CASE REPORT

Early diagnosis of progressive systemic sclerosis (scleroderma) from a panoramic view: report of three cases

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Systemic sclerosis (scleroderma) is a multisystem disease involving the skin, lungs, heart, kidneys and gastrointestinal tract. It is probably owing to a disorder of the immune system. It may also have some oral manifestations, including limited ability to open the mouth, widening of the periodontal ligament (PDL) space and mandibular bone resorption. We report here on three asymptomatic cases that were referred to our oral and maxillofacial radiology centre for another reason, and in which we suspected scleroderma based on the widening of the PDL in the panoramic view. The early diagnosis was finally confirmed by pathological findings.

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Introduction

The term scleroderma originates from two Greek words, “sclero” meaning hard and “derma” meaning skin. Scleroderma is a multisystemic disorder involving the immune system that leads to swelling and then thickening of the connective tissue. As a result, skin, usually of the hands and feet, becomes stiff, tight and shiny.^{1,2} Most patients are adults and women are affected three times more often than men.¹ The disease is associated with very large depositions of dense collagen into the tissues of the body.¹

The disease can occur in three forms: (1) morphea (circumscribed scleroderma), characterized by local thickening of the skin; (2) generalized or progressive scleroderma (diffuse form), characterized by tautness of the skin with distinctive involvement of the lungs, heart, kidneys, and gastrointestinal tract, and osteolytic changes in the skeleton; and (3) acrosclerosis, a combination of scleroderma of the extremities and Raynaud’s disease.³

Oral manifestations of progressive systemic sclerosis (PSS) may include limited ability to open the mouth; xerostomia; periodontal disease; increased periodontal

ligament (PDL) width; and osseous resorption of the mandible.² Facial skin becomes thin and taut, which leads to a mask-like appearance.⁴ Calcinosis cutis, Raynaud’s phenomenon, oesophageal dysfunction, sclerodactyly and telangiectasia (CREST syndrome) is an uncommon condition that may be a relatively mild variant of systemic sclerosis.¹ As with systemic sclerosis, most patients with CREST syndrome are women in the sixth or seventh decade of life.¹ Raynaud’s phenomenon, resulting from digital arterial spasm, is often the earliest manifestation and may precede extensive skin and internal organ involvement by months or years.⁵

The most common oral radiographic manifestation of PSS, which occurs in about two-thirds of patients, is an increase in the width of the PDL around the teeth.⁶ Widening of the PDL affects both anterior and posterior teeth, although it is more pronounced around the posterior teeth. Despite the widening of the PDL space, the lamina dura remains normal, the teeth are often not mobile and their gingival attachments are usually intact.⁶

Histologically, diffuse deposition of dense collagen within and around the normal structures is seen. This abnormal collagen replaces and destroys normal tissue, leading to loss of normal tissue function.¹ The clinical sign of stiffened skin texture is suggestive of the

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diagnosis and a skin biopsy may confirm the diagnosis.¹ Several serological tests are also useful for confirmation of the diagnosis and for classification.⁷

Case 1

A 37-year-old woman was referred to our clinic for a panoramic radiograph owing to tooth pain. From the panoramic radiograph we noted widening of the PDL with intact lamina dura in the mesial root of the mandibular left first and second molars, and in the mesial root of the maxillary second molars (Figure 1a). This led us to suspect PSS, and we therefore looked for signs of mandibular bone resorption; however, we did not find any evidence of that (Figure 1b). The patient did not have any positive medical history or symptoms related to PSS or CREST syndrome, except for Raynaud's phenomenon. As she presented with a mask-like face (Figure 1c) and a slightly shiny and tense appearance of the hands (Figure 1d), we referred her to a dermatologist for further evaluation. A biopsy of the skin revealed hyperkeratosis in the epidermal layer and increased collagen fibres in the dermal layer (Figure 1e), consistent with scleroderma. After a 6-month period of treatment we did not find any further widening of the PDL in follow-up radiological studies.

Case 2

A 23-year-old woman had a panoramic radiograph taken owing to pain in the posterior teeth of the maxilla. In the panoramic radiograph, in addition to caries of the right maxillary second premolar, we found widening of the PDL with normal lamina dura in the mesial and distal roots of the mandibular first molars. Periapical radiographs confirmed this (Figure 2a,b). We did not find any bone resorption in the mandible. The patient had had no symptoms suggesting PSS or CREST syndrome in the past, and her face and hands appeared normal. Considering a possible diagnosis of PSS, we referred her to a dermatologist and eventually scleroderma was confirmed by skin biopsy.

Case 3

Generalized widening of the PDL with intact lamina dura was incidentally found in a panoramic radiograph of a 30-year-old woman who presented for another reason (Figure 3). There were no signs of any mandibular bone resorption. In addition, we did not find any signs or symptoms suggesting PSS in her history or physical examination, with the exception of Raynaud's phenomenon. Consultation with a dermatologist and subsequent pathological studies led to the diagnosis of scleroderma.

Discussion

Scleroderma has been described since the eighteenth century; however, its aetiology still remains unclear

and probably involves immunological mechanisms. Unfortunately, the course of the disease is unpredictable, and no definite treatment is available. However, in recent years considerable progress has been made in the treatment of this condition.^{8,5}

Scleroderma is a disease of middle age. Indeed, it occurs mostly in patients aged between 30 and 50 years and is 3 times more frequent in women than in men.⁶ All of our cases were women, and while one of them was less than 30 years old, the others were in their early 30s.

PSS has oral manifestations that include decreased opening of the mouth, microstomia, xerostomia, PDL widening and osseous resorption of the mandible.² In addition to skin stiffness, involvement of the temporomandibular joint further reduces the oral opening. Oral ulcerations owing to gastro-oesophageal reflux and subsequent infections are common.⁴ However, our patients did not exhibit any of these manifestations, except for widening of the PDL, probably owing to the early stage of the disease at the time of diagnosis.

Collagen deposition in the subcutaneous tissue of the facial skin results in the characteristic smooth, taut mask-like face.¹ Similarly, atrophy of the nasal alae leads to a pinched appearance of the nose, (sometimes called a "mouse face").¹ Only one of our patients appeared to have atrophy of the nasal alae as well as the mask-like face that indicated to us a diagnosis of scleroderma. The others did not show any facial signs.

Involvement of the hands in PSS includes resorption of the terminal phalanges (acro-osteolysis) and flexion contractures that produce shortened claw-like fingers.¹ None of our cases displayed this involvement except for one whose hands had a slightly shiny and tense appearance.

Raynaud's phenomenon is defined as a paroxysmal vasospasm that is induced by exposure to cold or emotional stress.⁷ Raynaud's phenomenon is usually the first manifestation in systemic sclerosis⁹ and occurs in more than 95% of PSS patients.⁷ None of our patients presented with features suggestive of either of these syndromes, although two showed signs of Raynaud's phenomenon. The patient who did not is probably among the 5% of PSS patients without this manifestation.

A radiographic feature in some cases of PSS includes typically bilateral, sharply demarcated and relatively symmetric mandibular erosions at regions of muscle attachment, such as the angles, coronoid process, digastric region or condyles. This resorption may be progressive in the course of the disease.⁶ Auluck pointed out that these erosions are caused by exerting pressure on the bone *via* the atrophic muscles at their attachment site. Atrophy of the muscles is attributable to increased fibrosis of the muscles and a decrease in the vascularity of the muscles, secondary to the fibrosis of muscular walls of the arteries.¹⁰ Other authors have mentioned that mandibular resorption can also be caused by ischaemia of the bone secondary to vasculitis

a

b

c

d

e

Figure 1 A 37-year-old woman with mask-like facial appearance. (a) Panoramic view: note the widening of periodontal ligament (PDL) in mesial root of mandibular left first and second molars, and also mesial root of maxillary second molars. (b) No bone resorption is evident. (c) Mask-like appearance and atrophy of the nasal alae. (d) Slightly tense and shiny appearance of hands. (e) Photomicrograph of skin biopsy showed sclerotic derm with dense collagen fibre

Figure 2 A 23-year-old woman with pain in right maxillary second premolar. (a) Panoramic view: note caries of the maxillary right second premolar and widening of the periodontal ligament (PDL) in mesial and distal roots of the first mandibular molars. (b) Periapical view demonstrates PDL widening in mesial and distal roots of the first mandibular molars

associated with PSS. In addition, the connective tissue abnormality associated with the disease can cause entrapment of the blood vessels.¹¹ Ramon et al⁸ suggested that the condyle, coronoid and mandibular angles, areas that show resorption, are supplied by small arterial branches of the internal maxillary artery rather than the main inferior alveolar artery. PSS may be more likely to affect these smaller arterial branches rather than larger arteries, thus leading to bone resorption secondary to diminished vascularity in those areas.⁸ Tightness of the skin, which may lead to pressure resorption of the bone, can be another factor in mandibular resorption.¹¹ Overall, it seems that bone resorption in systemic sclerosis has a multifactorial aetiology.¹²

However, mandibular resorption is relatively uncommon and has been reported in only 10% of the cases in some studies¹² and 7% in others.¹³ Although Marmary et al did not find any correlation between alterations in the jaw and the patient's age, duration of illness or

laboratory findings,¹⁴ Marcucci and Abdala mentioned that mandibular osteolysis seems to develop in patients with a longer duration of the disease.⁹ Neither of our cases had erosions or bone resorption in the mandible, which probably implies that PDL widening is one of the earliest manifestations of the disease and occurs before mandibular bone resorption.

PSS may also affect the PDL space by increasing its width. Stafne and Austin¹⁵ first described this finding and noted an incidence of about 7%. They mentioned that the posterior teeth are involved more frequently than the anterior teeth. Other authors have confirmed both this incidence and more involvement of the posterior teeth.^{16,17} However, White et al reported an incidence of more than 37%¹⁸ and Vincent and Agard mentioned an incidence of 33%.¹³ Marmary et al¹⁴ found that all their patients with PSS had higher average PDL widths than control patients.

Auluck suggested that PDL widening is caused by involvement of the masticatory muscle, which becomes

Figure 3 A 30-year-old woman. Panoramic view: generalized PDL widening in both jaws

more bulky and leads to an increase in masticatory occlusal forces that may result in primary trauma from occlusion. Hence, widening of the PDL in patients with PSS occurred without noticeable deposits of plaque or calculus.¹⁰ However, Mehra found that atrophy of the muscles decreases the forces of mastication and thus the chances of trauma from occlusion. In his opinion, it seems more likely that increased collagen synthesis, which leads to an increase in PDL bulk, explains the aetiology of PDL widening in these patients.¹⁹ Regarding the latter hypothesis, however, it is difficult to explain why PDL widening does not affect all patients with PSS and why it occurs more often in the posterior teeth in some patients. There was no significant clinical involvement of the masticatory muscles in our patients. A number of authors have reported the presence of PDL widening in only some of the teeth in certain scleroderma patients.¹⁴ In our study, one case had a widening of the PDL with an intact lamina dura in all teeth; but in other cases, this phenomenon was seen in only some teeth, especially the posterior teeth. As with cases reported by Marmary et al,¹⁴ our cases had PDL widening but did not have signs of jaw erosion.

Widening of the PDL may also occur in malignancies such as osteosarcoma and trauma from occlusion. In malignancy, the lamina dura does not remain intact, and PDL widening takes place in the teeth located around the

lesion. Conversely, the lamina dura remains intact in PSS, and PDL widening occurs in more than one quadrant and usually in the posterior teeth.⁶ In trauma from occlusion, PDL widening occurs but in association with angular bone defects and mobility of teeth, conditions that are not seen in patients with systemic sclerosis.¹⁹ However, some authors have suggested that PDL widening can be localized or generalized in the repair phase of trauma from occlusion and only in advanced stages will there be bone loss or tooth resorption.²⁰ Therefore, it may be the case that tooth mobility and bone loss do not always occur.²¹ Certainly, we found no angular bone defects or mobility of teeth in our patients.

Conclusion

We conclude that PDL widening may be one of the first radiographic signs of PSS, even in the absence of other manifestations, in the early stages of the disease. In view of the cases reported here, with unexplained PDL widening that was finally diagnosed with PSS, we recommend that a careful evaluation for PSS should be carried out in all patients showing widening of the PDL with an intact lamina dura, especially if located in the posterior teeth and in more than one quadrant.

References

1. Neville BW, Damm DD, Allen CM, Bouquot JE. *oral and maxillofacial pathology* (3rd edn). St Louis: Elsevier/Saunders, 2009, p 799.
2. Wood RE, Lee P. Analysis of the oral manifestations of systemic sclerosis (scleroderma). *Oral Surg Oral Med Oral Pathol* 1988; **65**: 172–178.
3. Alexandridis C, White SC. Periodontal ligament changes in patients with progressive systemic sclerosis. *Oral Surg Oral Med Oral Pathol* 1984; **58**: 113–118.
4. Benetti R, Zupi A, Toffanin A. Prosthetic rehabilitation for a patient with microstomia: A clinical report. *J Prosthet Dent* 2004; **92**: 322–327.

5. Fanci A, Braunwald E. *Harrisons Manual of Medicine*, 17th edition. New York: McGraw-Hill, 2009, p 167.
6. White SC, Pharoah MJ, editors. *Oral radiology—principles and interpretation*, 6th edition. Saint Louis: Mosby, 2009, p 467.
7. Chung L, Lin J, Furst E, Florentino D. Systemic and localized scleroderma. *Clin Dermatol* 2006; **24**: 374–392.
8. 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**: 269–274.
9. Marcucci M, Abdala N. Clinical and radiographic study of orofacial alterations in patient with systemic sclerosis. *Braz Oral Res* 2009; **23**: 82–88.
10. Auluck A. Widening of periodontal ligament space and mandibular resorption in patients with systemic sclerosis. *Dentomaxillofac Radiol* 2007; **36**: 441–442.
11. Pogrel MA. Unilateral osteolysis of the mandibular angle and coronoid process in scleroderma. *Int J Oral Maxillofac Surg* 1988; **17**: 155–156.
12. Auluck A, Pai KM, Shetty C, Sheno SD. Mandibular resorption in progressive systemic sclerosis: a report of three cases. *Dentomaxillo Facradiol* 2005; **34**: 384–386.
13. 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. *La Revue de Medecine Interne* 2009; **30**: 5–11.
14. Marmary Y, Glaiss R, Pisanty S. Scleroderma: oral manifestations. *Oral Surg Oral Med Oral Pathol* 1981; **52**: 32–37.
15. Stafne EC, Austin LT. A characteristic dental finding in acrosclerosis and diffuse scleroderma. *Am J Orthod Oral Surg* 1944; **30**: 25.
16. Rowell NR, Hopper FE. The periodontal membrane in systemic sclerosis. *Br J Dermatol* 1977; **96**: 15–20.
17. Vareltzidou-Anagnou A, Mitsis FL. Scleroderma and periodontal lesions: a clinical study of 18 cases. *Odontostomatol Proodos* 1978; **32**: 115.
18. White SC, Frey NW, Blaschke DD, Ross MD, Clements PJ, Furst DE, et al. Oral radiographic changes in patients with progressive systemic sclerosis (scleroderma). *J Am Dent Assoc* 1977; **94**: 1178.
19. Mehra A. Periodontal space widening in patients with systemic sclerosis: a probable explanation. *Dentomaxillofac Radiol* 2008; **37**: 183–184.
20. Rout PG, Hamburger J, Potts AJ. Orofacial radiological manifestations of systemic sclerosis. *Dentomaxillofac Radiol* 1996; **25**: 193–196.
21. Auluck A. Letter to the editor. *Dentomaxillofac Radiol* 2008; **37**: 184.