.
5. Delantoni A, Matziari E. Osteolysis Affecting the Jaws in Systemic Sclerosis: Clinical and Osseous Changes Based on a Case Presentation. Clin Med Insights Arthritis Musculoskelet Disord. 2015;8:65-7.
6. MacIntosh RB, Shivapuja PK, Naqvi R. Scleroderma and the temporomandibular joint: reconstruction in 2 variants. J Oral Maxillofac Surg. 2015;73(6):1199-210.
7. Dagenais M, MacDonald D, Baron M, Marie H, Solène T, Russell S, et al. The Canadian Systemic Sclerosis Oral Health Study IV: oral radiographic manifestations in systemic sclerosis compared with the general population. Oral Surg Oral Med Oral Pathol Oral Radiol. 2015;120(2):104-11.
8. Leung WK, Chu CH, Mok MY, Yeung KW, Ng SK. Periodontal status of adults with systemic sclerosis: case-control study. J Periodontol. 2011;82(8):1140-5.
9. Vincent C, Agard C, Barbarot S, N'guyen JM, Planchon B, Durant C, et al. Orofacial manifestations of systemic sclerosis: A study of 30 consecutive patients. Rev Stomatol Chir Maxillofac. 2010;111(3):128-34.
10. Marcucci M, Abdala N. Clinical and radiographic study of orofacial alterations in patients with systemic sclerosis. Braz Oral Res. 2009;23(1):82-8.
11. Rout PG, Hamburger J, Potts AJ. Orofacial radiological manifestations of systemic sclerosis. Dentomaxillofac Radiol. 1996;25(4):193-6.
12. Wood RE, Lee P. Analysis of the oral manifestations of systemic sclerosis (scleroderma). Oral Surg Oral Med Oral Pathol. 1988;65(2):172-8.
13. Janssens X, Herman L, Mielants H, Verbruggen G, Veys EM. Disease manifestations of progressive systemic sclerosis: sensitivity and specificity. Clin Rheumatol. 1987;6(4):532-38.
14. Alexandridis C, White SC. Periodontal ligament changes in patients with progressive systemic sclerosis. Oral Surg Oral Med Oral Pathol. 1984;58(1):113-8.
15. Butts HC, Goetz JB, Spencer SH, Ross MD, Clements PJ, Furst DE, et al. Oral radiographic changes in patients with progressive systemic sclerosis (scleroderma). J Am Dent Assoc. 1977;94(6):1178-82.
16. Rowell NR, Hopper FE. The periodontal membrane in systemic sclerosis. Br J Dermatol. 1977;96(1):15-20.
17. Marmary Y, Glaiss R, Pisanty S. Scleroderma: oral manifestations. Oral Surg Oral Med Oral Pathol. 1981;52(1):32-7.
18. Hopper FE, Giles AD. Orofacial changes in systemic sclerosis--report of a case of resorption of mandibular angles and zygomatic arches. Br J Oral Surg. 1982;20(2):129-34.
19. Ramon Y, Samra H, Oberman M. Mandibular condylosis and apertognathia as presenting symptoms in progressive systemic sclerosis (scleroderma). Pattern of mandibular bony lesions and atrophy of masticatory muscles in PSS, presumably caused by affected muscular arteries. Oral Surg Oral Med Oral Pathol. 1987;63(3):269-74.
20. Jagger R, Absi E, Jeremiah H, Sugar A. Bilateral mandibular condylysis in a patient with systemic sclerosis. Dentomaxillofac Radiol. 2006;35(6):461-63.
21. Rahpeyma A, Zarch SH, Khajehahmadi S. Severe osteolysis of the mandibular angle and total condylolysis in progressive systemic sclerosis. Case Rep Dent. 2013;2013(1):948042.
22. Matarese G, Isola G, Alibrandi A, Alberto LG, Gianluca B, Giancarlo C, et al. Occlusal and MRI characterizations in systemic sclerosis patients: A prospective study from Southern Italian cohort. Joint Bone Spine. 2016;83(1):57-62.
23. Tolkachjov SN, Patel NG, Tollefson MM. Progressive hemifacial atrophy: a review. Orphanet J Rare Dis. 2015;10:1-13.
24. Baron M, Hudson M, Dagenais M, David M, Geneviève G, Tarek ES, et al. The Canadian Systemic Sclerosis Oral Health Study V: Relationship Between Disease Characteristics and Oral Radiologic Findings in Systemic Sclerosis. Arthritis Care Res. 2015;68(5):673-80.
25. Jung S, Minoux M, Maniere MC, Martin T, Schmittbuhl M. Previously undescribed pulpal and periodontal ligament calcifications in systemic sclerosis: a case report. Oral Surg Oral Med Oral Pathol Oral Radiol. 2013;115(4):e47-51.
26. Trainito S, Favero L, Martini G, Thomas KP, Vittorio F, Troels H, et al. Odontostomatologic involvement in juvenile localised scleroderma of the face. J Paediatr Child Health. 2012;48(7):572-76.
27. De Figueiredo MA, de Figueiredo JA, Porter S. Root resorption associated with mandibular bone erosion in a patient with scleroderma. J Endod. 2008;34(1):102-3.
28. Arroyo-Bote S, Bucchi C, Manzanares MC. External Cervical Resorption: A New Oral Manifestation of Systemic Sclerosis. J Endod. 2017;43(10):1740-43.

Cite this article as: Kaaki M. Oral and radiological features associated with systemic sclerosis: a literature review. Int J Community Med Public Health 2024;11:4508-11.