Previously undescribed pulpal and periodontal ligament calcifications in systemic sclerosis: a case report

Sophie Jung, DD,[a,g,h] Maryline Minoux, DD, PhD,[b,g,i] Marie-Cécile Manière, DD, PhD,[c,e,g] Thierry Martin, MD, PhD,[f,h] and Matthieu Schmittbuhl, DD, PhD[d,e,g,j]
University Hospital and University of Strasbourg, Strasbourg, France

Systemic sclerosis (SSc), a multisystem autoimmune disease characterized by widespread fibrosis, vascular alterations, autoimmunity, and inflammation, has effects on the hard and soft tissues of the orofacial region. The most common oral radiographic features correspond to widening of the periodontal ligament space and to mandibular resorption. In this report, cone-beam computerized tomography (CBCT) confirmed not only the well described periodontal features associated with SSc but also revealed previously undescribed calcifications within the periodontal ligament space of most maxillary teeth. Moreover, CBCT showed pulp calcifications in some incisors and premolars with these calcifications leading to root canal obliterations. Such manifestations (which could be linked to different major pathogenic features of SSc such as calcinosis, vasculopathy, and fibrosis) contribute to the phenotypic spectrum of the disease. (Oral Surg Oral Med Oral Pathol Oral Radiol 2013;115:e47-e51)

Systemic sclerosis (SSc) is a relatively rare multisystem connective-tissue disorder. This autoimmune disease is characterized by excessive production and deposition of collagen within the skin and internal organs associated with vascular abnormalities.[1] The adult annual incidence rate of SSc ranges from 0.4 to 2.4 per 100,000.[2] SSc preferentially affects women with a female-male ratio of ~7:1.[3] The peak age of onset is between 45 and 65 years, but any age group can be affected.[4] Progression and severity of the disease are variable and make prognosis uncertain. According to the extent of skin fibrosis and the pattern of organ-system involvement, SSc is classified into limited cutaneous SSc or diffuse cutaneous SSc.[4] Although the precise etiology of SSc has not been established, autoimmunity plays a pivotal role in its pathogenesis as evidenced by the presence of multiple SSc-specific autoantibodies.[1] Anticentromere antibodies are associated with limited forms of scleroderma, whereas anti-topoisomerase I antibodies (anti-Scl 70) are more often found in diffuse SSc.[1,3,5]

Patients with SSs may present with a wide array of clinical features, such as Raynaud phenomenon (which is often the earliest manifestation), skin thickening, esophageal dysmotility, pulmonary hypertension, arthralgia, and renal insufficiency.[4] Dystrophic calcinosis is a common finding in SSc, affecting ~25% of patients.[6,7] Calcified deposits are found not only in the skin and subcutaneous tissues but also in the periarticular regions, and they may preferentially occur at anatomic sites where tissue integrity has been compromised.[8]

The orofacial region is frequently involved in SSc. The clinical manifestations are well described and include skin and oral mucosa atrophy, limited mouth opening, mandibular resorption, and periodontal ligament space (PLS) widening.[5,9-18]

In the present report, a rare case of SSc exhibiting unusual dentoalveolar calcifications is presented and previously undescribed features of SSC are assessed relative to the literature.

CASE REPORT

The department of Internal Medicine and Clinical Immunology referred a 33-year-old white woman with the diagnosis of diffuse SSc to the National Reference Center for Dental Manifestations of Rare Diseases. She complained of recurrent diffuse maxillary pain that had persisted for several weeks.

The diagnosis of SSc was made 13 years earlier, ~2 years after the first signs of the disease (skin fibrosis and Raynaud phenomenon) had appeared. The patient exhibited severe skin involvement, with a current modified Rodnan Skin Score of 21/51 that had reached its highest level (37/51) 7 years before. Her medical history revealed that she suffered from polyar-

[a]Department of Oral Medicine and Oral Surgery, University Hospital.
[b]Department of Operative Dentistry, University Hospital.
[c]Department of Pediatric Dentistry, University Hospital.
[d]Department of Dentomaxillofacial Radiology, University Hospital.
[e]National French Reference Center for Dental Manifestations of Rare Diseases, University Hospital.
[f]Department of Internal Medicine and Clinical Immunology and National Referral Center for Systemic Autoimmune Diseases, University Hospital and Faculty of Medicine, University of Strasbourg.
[g]Faculty of Dentistry, University of Strasbourg.
[h]Research Team CNRS UPR 9021, Molecular and Cellular Biology Institute (IBMC), Strasbourg.
[i]Friedrich Miescher Institute for Biomedical Research, Basel, Switzerland.
[j]Research Team INSERM UMR 977, Strasbourg.
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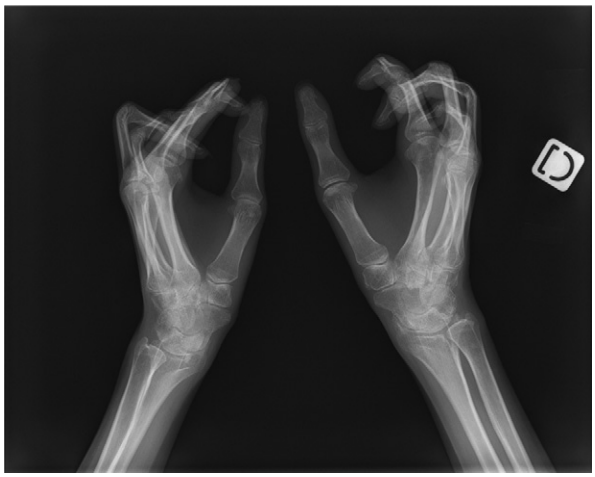

Fig. 1. Radiograph of the hands, showing the flexion contracture and claw-like deformity due to hand arthritis associated with sclerodactyly.

thralgia and sclerodactyly complicated by digital ulcers and moderate lung fibrosis with no evidence of pulmonary arterial hypertension. Anamnesis also revealed myocardial and gastrointestinal involvement manifested by gastroesophageal reflux and small bowel malabsorption. Hand arthritis associated with sclerodactyly led to flexion contracture and a claw-like deformity (Figure 1). Laboratory examination was positive for antitopoisomerase I (Scl-70) antibodies. Her medications included calcium channel blockers and an antiplatelet drug for treatment of Raynaud phenomenon, alginic acid for gastroesophageal reflux, and paracetamol/tramadol for analgesia. No immunosuppressive therapy was administrated.

Clinical examination revealed several common orofacial features of SSc. The facial skin was smooth and tight, with a loss of normal animation lines resulting in a mask-like facies. Multiple telangiectasias were seen on the face and the neck. The nasal alae were atrophied leading to a pinched appearance of the nose, and the lips were thin and sclerotic. The patient had microstomia with a maximal incisal opening of 20 mm and marked limitation of mandibular movements.

Intraoral examination revealed tongue rigidity and pale, sclerotic, dry mucosa with several telangiectasias. Two premolars (25 and 44) had been previously extracted. Despite the hand deformity and limited oral opening, her oral hygiene was good. The periodontal examination revealed no periodontal pocket nor tooth mobility.

A panoramic radiograph revealed a generalized PLS widening associated with an intact lamina dura (Figure 2). A carious lesion involving the pulp was diagnosed on the maxillary right second molar. To check for periapical inflammatory lesions, a cone-beam computerized tomography (CBCT; NewTom 5G; QR, Verona, Italy) examination of the maxilla was performed. The CBCT images revealed irregular densely calcified islets of various sizes in the PLS of most maxillary teeth. These calcifications were most conspicuous in teeth with enlarged periodontal ligaments (Figure 3, *B*), and were not visible on a periapical radiograph of upper right premolars and molars (Figure 3, *A*). The CBCT images also contained

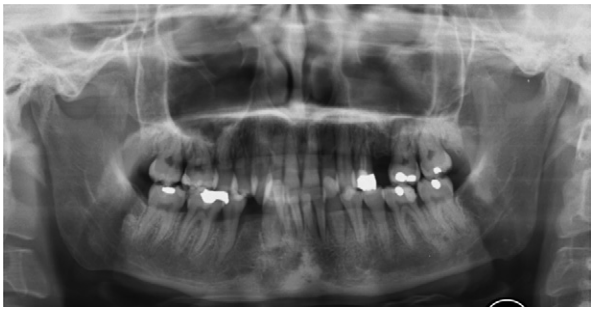

Fig. 2. Panoramic radiograph revealing generalized periodontal ligament space widening as well as pulp obliterations involving incisors and premolars.

pulp calcifications involving incisors and premolars, with these leading to complete root canal obliterations (Figure 3, *B*). No sign of root resorption or osteolysis of the mandibular angles, condyles, coronoid processes, or posterior borders of the ascending rami was observed. No other radiographic calcification was identified at the sites frequently involved in dystrophic calcinosis (forearms, elbows, and fingers).

Given limited oral opening, endodontic treatment was not feasible on the decayed tooth. A pulpotomy was performed with Biodentine (Septodont, Saint-Maur-des-Fossés, France) and resulted in dental pain relief.

In compliance with the Helsinki Declaration, written informed consent was obtained from the patient for publication of this report and accompanying images.

DISCUSSION

Orofacial features are found in ~80% of SSc patients.[12] Three pathologic processes contribute to tissue damage: vasculopathy, fibrosis, and inflammation/autoimmunity.[19,20] The effects of the disease on the facial region are mainly the result of collagen deposition in the subcutaneous tissue of the skin, facial muscle atrophy, and skin fixation to the underlying structures. Hardening and tightening of the skin induce changes in the face, and these result in a characteristic mask-like facies. Fibrosis of the lips causes reduced mouth opening. Limited oral opening impairs oral hygiene and makes dental procedures and intraoral radiographs difficult.[12,14,21] Nasal alae are frequently atrophied resulting in a mouse-like facies and a nose that appears pinched.[21] Oral mucosa often has telangiectasias and is pale, taut, and atrophic with a loss of elasticity.[21] Another usual oral manifestation is fibrosis of the salivary glands, resulting in xerostomia.[22] Resorptions of the mandibular angles, posterior borders of the ascending rami, coronoid processes, condyles, and zygomatic arches have also been associated with SSc[11-13,21,23] and can be due to excessive pressure exerted by hardened overlying tissues.[10]

The most common dental radiographic finding (which occurs in more than one-third of patients with SSc) is PLS widening.[5,9] The present radiologic examination revealed