

Case report of localized scleroderma in the scalp and forehead (en coup de sabre)

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ABSTRACT

Localized scleroderma can occur at any age and in any race, but is more common in Caucasians. Environmental factors, such as trauma, infections, or drug or chemical exposure, may play a role, but not for most patients. The disease is not contagious. The disease is not passed on directly from parent to child by any one gene, though certain genes may make a child more likely to develop localized scleroderma.

BACKGROUND

Scleroderma, or systemic sclerosis, is a chronic connective tissue disease generally classified as one of the autoimmune rheumatic diseases. The word “scleroderma” comes from the Greek word “sclero” meaning hard, and the Latin word “derma” meaning skin. Hardening of the skin is one of the most visible manifestations of the disease¹. Scleroderma is a rare disease of unknown etiology, characterized by thickening and hardening of skin resulting from increased collagen production. The term include a variety of diseases, from localized to systemic sclerosis².

Localized scleroderma is generally divided between morphea, linear scleroderma, and scleroderma en coup de sabre. Each type can be subdivided further and some children have more than one type³.

Localized scleroderma incidence range from 0.4 to 2.7 per 100,000 people⁴. Females are primarily affected , and a similar distribution between children and adults occurs^{1,3}. 90% of Diseased children are diagnosed between 2 and 14 years of age^{1,3,5}. It involves three features:

- ◆ an overproduction of collagen.
- ◆ an autoimmune process.
- ◆ blood vessel damage⁶.

Children with linear scleroderma en coup de sabre are a very diverse group. Some children appear to have the classically described disease with lesions only on the scalp and forehead, while other children may have lesions only on the chin or lip. There is another group of children who are termed as having Parry Romberg syndrome. Children with this condition have similar skin lesions, but may have involvement of the whole side of the face and even involvement of the tongue^{3,7}

PATHOGENESIS

Linear scleroderma is a form of localized scleroderma which frequently starts as a streak or line of hardened, waxy skin on an arm or leg or on the forehead. Sometimes it forms a long crease on the head or neck, referred to as **en coup de sabre** because it resembles a saber or sword wound³. It primarily affects the pediatric population. Up to two-thirds of patients given this diagnosis are under the age of 18 years¹, skin Pathogenesis seems to be similar between localized scleroderma en coupe de saber, localized scleroderma and systemic sclerosis, although not fully understood^{1,8-10}.hypothesis that vasculature is the primary target in localized sclerosis^{8,9,11}. Early skin biopsies revealed damaged endothelial cells preceding the development of fibrosis by months to years. Small-artery occlusion is follow, which increase with time by thrombotic events driven by platelets activation, resulting in fibrosis and end-organ damage. The inciting event for microvascular damage remains unknown^{11,12}. Available data suggests a complex pathogenesis of scleroderma, in which blood vessels, the immune system and extracellular matrix are affected and may contribute to the development of the disease till now^{13,14}.

CLINICALLY

Female patient, of 12 years of age, presented with ipsilateral, near the midline of the scalp, with a lesion that is started before two and half years previously, as linear red mark, then linear scar like lesion of about 1.5 cm in width and 6 cm in length and of 0.5 cm depression, associated with hair loss and tethering of the skin to the underling connective tissues, the skin is thin, hypopigmented silver in color, and violaceous in appearance. The lesion is located in the scalp on the top of the head, figures (1- A,B,C).



A



B



C

Figure (1) The lesion in the scalp

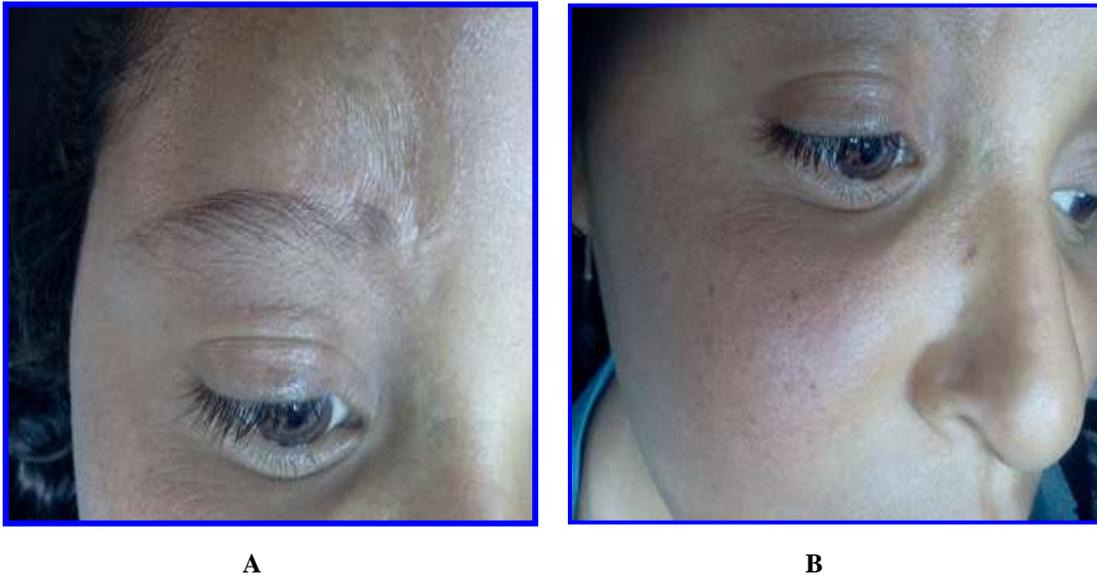


Figure (2) The lesion in the forehead

The lesion extended to the forehead figure(2,A) on the right side of the face till the medial canthus of the right eye figure(2,B). No any associated skin lesion or clinical presentation is noted. Treatment varies depending on the patient's disease activity, lesion location and extent, and whether there are related problems. Careful clinical evaluation is the primary method for monitoring scleroderma. X-rays and computerized tomography (CT) scans are used to look at bone abnormalities. Thermography can detect differences in skin temperature between the lesion and normal tissue. Ultrasound and magnetic resonance imaging (MRI) can aid soft tissue assessment, patients with mild superficial disease, topical medications often are used to control the inflammation and soften the skin. like corticosteroids.

DISCUSSION

Typically skin lesion develop prior to neurological manifestations, although cases of the reverse have certainly been described. A literature review done by Kister et al¹⁵. found that the skin lesion preceded the onset of neurological symptoms by an average of several years, although in 29% of the patients studied the two occurred within one year of each other⁹.

The lesion looks to be of the same size from about six months till now, so no further treatment is given for the patient.

POINTS TO REMEMBER

- Treatment for scleroderma should start as soon as possible. Treatment is more effective during the early inflammatory stage as the medicines do not directly target fibrosis.
- Children are at risk for growth problems and internal tissue involvement, so regular follow-up visits with the rheumatologist are essential to ensure that treatment is controlling inflammation and to minimize side effects from treatment.

The cause is not clear. What is known is that cells called fibroblasts make too much of a protein called collagen. The collagen gets deposited in the skin causing scarring and thickening (fibrosis). It is not known why the fibroblasts produce too much collagen in the areas of affected skin. It is probably some fault with the immune system. It is sometimes seen after the development of diseases in which the immune system attacks the body's own cells (autoimmune conditions), such as lichen sclerosus and lichen planus.

CONCLUSION

The patient kept under observation for further presentations of the disease with minor cosmetic correction of the lesion. Juvenile scleroderma can be unsettling for the child and his/her family, but if treated properly by an experienced physician, it is a condition that can be managed.

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