En coup de sabre morphea: an uncommon condition in Africa

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Abstract

The term *en coup de sabre* morphea refers to a lesion of linear morphea typically located in the frontoparietal scalp and/or the paramedian forehead, often resembling a strike with a sword. In literature, *en coup de sabre* morphea, and *en coup de sabre* scleroderma are terms used interchangeably and synonymously. Due to the rarity of this condition, treatment is largely based on case report series, leaving much room for speculation in terms of drugs of choice, duration of treatment, and dosages. Although it typically leaves behind notable and often permanent skin pigmentary changes and indentation of the affected areas, this condition usually remits spontaneously, even in the absence of an active form of treatment. The disease severity and prognosis vary according to the subtype: circumscribed morphea has a generally more benign course when compared with linear scleroderma and generalized morphea.

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Case Report

A 31-year-old female, with a 2-year history of a lesion that started as an asymptomatic macule on her forehead that progressively extended to the left nasal bridge and parietal aspect of the scalp, was referred to the Dermatology Outpatients Department (OPD) at Universitas Academic Hospital. She gave no history of convulsions, persistent headaches, or visual disturbances.

On clinical examination, she had a strikingly linear and poorly demarcated hyperpigmented scar that had marked depression, and was non-inflamed, affecting her left nostril all the way to her mid-scalp causing secondary alopecia and a subtle facial disfigurement. Clinically, the lesion felt firm, atrophic, and indurated (Figure 1).

She had no systemic symptoms, nor visual or neurological signs to indicate any extracutaneous involvement.

Except for a positive antinuclear antibody of low titers of 80, all other related serological tests came back negative for systemic connective tissue disease, namely rheumatoid factor, erythrocyte sedimentation rate, and the antineutrophil cytoplasmic antibodies (C- and P-ANCA's). The extractable nuclear antigen test and C-reactive protein results were also normal, and so was her full blood count blood test.

Histological examination of sections of the punch biopsy of the skin showed the presence of skin consisting of the epidermis, dermis, and underlying subcutis. The epidermis appeared atrophic, there was no hyper- or parakeratosis is noted. Extensive solar elastosis was present in the dermis, dense collagen bands were also noted in the dermis and the subcutis and atrophy of the skin adnexal structures were evident. A histochemical stain for acid mucin was negative and no tumor or dysplasia was present. The morphological features were indeed consistent with the clinical impression of morphea (Figure 2). She was started on chloroquine sulfate 200 mg daily and medium potency topical corticosteroid in the form of betamethasone cream was also used daily with monthly follow-ups. She was further referred to the Ophthalmology and Neurology Departments as a precaution, with no pathological findings found. Ophthalmology is also important for baseline eye examination prior to chloroquine sulfate commencement.

She showed clinical signs of improvement: the disease was halted from further progression (indentation and deepening in hyperpigmentation) and the affected skin had less induration and felt softer in texture 6 months after treatment (at the time of the article write-up). She remained with no extracutaneous symptoms, while her main concerns were that of cosmetic appeal.

Discussion

Scleroderma is a rare auto-immune disease of unknown etiology in which increased collagen deposition occurs and results in dermal thickening with a typical loss of subcutaneous tissue, sometimes affecting the underlying musculoskeletal structures. Involvement may be diffuse (systemic sclerosis) or localized to the skin (localized scleroderma). Linear scleroderma represents a unique form of localized scleroderma that primarily affects the pediatric population, with 67% of patients diagnosed before the age of 18.[1]

As is widely the case with most connective tissue diseases, morphea is more common in women (2.6-3.1). The mean average age of onset in pediatrics is around adolescence, but adult onset is not uncommon.[2]

Due to the rarity of the two conditions, most of the available literature is based on case reports, case series, and clinical surveys.[3]

Lesions of morphea can be classified into one of the following 3 categories based on physical examination findings and history: active, regressing, and burnt out. Active disease is defined as the presence of cutaneous areas of active sclerosis with or without a violaceous border, and with or without the extension of disease into new areas. Lesions are classified as regressing if there is a notable decrease in sclerosis and sometimes changes in pigmentation to a pre-disease state. Burnt-out lesions can have no sclerosis and still remain pigmented.[3-5]

The diagnoses of *en coup de sabre* are typically made on clinical grounds; however, antinuclear antibody with homogeneous and speckled patterns may be positive in 37-50% of patients with linear scleroderma, and anti–single-stranded-DNA antibodies may be present. More specific autoantibodies such as Scl-70, anticentromere, Ro/La, and U1RNP may precede the development of systemic disease and patients with these markers should be followed closely for several years as they remain at high risk for further disease progression and systemic disease.[1,6]

Despite numerous studies trying to elucidate the mechanisms of morphea, its causes are still unknown but generally accepted to be auto-immune. Other associations included mechanical events (67%), followed by infections (25%), drugs (5%) and psychological distress (3%).[7]

Dermoscopic examination of affected sites typically shows loss of follicular openings on a whitish skin surface; scattered black dots, broken hairs, pili torti, and short thick linear, and branching tortuous vessels on the periphery of the lesion.[8]

Common eye signs of this condition include keratopathy, retinal detachment in childhood, restricted eye motility, and diplopia.[9]

A significant number of patients, especially children, will have central nervous symptoms to a varying degree, this will commonly include headaches and convulsions. Bone and dental abnormalities are detected with magnetic resonance imaging scans, computed tomography scans, and electroencephalogram testing.[3,10,11]

Lesions of localized scleroderma are histologically characterized by perivascular lymphocytic infiltrate in the reticular dermis and swollen endothelial cells.[12]

Treatment

Children and teenagers with morphea and scleroderma of different forms generally tend to have a better prognosis as compared with their adult counterparts. Methotrexate and oral prednisone are yielding the best clinical outcome when used in combination, but they have also been found to have a high side-effect profile. Antimalarial drugs such as chloroquine have also been favored with less side effect profile, along with topical, oral, and injectable corticosteroids.[13]

Narrow-band ultraviolet can also be used to treat morphea. Because of some similarities in this group of conditions, medications used in the treatment of other forms of morphea and scleroderma are commonly used to treat *en coup de sabre* morphea and Parry-Romberg syndrome.[13,14]

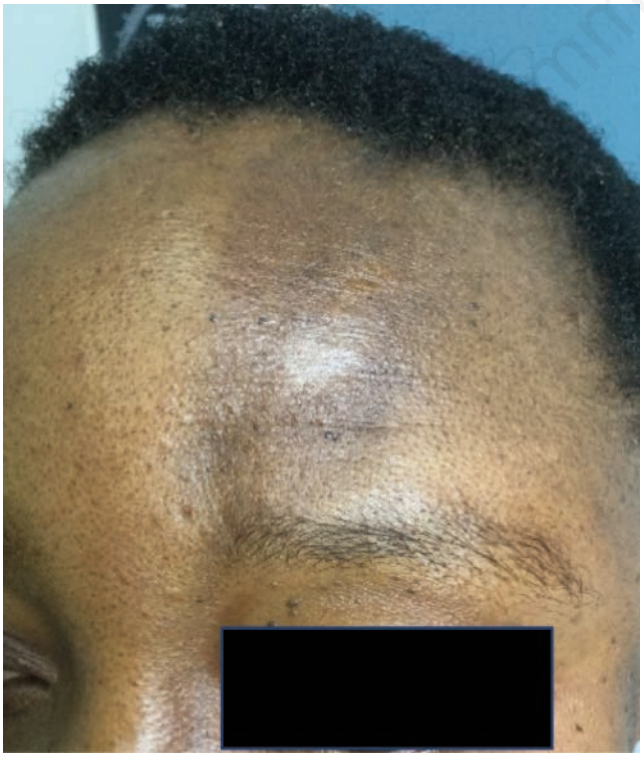

Figure 1. Picture of *en coup de sabre* morphea, courtesy of The South African Institute of Dermatology.

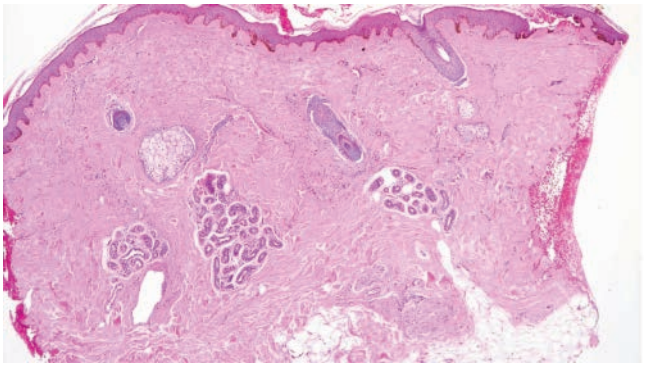

Figure 2. Morphea on a hematoxylin and eosin staining. Courtesy of Dr. Michelle du Preez (Department of Anatomical Pathology, University of the Free State, South Africa) and Professor Wayne Grayson (Ampath Laboratories, Gauteng province, South Africa).

Conclusions

Localized forms of morphea commonly tend to burn out even in the absence of active treatment, leaving permanent scarring, and pigmentary changes with or without disfiguration of the underlying structures. However, treatment is highly recommended to halt the progression of the disease.[15]

The rarity of this form of scleroderma has resulted in the paucity of documented knowledge about the drug of choice, duration of treatment and dosages, resulting in treatment being individualized in the case series type of literature that is currently available.

Early recognition of this condition can prevent long-term complications in the form of facial deformities, including severe eye involvement, sometimes even warranting intervention by plastic surgeons. Relapses are uncommon.[9]

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