Linear Scleroderma ‘en coup de sabre’ and Epilepsy: A Case Report

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This article is available from: http://www.jtad.org/2013/2/jtad1372c3.pdf

Key Words: Linear scleroderma, en coup de sabre, epilepsy

Abstract

Observation: Linear scleroderma is a form of localized scleroderma characterized by sclerotic lesions distributed in a linear, band-like pattern. The “en coup de sabre” subtype of linear scleroderma is more often associated with systemic manifestation, including epilepsy. Here, we report a case of typical linear scleroderma “en coup de sabre” with epilepsy.

Introduction

Linear scleroderma, a variant of localized scleroderma, is a disorder of unknown origin and characterized by fibrosis of the skin and underlying tissue [1]. When linear scleroderma occurs on the head, it is called as linear scleroderma “en coup de sabre” (LSCS). Depressed skin lesions of LSCS are generally located near the midline of the forehead with extension into the frontoparietal scalp. The disease is commonly associated with neurological symptoms, including epilepsy. We present a LSCS patient with generalized tonic-clonic seizures developed three years after the characteristic skin lesion on his head occurred.

Case Report

A 22-year-old man presented to our outpatient clinic with a 19-year-history of atrophic patches on his head. On dermatologic examination, he had band-like depressed and deep atrophic skin lesion with scarring alopecia on the left aspect of his forehead extending to left parietal scalp region. (Figures 1 and 2) He had no history of preceding infection or trauma and had no family history of connective-tissue disease. A previously conducted skin biopsy of his forehead confirmed the diagnosis of localized scleroderma. After the disease had been diagnosed, he used topical steroids and topical calcipotriol for two years to his lesional region. However, he discontinued the topical treatment after the age of five. Three years after onset of the disease, at the age of six years, the patient began to experience generalized tonic-clonic seizures. He was diagnosed with epilepsy and used various anti-epileptic drugs. Since last five years, he has...
used valproic acid 1000 mg/day. The atrophic changes of the skin remained the same since about last 15 years and his seizures had not worsened at last five years.

Discussion

The term of LSCS was first used in 1854 by Addison, given the resemblance of the skin lesions to the stroke of a sabre [2]. The reported incidence of LSCS is 0.13 cases per 100,000 population [3]. Females are primarily affected and a similar distribution between children and adults occurs. In adults, disease incidence peaks in the fifth decade, whereas 90% of children are diagnosed between 2 and 14 years of age [4].

As in other types of scleroderma, the etiology of LSCS is not known exactly. However, endothelial cell damage leading to increased fibroblast activity and ischemia as a result of luminal narrowing and following modification of collagen production has been proposed as the pathogenic mechanism [2]. Viral infections, particularly due to *Borrelia burgdorferi*, genetic factors, and preceding trauma may also play roles in its development. Neurologic abnormalities, particularly seizures, have been described in association with LSCS [5].

In different studies the incidence of patients with epileptic seizures in all LSCS patients varies between 8% [6] with 13% [7]. Other neurologic abnormalities described in LSCS are focal neurologic deficits, movement disorders, trigeminal neuralgia, and mimics of hemiplegic migraines. In the differential diagnosis, the firstly considered disease is progressive hemifacial atrophy (*Parry-Romberg* syndrome) characterized by progressive hemifacial atrophy without cutaneous sclerosis [8]. However, many authors believe that both diseases are different clinical variants of the same disease [9]. Additionally, LSCS and *Parry-Romberg* syndrome coexist in 20–37% of the patients with LSCS diagnosis, and both conditions have similar age of onset and disease course [4].

The treatment of LSCS remains difficult. At the active stage of the disease agents such as D-penicillamine, systemic and topical steroids, methotrexate and topical calcipotriol with PUVA may be effective in the treatment of LSCS. When the progression of the disease is completed injection of autologus fat tissue, silicon, bovine collagen and inorganic implants to atrophied lesions may be used for cosmetic revision [9].

In our patients, neurologic manifestations occurred after the diagnosis of LSCS as in many patients. As a conclusion, we suggest a long and careful follow-up of LSCS patients because of the accompanying abnormalities such as epilepsy.

References

