EN COUP DE SABRE- A CASE REPORT

SM YUSUF*MB BS, MSc, FWACP, **MU SANI*** *MBBS, FWACP, MS* **MIJINYAWA*** *MBBS, FWACP,* **AZ MOHAMMED**** *MB BS, FMCPath,*

**Dermatology unit, Department of Medicine, Aminu Kano Teaching Hospital, Kano- Nigeria*
***Department of Histopathology, Aminu Kano Teaching Hospital*

Correspondence- SM YUSUF, E-mail shehumy@yahoo.com

Abstract

En coup de sabre (ECDS) is a peculiar variant of morphea characterized by linear, atrophic cutaneous depressed plaques that usually develops during childhood. We present a case of a 5year old boy whose lesion started at the age of 3.

Introduction

Scleroderma may occur as a systemic disease or as a localised form.[1] Localised scleroderma presents in three clinical forms: generalised, morphoea (atrophic and sclerotic skin lesions), and linear scleroderma.[2] Linear scleroderma is characterized by localized fibrosis of skin, blood vessels, subcutaneous fat, muscle and sometimes bone. It primarily affects the population during the first and second decade. Upper limbs are the most commonly affected but the fronto-parietal area of the forehead and scalp may also be involved initially, this is referred to as en coup de sabre (ECDS).[3]

Case report

A5-year-old woman presented with a 2-year history of a new scar on the right side of his forehead. The patient reported no history of abrasions from minor traumas to the forehead, seizures, dysphagia or Reynaud's phenomenon. His past medical, social and family histories were unremarkable. On physical examination, there was a hypopigmented, atrophic, linear plaque in the centre of the left forehead. The plaque extended superiorly to the frontal scalp. There was alopecia in the frontal scalp in the area of the plaque. The face was normal and symmetric. There were no signs of neurological abnormalities.

Total blood count including eosinophil count, were normal. Rheumatoid factor and serum protein electrophoresis were also normal. A biopsy specimen obtained from an area of a plaque on the frontal scalp showed epidermal atrophy and thick collagen bundles in the deeper portions of the reticular dermis. There is an associated sparse, superficial and deep, perivascular and periadnexal infiltrate of lymphocytes with a few plasma cells. He was subsequently treated with topical mometasone furoate and oral prednisolone 10mg daily without appreciable response.

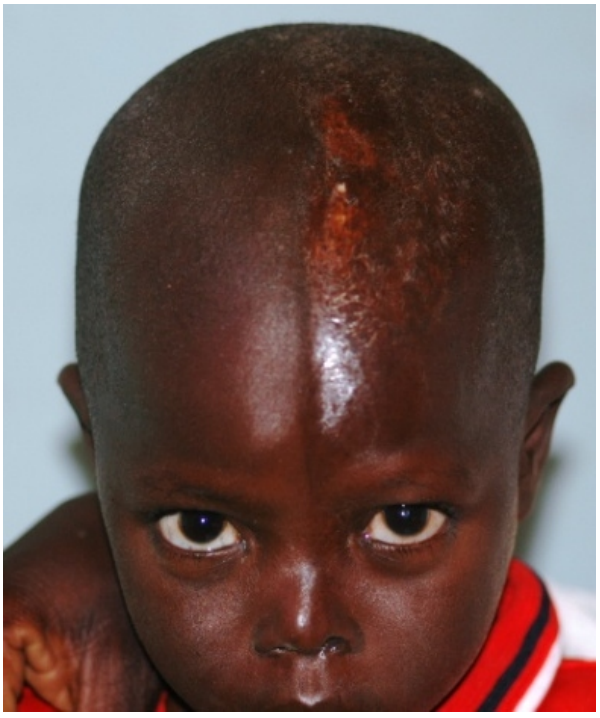

Fig1. Atrophic plaque of En coup de sabre

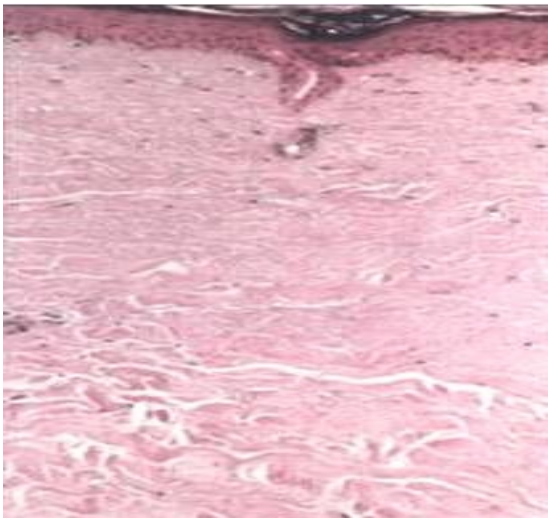

Fig. 2 Histology showing atrophic epidermis and thick collagen bundles in the dermis X 100

Discussion

En coup de sabre (ECDS) is a rare variant of morphea which presents on the frontal or frontoparietal scalp suggestive of a stroke from a sword.[2] Affected children often have associated asymmetric growth and progressive facial disfigurement. The skin is involved first and appears indurated. An ivory coloured, band-like depression of the frontoparietal region is characteristic. The lesions can result in scarring alopecia of the scalp, eyebrows, or eyelashes.[3] Although morphoea is generally more common in whites than in blacks and more common in women than in men, linear morphea seems to occur equally in both sexes.[2] En coup de sabre develops during childhood, usually at about 7 or 8 years.4

En coup de sabre may be associated with specific problems which include refractory partial motor dextrolateral seizures, hemiparesis, and progressive intellectual deterioration. Intracerebral and orbital lesions, impaired vision, and retro-ocular pain of the affected eye. Malalignment of the jaw and unilateral atrophy of the tongue have also been reported.[5] Alterations in antinuclear antibodies, anti-single-stranded DNA antibodies, rheumatoid factor and eosinophilia may accompany en coup de sabre.[6] The exact cause of morphoea is not known, however the proposed aetiologies include a blastoid process, in which an abnormal clone present in one area of embryologic development results in a linear array of cells susceptible to morphea; infection, specifically due to Borrelia; microchimerism or an alteration in antigens caused by ischemic damage; and autoimmunity, perhaps involving increased secretion of interleukin (IL)-1 and IL-2.[2,7,8]

Clinical mimics of en coup de sabre include atrophoderma of Pasini and Pierini, but, patients with atrophoderma of Pasini and Pierini have tan-coloured, depressed atrophic plaques that occur usually on the back, lack erythema, and have well-circumscribed edges (cliff-drop borders). Eosinophilic fasciitis (Shulman's syndrome) is morphea of the fascia and primarily affects the extremities. The condition can be painful and manifests with cutaneous cobblestoning, induration, and oedema; Parry-Romberg syndrome, or progressive facial hemiatrophy, is a unilateral, slowly progressive atrophy that affects the skin, subcutaneous tissues, muscles, and bones and can overlap en coup de sabre.[9,10] ECDS can be diagnosed clinically and confirmed histologically. Histologically, ECDS is characterized by the deposition of collagen in the dermis and subcutis, inflammatory perivascular and diffuse infiltrate (predominantly seen in early lesions and composed of lymphocytes, macrophages and plasma cells), small vessel wall thickening and lumen narrowing. The collagen bundles become hyalinized, thus replacing subcutaneous fat and muscle, characterize the late sclerotic phase. Importantly, the elastic tissue is absent.[11]

Treatment included topical, intralesional, or systemic glucocorticoids; vitamin E; vitamin D3; phenytoin; retinoids; penicillin; griseofulvin; interferon-*a*, D-penicillamine; antimalarials; ultraviolet A phototherapy with or without psoralens; and surgery.[12]

References

1. Katz KA. Frontal linear scleroderma (en coup de sabre). *Dermatol Online J.* 2003;9(4):10.
2. Tuffanelli DL. Localized scleroderma. *Semin Cutan Med Surg*. 1998;17:27-33.
3. Peterson LS, Nelson AM, Su WP: Classification of morphea (localized scleroderma). Mayo Clin Proc 1995: 70:106876.
4. Peterson LS, Nelson AM, Su WP, et al. The epidemiology of morphea (localized scleroderma) in Olmsted County 1960-1993. *J Rheumatol.* 1997;24:73-80.
5. 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.
6. Amento EP. Immunologic abnormalities in scleroderma. Semin Cutan Med Surg 1998;17:18.
7. Soma Y, Fujimoto M. Frontoparietal scleroderma (en coup de sabre) following Blaschko's lines. *J Am Acad Dermatol.* 1998;38 (2 Pt 2):366-368.
8. Holland KE, Steffes B, Nocton JJ, Schwabe MJ, Jacobson RD, Drolet BA: Linear scleroderma en coup de sabre with associated neurologic abnormalities.
9. Emery H. Pediatric scleroderma. *Semin Cutan Med Surg.* 1998;17:41-47.
10. Orozco-Covarrubias L, Guzman-Meza A, Ridaura-Sanz C , Carrasco DD (?) , Sosa-de-Martinez C, Ruiz-Maldonado R. Scleroderma "en coup de sabre" and progressive facial hemiatrophy. Is it possible to differentiate them? J Eur Acad Dermatol Venereol 2002;16:361366.
11. Serup J. Localized scleroderma (morphoea). Acta Derm Venereol 1986; 122: 1
12. Hunzelmann N, et al. Management of localized scleroderma. Semin Cutan Med Surg 1998;17:34-36.