Progressive systemic sclerosis in a child: case report
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Abstract
A case of pediatric progressive systemic sclerosis is reported and a literature review concerning medical and dental aspects of this condition is provided. Systemic features include sclerodactyly, Raynaud's phenomenon, telangiectasia, calcinosis, myositis, arthritis, tenosynovitis, renal failure, esophageal hypomotility, pulmonary fibrosis and heart failure. Oral manifestations include reduced interincisal distance, xerostomia, telangiectasia, increased periodontal ligament width, osseous resorption of the mandible, periodontal disease, and increased decayed, missing, and filled teeth (DMFT). The prognosis is difficult to predict because spontaneous remission has been documented, but death may result from extensive visceral involvement (heart, kidney, and lung). (Pediatr Dent 15:275-79, 1993)

Introduction
Progressive systemic sclerosis (PSS) is a rare connective tissue disease of unknown etiology characterized by increased collagen deposition leading to fibrosis and subsequent degeneration of the skin and internal organs. Approximately 12 new cases of PSS per million of population are diagnosed in the United States each year, with females being affected three to four times more frequently than males. There is no racial predilection. It most commonly occurs between 30 and 50 years of age. PSS in children is very rare. One and one-half per cent of all patients diagnosed are younger than 10 years old, and 7% of cases occur in patients between 10 and 19 years old. Childhood PSS also has a female predominance of more than 75% and usually begins before puberty. There is considerable variation in both the rate of progression and clinical severity of this disease. The most prominent clinical observation is thickened, hidebound skin—especially around the fingers and hands (sclerodactyly). Other features of the disease include: Raynaud's phenomenon, telangiectasia, calcinosis, myositis, arthritis, tenosynovitis, renal failure, esophageal hypomotility, pulmonary fibrosis, and heart failure. A more localized variant of PSS is termed the CREST syndrome (calcinosis, Raynaud’s phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia).

The precise etiology of PSS is unknown, however several factors seem to play a key role. Immunologic studies suggest that the pathogenesis of the disease may be an autoimmune response directed against endothelium. Furthermore, the early stages of the disease also may be characterized by the presence of an inflammatory cell infiltrate in the dermis. Others have shown coagulation abnormalities that result in thrombosis, occlusion, and extensive ischemic tissue damage. Fibroblast tissue culture studies have confirmed increased secretion of collagen proteins. In vitro studies showed that increased collagen production is due to an overexpression at the transcriptional level. The fibroblasts in these patients exhibit growth abnormality.

Information on oral aspects of pediatric scleroderma is limited in the dental literature. Hoggins and Hamilton reported a case of a 4-year-old white female suffering from localized scleroderma involving the face, chin, and ear that caused facial asymmetry. Rabey reported a case of a 14-year-old white male with proptosis, tightly stretched skin around the face and neck, upper teeth revealed due to an inability to close his mouth, and loss of mandibular condyles bilaterally. The only paper published in a pediatric dental journal on pediatric scleroderma was by Anderson and Ewell-Jackson.

On the other hand, numerous studies on adult patients have been published. The oral manifestations of PSS in adult patients include impaired mouth opening with reduced interincisal distance, xerostomia, telangiectasia, increased periodontal ligament width, osseous resorption of the mandible, periodontal disease, and increased decayed, missing, and filled teeth (DMFT). Taveras reported the first documented case of osteolysis of the mandibular angle and condyle in adult patients with scleroderma. There has been only one reported case of a child suffering from PSS with bilateral resorption of the mandibular condyles. The etiology of the osteolysis is unknown but there are three proposed theories.

1. Tightening of the facial skin may exert excessive pressure on the mandible and induce the bone loss.
2. The vasculopathy associated with this disease may diminish the blood supply to the mandible resulting in ischemia and bone necrosis.
3. Atrophy of the muscles of mastication such as the masseter muscle may lead to bone resorption. It is interesting to note that all bone resorption in these
patients occurs in areas of muscle attachment.

Oral radiographic examination of these adult patients often reveals generalized enlargement of the periodontal ligament space. Widening of periodontal ligament space also may be observed in traumatic loosening of teeth, periodontal inflammation, diabetes mellitus, cystinosis, osteomyelitis, and even malignant tumors. Studies have shown that this finding in PSS patients is not due to extrusion of teeth, the presence of periodontal inflammation, or resorption of roots. Rather, the enlarged periodontal space is caused by excess deposition of collagen and oxytalan fibers and subsequent resorption of alveolar crest bone surrounding the roots. Stafne and Austin were the first to report involvement of the periodontal membrane in 9 of 127 (7.1%) adults with scleroderma. Subsequent studies have shown the percentage of scleroderma patients with periodontal ligament involvement to be greater. Wood and Lee reported an increased periodontal membrane width in adult patients with PSS involving all groups of teeth. White et al. documented thickened periodontal ligament space in 13 of 35 (37%) adult patients. Posterior teeth were affected more frequently than anterior teeth. Alexandridis and White reported similar findings, but with a much higher prevalence of thickened periodontal ligaments (65%). Marmary et al. found increased periodontal ligament width in all of the patients in their study with a greater degree of involvement in the posterior teeth.

The oral mucosa may undergo changes similar to those found in the skin. Mild edema followed by gradual atrophy and induration of the mucosal tissues may be present in some scleroderma patients. Also, the color of the mucosa may change to a more whitish tinge. About 26% of patients may develop lingual and buccal mucosal crenation and 16% may develop focal gingival recession. The gingival recession can be attributed to fibrotic stricture along the mandibular mucobuccal fold, thus causing stripping of the attached buccal gingiva adjacent to the lateral incisor, canine, or premolar.

Due to the various forms and extent of systemic involvement, the prognosis in scleroderma is difficult to predict. There have been documented cases of spontaneous remission in children but the lowest survival rate was found in older patients, especially in males, even though the disease has a female predominance. In a study of 237 adult patients with PSS over 11 years, the mortality rate was 25.7%. The overall 3-, 6-, and 9-year survival rates of PSS were 86, 76, and 61% respectively. Pulmonary manifestation was the most frequent cause of death. However, adverse prognostic factors include renal, cardiac, and pulmonary involvement in decreasing order of importance. There have been no studies detailing the exact nature of changes in child patients owing to the paucity of cases at any one center. The purpose of this paper is to relate the findings, both medical and dental, of a child afflicted with PSS.

Case report

A 9-year-old white female complained of pain in her hands while washing dishes. She subsequently developed ulceration of the fingertips and Raynaud's phenomenon in her hands; a diagnosis of scleroderma was made in March 1984.

When seen in the Rheumatology Clinic in May 1984, she had thickening and tightening of the skin around her fingers, hands, forearms, lower limbs, and feet. The face and trunk were spared. She had flexion deformities of all her fingers with finger tip pulp space wasting and pitting scars at the tips. She was started on penicillamine at (375 mg/day).

Over the next several months, she had progressive skin tightening with both hyper- and hypopigmentation, new ulcers on her hands, and boney prominences. In addition, she developed polyarthritis involving her ankles, knees, hips, and elbows along with weight loss. Her face and trunk remained normal. On maximal opening of her mouth, the interincisal distance was reduced to 34 mm.

Due to the continued progression of the disease, she was hospitalized in November 1984. She complained of shortness of breath upon exertion and noticed decreased strength. She was having difficulty dressing and holding onto objects because of the flexion deformities of her fingers. At this point skin thickening was more extensive, involving her face, chest, and abdomen as well as the distal extremities. Her interdental distance on mouth opening had further decreased to 28 mm. She had 90° flexion deformities of all the proximal interphalangeal joints of her hands and pitting scars at the tips of all 10 digits (Fig 1).

Skin ulceration was present over the dorsum of several fingers, both elbows, and medial malleoli. She had 14 active joints with stress pain of both shoulders, elbows, wrists, hips, knees, and ankles. Tendon crepitu was present at the wrists, knees and ankles. Her chest radiograph was normal. Pulmonary function tests showed a restrictive defect, with a total lung capacity of 59% of predicted norm and carbon monoxide diffusing capacity of 60% of pre-

Fig 1. Clinical photograph demonstrating marked skin involvement, flexion deformities, and ulceration of both hands.
dicted norm. Skin biopsy showed an increase in dermal and subdermal collagen, but no inflammatory infiltrate. Muscle biopsy was normal. While in the hospital, a three-day IV infusion of prostaglandin $E_2$ was administered, after which the ulcers on the hands healed gradually. Despite the penicillamine, her condition continued to progress rapidly and a decision was made to treat her with cyclosporin on an experimental basis.

The patient presented to the dental clinic January 1985 for a dental examination. Extraoral examination revealed smooth, tightened skin around her face and signs of telangiectasia on her cheeks. Intraoral examination proved to be difficult since her interincisal distance was reduced to 23 mm. She was in mixed dentition stage with the right and left maxillary deciduous canine, first and second molar, the right deciduous second molar, and the left deciduous first and second molar still present. These deciduous teeth were not clinically mobile. Oral hygiene was well maintained and there was little plaque or calculus present. A radiographic intraoral series was exposed that showed enlarged periodontal ligament spaces in both permanent and primary teeth (Fig 2). The first permanent molars, the mandibular permanent central incisors, and the mesial root of the partially resorbed mandibular right second deciduous molar all exhibited thickening of the periodontal ligament space. Increased lamina dura width was found in many of the other teeth. Unfortunately, the patient had not had prior dental radiographs that would definitively prove the existence of widened periodontal ligament spaces on primary teeth. Transpharyngeal radiographs showed no sign of osseous resorption of the mandibular condyles, coronoid processes, ramus, or body of the mandible.

In September 1985, a recall examination showed that her condition had stabilized with no new areas of skin involvement or active digital ulcers. Fixed flexion deformities persisted in the joints of the fingers and both elbows and with limited range of motion at the wrists, shoulders, hips, and knees.

She was hospitalized again in January 1986. After stopping cyclosporin, she had become wheelchair bound because of progressive skin changes with flexion contractures of the knees and hips. Head and neck examination revealed an interincisal distance of 20 mm. The mucous membranes appeared normal. Skin examination showed progression of the disease over her arms and chest. Musculoskeletal examination revealed marked reduction in range of movement of the elbows, wrists, shoulders, and especially the hips and knees. She was healed with aggressive physiotherapy and splinting. Cyclosporin therapy was recommenced.

Her annual recall examination in April 1987 showed remarkable improvement. The previously noted dyspnea had resolved. She had gained 8.5 kg, was no longer confined to a wheelchair, and the flexion deformities of her knees and elbows had lessened considerably. Examination showed some resolution of the skin changes on the arms, forearms, hands, and feet. Incisal distance was 25 mm. Radiographic examination revealed soft tissue calcification and mild terminal tuft resorption in both index fingers (Fig 3).

The patient returned to the Dental Clinic for a recall examination in January 1989. Nothing new was noted during the intra- and extraoral examination except that her interincisal distance had improved to 26 mm from the previous evaluation. She remained caries free despite her difficulty in gripping a toothbrush. Posterior intraoral radiographs showed that the enlarged periodontal ligament space had persisted around the first molar teeth.

The patient is now 18 years old and coping well, despite severe residual joint deformities—especially of the hands. Despite discontinuation of cyclosporin, the scleroderma appears to be in remission.

Discussion

Comparing this case report with other literature is difficult because of the limited information available in the
pediatric dental literature. The two female patients reported by Anderson and Ewell-Jackson were adult patients who suffered from PSS in childhood so it would be inappropriate to draw comparisons to this patient.12 This patient exhibited reduced interincisal distance and increased periodontal ligament width, but no oral mucosal lesions or resorption of the mandible were evident. The degree of interincisal distance reduction progressed as the disease progressed and the patient never fully recovered the ability to open her mouth. The changes in interincisal distance mirrored the course of the disease although this is of limited value in a single case report.

Radiographically, this patient's permanent posterior teeth exhibited widening of the periodontal ligament (Fig 2). No studies have examined the natural history of the periodontal involvement, but a case has been reported in the dental literature that suggests that widening of the ligament space may be reversible.21 Even if this periodontal condition is permanent, these teeth do not exhibit signs of mobility27 and therefore require no intervention. This case represents the first report of widening of the periodontal ligament space and lamina dura in the primary dentition. Although the right mandibular primary second molar was resorbed partially by the erupted premolar, the primary tooth had an enlarged periodontal ligament and was not clinically mobile.

These patients often suffer from severe flexion deformities of their fingers and other body joints and thus have reduced manual dexterity. Maintenance of oral hygiene may be difficult due to the patient's inability to grip and manipulate a toothbrush. A larger-handed toothbrush or flossing aids may allow easier grasping and facilitate maintenance of good oral hygiene.20 This patient managed to keep her oral hygiene level quite high despite serious digital abnormalities.

Intraoral examination and intraoral procedures such as exposure of intraoral radiographs and extractions are difficult to perform due to tightening of skin, reduced interincisal distance, and diminished oral aperture. Surgical intervention such as bilateral commissurotomies can correct the microstomia.27 However a nonsurgical alternative was proposed by Naylor et al.,26,28 which involves cheek and mouth stretching exercises and tongue depressor exercises to increase interincisal distance to facilitate oral hygiene, mastication, and dental access. These exercises were suggested to the patient but she neglected to follow through with them.

Another factor that may be important to the practicing dentist is the extensive visceral involvement. Heart, lung, kidney, and GI involvement may complicate dental procedures. Consultation with the patient's rheumatologist or internist is suggested prior to engaging in any invasive dental procedure.

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Vaccine experts formulate new standards for child immunizations

Guidelines represent key element in national strategy for vaccinations

A recent resurgence of measles in the United States prompted the creation of new standards for pediatric immunizations, published in this week's Journal of the American Medical Association.

According to the Centers for Disease Control and Prevention (CDC), there were 55,622 reported cases of measles in the United States between 1989 and 1991, resulting in 11,251 hospitalizations and 166 suspected deaths. The main cause of the epidemic was failure to vaccinate children at the recommended ages, 12 to 15 months.

In an effort to stop the spread of measles, the CDC and a 35-member working group representing 22 public and private agencies developed Standards for Pediatric Immunization Practices. The standards are being encouraged as a key element in the national strategy to administer vaccines more efficiently and effectively to the nation's children.

Key points of the 18 standards include:

- Immunization services should be responsive to the needs of patients.
- Immunization services should be available on a walk-in basis at all times for both routine and new-enrollee visits. Waiting time should be minimized and generally not exceed 30 minutes.
- In the public sector, immunizations should be free of charge. If fees must be collected, they should be kept to a minimum.
- Each encounter with a health care provider, including an emergency department visit or hospitalization, is an opportunity to screen immunization status and, if indicated, administer needed vaccines.
- Providers should educate parents and guardians in a culturally sensitive way, preferably in their own language, about the importance of immunizations.
- Public providers should routinely seek the input of their patients on specific approaches to better serve their immunization needs and implement the changes necessary to provide more user-friendly services.
- Only properly trained individuals should administer vaccines.