Journal of the Turkish Academy of Dermatology
eISSN 1307-394X

Case Report

DOI: 10.6003/jtad.1372c3

Linear Scleroderma '*en coup de sabre*' and Epilepsy: A Case Report

Mutlu ÇAYIRLI,[1] MD, Gürol AÇIKGÖZ,[2] MD

Address: [1]Ağrı Military Hospital, Dermatology Service, Ağrı, Turkey, [2]Gülhane School of Medicine, Department of Dermatology, Ankara, Turkey

E-mail: mutlu78tr@yahoo.com

Corresponding Author: Dr. Mutlu Çayırlı, Ağrı Military Hospital, Dermatology Service, Ağrı, Turkey

Published:
J Turk Acad Dermatol 2013; **7 (2)**: 1372c3.
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 skins 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

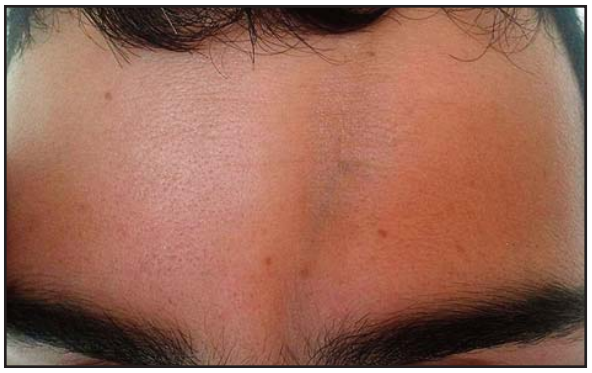

Figure 1. Band-like, depressed and atrophic lesion on forehead

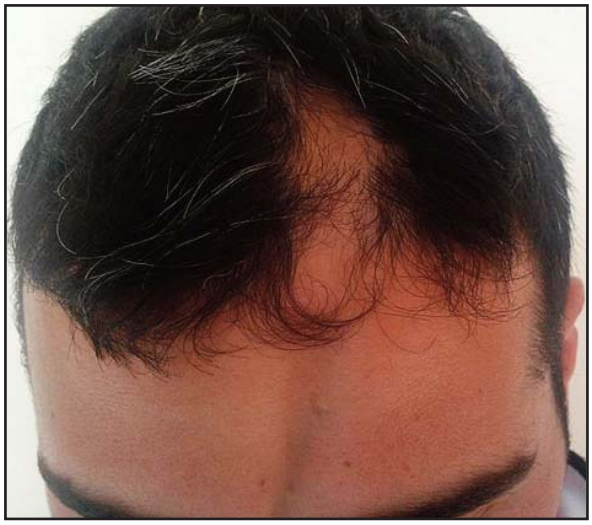

Figure 2. Scarring alopecia on the left parietal scalp region

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 autologous 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

1. Obermoser G, Pfausler BE, Linder DM, Sepp NT. Scleroderma en coup de sabre with central nervous system and ophthalmologic involvement: Treatment of ocular symptoms with interferon gamma. J Am Acad Dermatol 2003; 49: 543-546. PMID: 12963929

2. Chiang KL, Chang KP, Wong TT, Hsu TR. Linear Scleroderma "En Coup De Sabre": Initial Presentation as Intractable Partial Seizures in a Child. Pediatr Neonatol 2009; 50: 294-298. PMID: 20025145

3. Peterson LS, Nelson AM, Su WP, Mason T, O'Fallon WM, Gabriel SE. The epidemiology of morphea (localized scleroderma) in Olmsted County 1960-1993. J Rheumatol 1997; 24: 73-80. PMID: 9002014

4. Amaral TN, Marques Neto JF, Lapa AT, Peres FA, Guirau CR, Appenzeller S. Neurologic involvement in scleroderma en coup de sabre. Autoimmune Dis 2012; 2012: 719685. PMID: 22319646

5. Garófalo Gómez N, Novoa López L, Gómez García AM, Méndez Méndez M. Linear scleroderma en coup de sabre and epilepsy: Presentation of a case in a child. Neurologia 2012; 27: 449-451. PMID: 22178046

6. Zulian F, Athreya BH, Laxer R, et al. Juvenile Scleroderma Working Group of the Pediatric Rheumatology European Society (PRES). Juvenile localized scleroderma: clinical and epidemiological features in 750

children. An international study. Rheumatology (Oxf) 2006; 45: 614-620. PMID: 16368732

7. Tollefson MM, Witman PM. En coup de sabre morphea and Parry-Romberg syndrome: A retrospective review of 54 patients. J Am Acad Dermatol 2007; 56: 257-263. PMID: 17147965

8. Holland KE, Steffes B, Nocton JJ, Schwabe MJ, Jacobson RD, Drolet BA. Linear scleroderma en coup de sabre with associated neurologic abnormalities. Pediatrics 2006; 117: e132-136. PMID: 16326691

9. Jun JH, Kim HY, Jung HJ, Lee WJ, Lee SJ, Kim do W, Kim MB, Kim BS. Parry-Romberg Syndrome with En Coup de Sabre. Ann Dermatol 2011; 23: 342-347. PMID: 21909205