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Localized scleroderma in childhood: review of the literature and case report

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Abstract

Although the dental manifestations of the rare dermatologic disorder generalized scleroderma have been well described in adults, the oral manifestations of localized scleroderma in childhood are not well documented. A case of localized scleroderma in a female child is described which highlights the problems of dental management of the defects that interfere with dental and alveolar development.

Literature Review

Scleroderma, a relatively uncommon collagen disease of unknown etiology, was recognized more than 2 centuries ago (Capusan 1972). It is characterized by induration and fixation of the skin to the deeper subcutaneous tissues. It may begin in children and young adults with females being twice as frequently affected as males (Weisman and Calcaterra 1978).

The disease may occur in both the localized and systemic forms. In the localized form, also known as morphea, the changes are confined to skin and subcutaneous tissue with occasional involvement of deeper tissues (Kornreich et al. 1977). The cutaneous lesions are described as white or yellowish plaques, slightly elevated or depressed, and may occur as linear bands or ribbons, often termed *en coup de sabre* since they resemble the marks produced by a sabre sword (Shafer et al. 1983). The systemic form of the disease is characterized by diffuse fibrosis of the skin and certain internal organs, primarily the blood vessels, gastrointestinal tract, lungs, heart, and kidneys (Eversole et al. 1974; Shafer et al. 1983). The chief manifestations include thickening and induration of the skin, digital pitting, sclerodactyly, pigmentations, telangiectasia, and Raynaud's phenomenon.[1]

The oral manifestations of scleroderma include fibrosis and rigidity of facial skin, tongue, soft palate, larynx, salivary glands, and buccal mucous membrane[2] leading to microstomia, dysarthria, dysphagia, and xerostomia. Periodontal manifestations such as loss of attached gingiva and multiple foci of recession have been reported (Eversole et al. 1974). It is suggested that the external pressure created by the skin changes may result in resorption of the alveolar bone, tilting of the teeth, delayed tooth eruption and early exfoliation of the teeth (Looby and Burket 1942).

Radiographic signs include characteristic widening of the periodontal ligament space without increased tooth mobility and resorption of the coronoid process, condyle, and posterior aspect of the ascending ramus of the mandible.[3] These changes in the mandible and TMJ cause impaired growth and development as well as functional problems such as subluxation and abnormal motion (Nandakumar et al. 1983).

Although scleroderma usually is classified into clinical categories of localized and systemic forms, this distinction is not always clear-cut. Several authors now have suggested that localized cutaneous scleroderma in the form of morphea or linear bands does not preclude the development of progressive systemic sclerosis.[4]

The oral signs of scleroderma have been described mainly in adults. Although there are several reports on childhood scleroderma in the medical literature, only minimal information is available on the oral aspects. The first case report on the dental manifestations of localized childhood scleroderma was presented by Looby and Burket in 1942. Two decades later, Hoggins and Hamilton (1969) described a similar case. This paper presents a patient with localized scleroderma, emphasizing the early features and dental management of this rare dermatologic disorder.

Clinical Report

A Caucasian female patient 5.5 years of age was referred to the University Dental School by her derma-

[1] Kass et al. 1966; Dabich et al. 1974; Eversole et al. 1974; Kornreich et al. 1977; Shafer et al. 1983.

[2] Eversole et al. 1974; Weisman and Calcaterra 1978; Marmary et al. 1981; Naylor 1982.

[3] Seifert et al. 1975; White et al. 1977; Marmary et al. 1981; Nandakumar et al. 1983.

[4] Rodnan and Fennell 1962; Kass et al. 1966; Kornreich et al. 1977; Masi et al. 1980; Kuto et al. 1985.

tologist who had diagnosed a condition of localized scleroderma after biopsy of a skin lesion on her cheek.

She was of normal intelligence, with height 113 cm (50th percentile) and weight 17.0 kg (25th percentile). A linear, flat, white scar-like lesion, approximately 0.5 cm wide, extended from the bridge of the nose through the right ala and the right side of the philtrum of the lip (Fig 1). A 2-cm white, non-raised, indurated plaque with superficial hemorrhagic crusting (which was the site of the biopsy, Fig 2) was noted on her right cheek just below the malar eminence. No other skin lesions were apparent and no developmental dysmorphological features were noted.

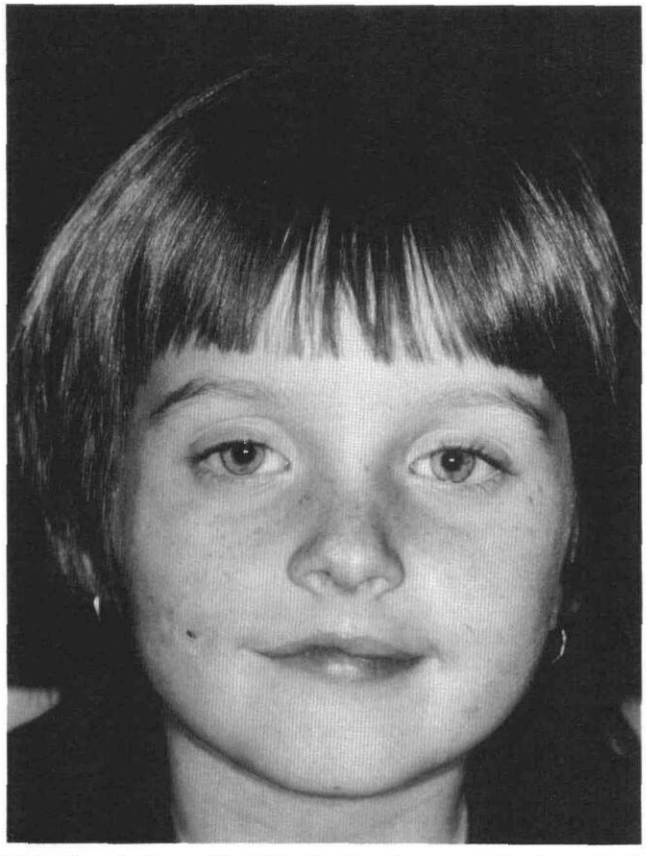

FIG 1. Two lesions of localized scleroderma on the face of a 6-year-old girl. A *coup de sabre* linear scar extends through the right ala and philtrum and a plaque is present on the right cheek.

Medical History

The patient was the product of a normal full-term pregnancy of a nonconsanguinous marriage. She weighed 3.1 kg at birth. Apart from mild jaundice at birth and occasional asthma which was controlled by Ventolin® syrup, her medical history was insignificant.

The skin lesions and dental problems appeared to date from about 2 years previously when she sustained a fall, striking and loosening the root of her maxillary right primary central incisor. She was taken to a medical general practitioner who in turn referred her to a dermatologist. A punch biopsy of skin was performed and diagnosis of scleroderma was confirmed by histopathology.

Tests to exclude systemic involvement of the disease such as anticentromere antibody were negative. However, blood tests revealed raised erythrocyte sedimentation rate as well as increased neutrophil and eosinophil counts. Antinuclear antibody tests were done to exclude systemic lupus erythematosus and these were negative. As there appeared to be no systemic involvement, no medical treatment was given at this stage.

Dental Manifestations

The patient's dental problems first were noted by her general dental practitioner when, after her fall, he observed that the maxillary right primary central incisor

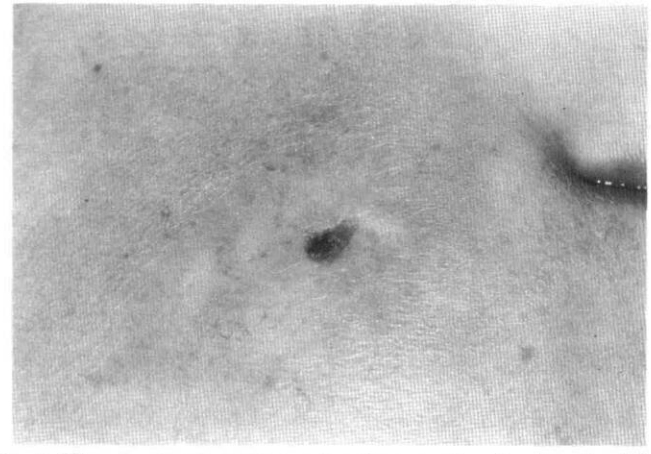

FIG 2. The plaque was an irregular, ivory-colored sclerosis of the skin with loss of normal skin creases. The scab is at the site of the biopsy (2x).

was loose and displaced mesially. There was a deep periodontal pocket on the mesial aspect of the tooth. A periapical radiograph taken at that time revealed extensive bone loss around the maxillary incisors (Fig 3, next page). The maxillary right primary central incisor was subsequently extracted.

When the patient first presented to the University Dental School, it was observed that there was a large defect of the labial gingiva and maxillary alveolus where the right primary central incisor had been lost (Fig 4, next page). At approximately 10 mm from the alveolar crest, the labial surface of the crown of the permanent right central incisor was exposed about 5 mm. The alveolar defect appeared to coincide in line with the scar on the philtrum.

The maxillary right primary lateral incisor and the left primary central incisor appeared to be tilted toward the alveolar defect. The maxillary right primary lateral incisor was only slightly mobile despite a large area of exposed root surface on its mesial and labial aspect. Periodontal probing did not reveal any pockets greater than 2 mm.

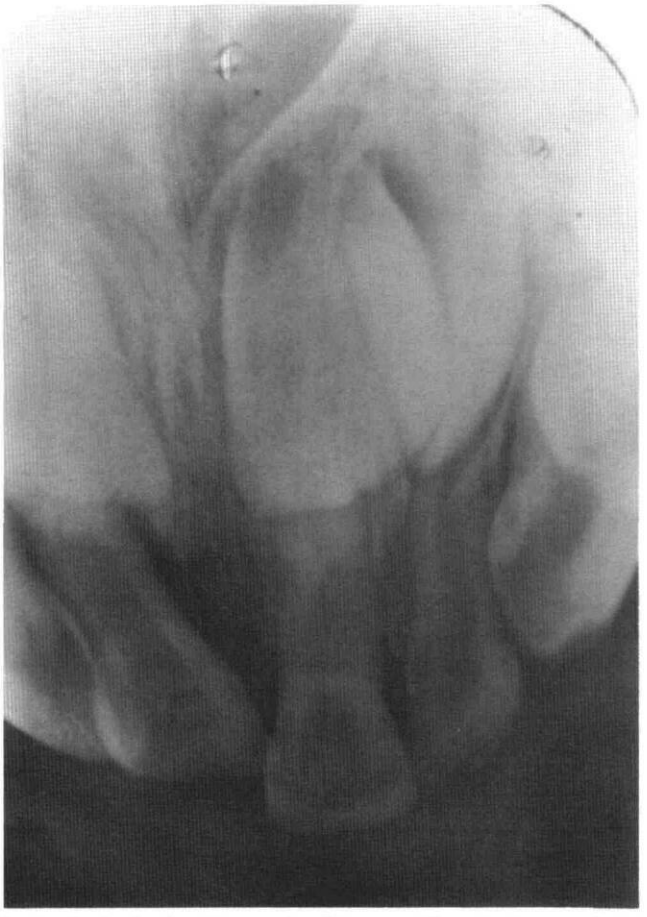

FIG 3. Periapical radiograph of the maxillary incisors taken at first detection of scleroderma. The right central incisor was loosened secondary to a fall. There is widening of the periodontal membrane and proximal bone loss between the primary incisors.

An orthopantogram revealed all permanent teeth except the third molars to be present. An anterior maxillary occlusal radiograph showed that the permanent maxillary right central incisor had a slight palatal inclination relative to the left incisor (Fig 5). No bone abnormalities were detected in those radiographs or in full-mouth periapical radiographs. In addition, the periapical films showed no evidence of an increase in periodontal membrane spaces frequently reported in diffuse scleroderma (Marmary et al. 1981). A soft-tissue radiograph revealed no evidence of calcification in the cheek lesion.

The lips, tongue, floor of the mouth, soft palate, and pharynx were of normal mobility. No dysphagia was reported and speech was normal.

Histopathology

Sections stained with H&E (Fig 6) showed dense sclerosis of dermal collagen extending into superficial subcutaneous fat. There was loss of dermal appendages — particularly the sweat glands — and atrophy of the epithelium with loss of rete pegs. Blood vessels were few and sclerotic changes were seen in their walls.

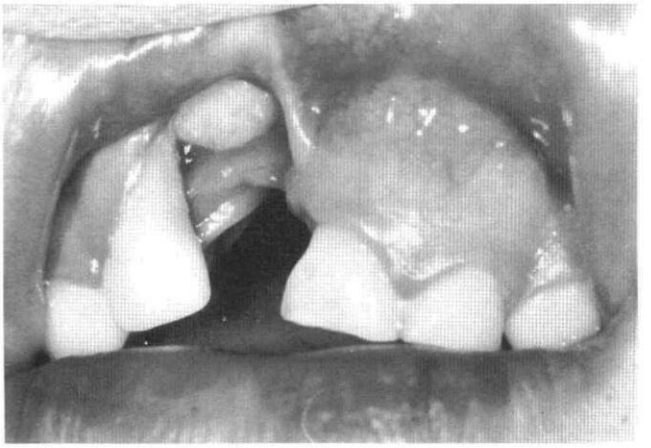

FIG 4. The maxillary anterior dentition 2 months after tooth exfoliation. There is a large alveolar defect on the right, the permanent central incisor crown is exposed, and there is loss of periodontal support on the mesial of the right primary lateral incisor.

The maxillary right primary central incisor had been kept in a dried state by the patient's mother. A ground section of this tooth showed no defect in enamel or dentin formation. The apical third of the root was tapered and the dentin was thinner than in the mid-third. Over this thinned dentin a thick layer of normal cellular cementum was present and showed evidence of a moderate amount of external resorption along the lateral aspects of the root and mildly at the apex. These findings were consistent with tooth exfoliation secondary to loss of periodontal support.

Dental Management

The immediate dental management was directed at the prevention of further loss of support surrounding the maxillary incisors. Although this loss of support was thought to be due to scleroderma, the prevention of further resorption secondary to periodontal inflammation from plaque on the primary lateral and left central

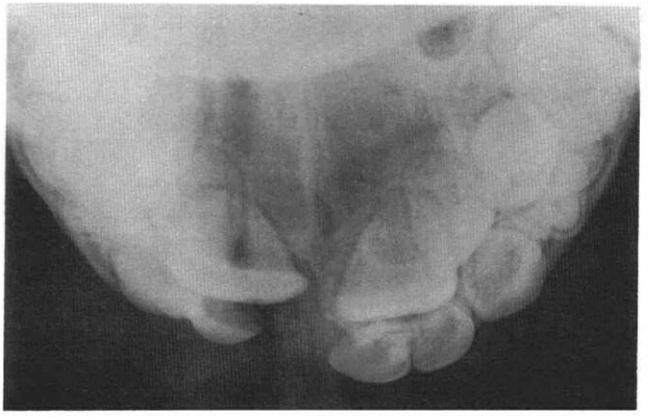

FIG 5. Anterior maxillary occlusal radiograph of the affected area. Note palatal inclination of the permanent right central incisor which may be the result of a dilaceration. The radiolucency on the cingulum of this tooth may be a developmental pit.

incisor was the first priority. The patient and her mother were instructed in meticulous oral cleaning of the site. She was recalled monthly and no further progression of bone resorption was found.

The permanent maxillary right central incisor which is due to erupt within the next 2 years may present several problems. First, it is possible that local fibrosis may impede tooth eruption and alveolar growth. Second, if orthodontic movement is required to encourage eruption or to correct the slight palatal inclination of this tooth, it may not respond normally to orthodontic forces. Third, the lack of labial attached gingiva may present an esthetic problem, already compromised by lack of alveolar growth in this region. This may be corrected by gingival grafts; however, the success of these grafts in scleroderma is unknown.

Discussion

Although the oral manifestations of systemic scleroderma have been well documented, particularly in the adult patient, there are only 2 reports in the dental literature of the dentofacial aspects of childhood scleroderma. Manifestations similar to those in this case such as localized facial skin fibrosis, loss of periodontal support, early exfoliation of primary teeth, lack of alveolar growth, and eruption of permanent teeth have been reported previously in an 8-year-old female patient (Looby and Burket 1942). Lack of alveolar growth in the localized affected region has been described in a 6-year-old female. Her left primary lateral incisor was tilted distally and the primary canine and first molar failed to reach occlusion (Rodnan, 1979).

Since the histologic features of the involved tooth are normal in this case, the lesion most likely developed with the local scleroderma after dental formation had occurred.

Children presenting with localized scleroderma must be assessed carefully, not only for oral problems but also for systemic manifestations as well. Long-term follow-up into adulthood is necessary since systemic involvement may supervene.

The present report appears to conform to the clinical and histological parameters of localized scleroderma with no attendant features of the generalized form. Accordingly, no systemic treatment is underway and the patient is under 6-month observation by her dermatologists. She will be monitored for dental and alveolar development and esthetics.

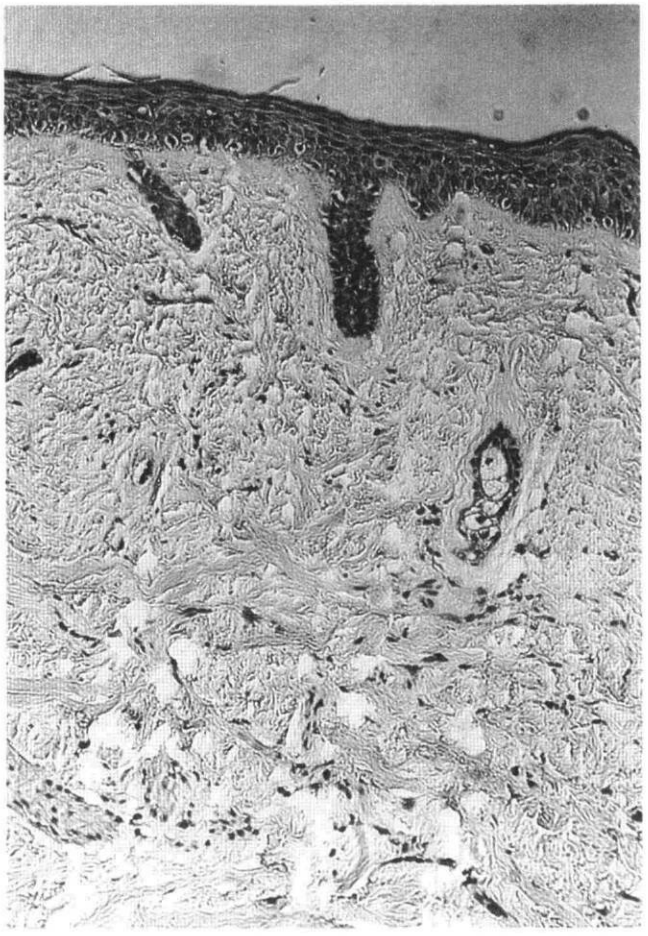

FIG 6. Section of affected skin of right cheek showing thinned epidermis, atrophy of skin adnexal structures and rete ridges, dermal sclerosis with excess collagen, and avascularity (H&E, 200x).

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Immunization: new U.S. schedule

Getting your children fully immunized has become easier. The most recent guidelines from the U.S. Centers for Disease Control alter the schedule of vaccinations, reducing the number of visits needed to complete the process.

Until now, a child had to be taken for immunization at 15 months for the combined measles-mumps-rubella (MMR) vaccine, and again at 18 months for the third dose of oral poliovirus (OPV) vaccine and the fourth dose of diphtheria-tetanus-pertussis (DTP) vaccine.

Now, children 15 months or older can be given these three immunizations simultaneously. One less visit may also prevent overlooking a vital immunization, thereby increasing the number of children who receive the complete series of vaccinations.

It is important for parents to consult their doctor for his/her recommendations for a child's immunization schedule. Some physicians may prefer to continue giving MMR at 15 months, followed by DTP/OPV at 18 months, especially if other purposes are served by the additional visit to the doctor.

New Immunization Schedule for Healthy Infants and Children

2 months	First diphtheria-tetanus-pertussis (DTP) and oral polio (OPV)
4 months	Second DTP and OPV
6 months	Third DTP
15 months	Measles-mumps-rubella (MMR), fourth DTP, and third OPV
24 months	Hemophilus influenza type b (HBPV)
4-6 years	Fifth DTP and fourth OPV
Every 10 years	Diphtheria/tetanus booster