Frontal Linear Scleroderma (En Coup de Sabre): A Case Report

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ABSTRACT

En coup de sabre is a type of linear scleroderma which presents on the frontal or frontoparietal scalp. En coup de sabre in children is associated with asymmetric growth and progressive facial disfigurement. The purpose of this report was to present the case of a 4-year-old girl with a 2-year history of en coup de sabre. The clinical presentation and radiographic findings are discussed. (J Dent Child 2006;73:175-178)

KEYWORDS: LINEAR SCLERODERMA, EN COUP DE SABRE, MALOCCLUSION, ATROPHY

Scleroderma is a relatively rare condition that probably has an immunologically mediated pathogenesis, characterized with extraordinary amounts of dense collagen deposition in body tissues. The skin develops a diffuse, hard texture ("sklero" is Greek for "hard"; "derma" means "skin"), and its surface is usually smooth. Involvement of the facial skin by subcutaneous collagen deposition results in the characteristic smooth, taut, mask-like facies.[1] Two basic forms are recognized:

1. a relatively inconsequential but disfiguring localized cutaneous form known as localized scleroderma; and
2. a potentially life-threatening form known as systemic scleroderma.[2]

Linear scleroderma, morphea, and generalized morphea belong to the so-called localized scleroderma group of diseases.[3,4] Linear scleroderma occurs primarily in children and usually affects the limbs, especially the legs, unilaterally.[3-5] Skin abnormalities can be found on the trunk, abdomen and buttocks.[6] Lesions are characterized by bands of fibrous pigmented skin mainly involving the extremities.

When linear scleroderma occurs on the face or scalp, it is referred to as "en coup de sabre" (the stroke of a sword).[3-7] En coup de sabre (ECDS) develops in the first or second decade of life, presents as band-like sclerotic lesions with more or less marked skin discoloration of the frontoparietal area.[8] Characteristically, ECDS manifests unilaterally and does not extend below the eyebrow. The active stage usually lasts 3 years or longer.[3,8] Involutionary atrophy of skin, muscle, and even bone may occur.

Neurological and ophthalmological abnormalities are not infrequent in ECDS.[8] A more extensive involvement of the subcutaneous tissue and underlying bony structures of the face, is known as progressive facial hemiatrophy or Parry-Romberg Syndrome.[1-3] Some authors consider both ECDS and Parry-Romberg Syndrome to be overlapping conditions.[9] In linear scleroderma, lesions may extend to and can involve the underlying muscles and bones with disturbances in growth and ankylosis. The lesion may extend into the scalp, causing alopecia (loss of hair). Raynaud phenomenon, which can be defined as an abnormal vascular reaction to cold with vasoconstriction, presents sequentially as white, blue, and red color changes of the hands. It is usually observed with systemic forms of scleroderma, not with localized scleroderma.

As in other types of scleroderma, the etiology of ECDS is unknown.[4,7] Hypotheses include microchimerism, which leads to a chronic, low-grade graft-versus-host-like disease or an alteration in antigens caused by ischemic damage.[3,7] *Borrelia burgdorferi* DNA has been identified by polymerase chain reaction assays in tissue sections from some, but not all, localized scleroderma patients.[4,7] Other suggested etiologies include trauma, surgery, and psychological stress.[4]

Serologic abnormalities may include: (1) antinuclear antibodies (ANA); (2) anti-single-stranded DNA antibodies; and (3) rheumatoid factor.[2] Eosinophilia may be present and may correlate with disease activity. A polyclonal IgG and IgM hypergammaglobulinemia may also be present

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and is found more often with severe cases and with clinical progression.[10]

Oral alterations like atrophy of the upper lip, atrophy of the tongue, delayed eruption of the teeth, deficient root development, and root resorption may develop.[1] In some cases, a lesion localizes unilaterally in the cheek, extending to the upper lip and giving the aspect of hemifacial atrophy or a mask-like appearance. It reaches the depth of the labial mucosa and, occasionally, the gingiva and tooth buds, affecting root formation and consequently dental eruption. Structural change may occur in the osseous tissue and result in mandibular joint restriction (pseudoankylosis) and facial and occlusal disharmonies.[11] Orthodontic therapy may be helpful to treat any associated malocclusion.[1]

CASE REPORT

The patient was a 4-year-old girl born to nonconsanguineous, healthy parents. There was no family history of scleroderma. She was clinically well until the age of 2 years, when she presented with a bruised plaque in the left cheek and mandible region. The patient was admitted to a regional hospital, where a skin biopsy led to a diagnosis of chronic dermatitis. She was prescribed many ointments, but without any success.

Subsequently, extension of the lesion to the left side of her forehead and sclerotic cutaneous changes with peripheral hyperpigmentation accompanying the facial hemiatrophy were noted. No alopecia, however, was observed on the frontal scalp. At the age of 3½ years, the child was admitted to the Department of Dermatology, School of Medicine, Marmara University, Istanbul, Turkey. A repeat skin biopsy confirmed the diagnosis of ECDS.

In histopathological examination, hematoxylin- and eosin-stained sections showed epidermal atrophy and thick collagen bundles in the deeper portions of the reticular dermis (Figure 1). A sparse lymphocytic infiltration with a few plasma cells was seen in the superficial and deep perivascular areas and also around the adnexa. A Verhoeff-van Gieson stain showed a decrease in elastic fibers in the deep reticular dermis, with the remaining fibers arranged parallel to the epidermal surface.

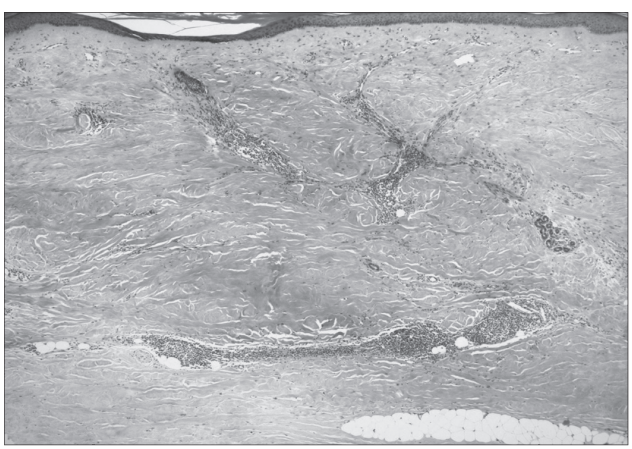

Figure 1. Histopathology of the skin showing thick collagen bundles in deep dermis with an associated superficial and deep perivascular lymphocytic infiltrate (hematoxylin–eosin stain, X200).

The patient had no history of epilepsy or Raynaud phenomenon. Neurological examination was within normal limits. Magnetic resonance imaging (MRI) of the brain was normal. Thinning of the cutaneous and subcutaneous tissues, with no abnormalities in the bony structures, was noted. Orbital computed tomography (CT) revealed no eye involvement, but atrophy of the left medial rectus muscle was observed. Laboratory investigations, including rheumatoid factor, eosinophil count, serum protein electrophoresis, antinuclear antibody, and antibodies to *Borrelia burgdorferi*, were either in the normal range or negative. Electroencephalogram (EEG) recording was normal. A computed tomography (CT) scan of the left scalp demonstrated decrease of cutaneous and subcutaneous fat.

Initially, the patient was treated with colchicine 1 mg/day, but this treatment regimen was not successful. Next, she was put on corticosteroid therapy (1 mg/kg/day of methylprednisolone). That dose was continued for 3 months and, thereafter, was tapered down slowly until complete cessation. No disease progression was noted during this therapy, and no abnormalities of the brain, eye, or bony structures were seen on a repeat cranial and maxillofacial CT done at 4½ years.

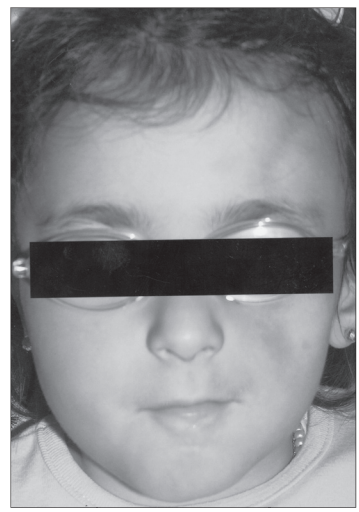

Figure 2. Lesion on the left side of forehead, cheek, and chin, with cutaneous changes accompanying the facial hemiatrophy.

One year later, the 4-year-old female patient was referred to the Department of Oral Diagnosis and Radiology, Dental Faculty, Marmara University.

Extraoral examination revealed that involvement of the left side of the forehead, cheek, and chin, with cutaneous changes accompanying the facial hemiatrophy, was remarkable (Figure 2). Intraoral examination revealed healthy dentition. Although there was no evidence of malocclusion, deviation of the maxillary midline towards the affected side as an effect of ECDS was observed (Figure 3). Atrophy on the tongue's left side was also observed (Figure 4). The panoramic and cephalometric radiographs revealed no anomalies.

The patient is now 5 years old, and she has been placed under periodic follow-up for occlusal development. If necessary, corrective orthodontic treatment will be carried out.

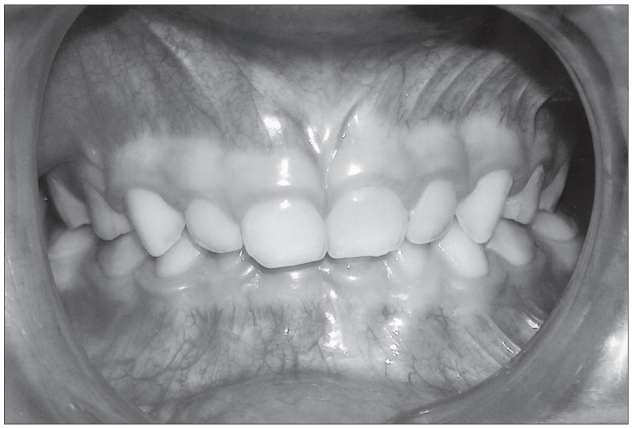

Figure 3. No evidence of malocclusion, but deviation of maxillary midline towards the affected side as an effect of ECDS.

DISCUSSION

Localized scleroderma is an uncommon disease characterized by sclerosis of the skin and subcutaneous tissue, without systemic involvement. Three major types are recognized, based on the extent and morphology of disease: (1) linear scleroderma; (2) morphea; and (3) generalized morphea.[4] Peterson et al published that the incidence of ECDS was 0.13 cases per 100,000 on the basis of a retrospective analysis of patients who developed morphea between 1960 and 1993 in Olmsted County, Minn.[12] Of the 82 cases of morphea identified in that study, 16 had linear scleroderma, including 4 with ECDS and 2 with Parry-Romberg syndrome.

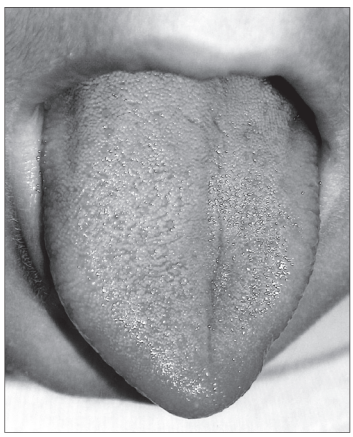

Figure 4. Atrophy on the tongue's left side.

Linear scleroderma typically occurs during childhood and exhibits a 3:1 female predominance.[4] In this report, the case of a 4-year-old female patient with ECDS was reviewed.

ECDS lesions on the forehead may develop along the midline or in a parasagittal location. Involvement of the scalp (with resultant alopecia), nose, and lip sometimes occurs. Bilateral linear morphea is particularly rare, with fewer than 10 cases reported in the literature.[4,8]

In this case, expansion of the lesion on the forehead's left side and cutaneous changes accompanying the facial hemiatrophy was observed, but no alopecia was found in the frontal scalp in the area of the plaque.

The tongue may undergo atrophy, and teeth may develop abnormally. Bones of the skull may become thin. Central nervous system findings include intracerebral calcifications, seizures, and changes in EEG patterns. Eye structures also may be affected, with patients having uveitis, enophthalmos or exophthalmos, and extraocular muscle myopathy.

In this case report, atrophy was present on the tongue's left side and dentition was normal except for a slight deviation of the maxillary midline towards the affected side. Eye structures were not affected; only left medial rectus muscle atrophy was evident. No focal signs were observed upon neurological examination and EEG. Additionally, Raynaud's phenomenon was not observed.

In the disease's early phases, skin biopsy specimens may reveal only inflammatory changes with little or no sclerosis. As the disease progresses, however, sclerosis—which is an increase and homogenization of collagen bundles—is noted. The patient's first biopsy was interpreted as chronic dermatitis, which may correspond to the disease's early inflammatory phase when no sclerosis is expected yet. A repeat biopsy, however, revealed the characteristic changes as the sclerosis developed over time.

There is no uniformly successful therapy for ECDS. Management modalities have included topical, intralesional, or systemic corticosteroids, methotrexate, vitamin E, vitamin D, phenytoin, retinoids, penicillin, griseofulvin, interferon, d-penicillamine, antimalarials, UVA phototherapy, and surgery.[13] Systemic therapy may be helpful in the inflammatory stages, especially for the progressive and destructive variants, but does not improve established sclerosis. Systemic corticosteroids, at the dose of 1 mg/kg/day, were given to the authors' patient, and no more progression of disease was noted thereafter. It cannot be told for sure, however, that it was solely due to the effect of treatment—as the disease itself is said to lose its activity after 3 years.

Linear scleroderma tends to follow a progressive course. Multidisciplinary care, including dermatology, ophthalmology, neurology and dentistry is necessary in the diagnosis and management of the disease.

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