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Case Report

'En coup de sabre' extending from scalp to neck: an extremely rare presentation

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Abstract 'En coup de sabre' is characterized by sclerotic lesions distributed in a linear, band-like fashion on the frontoparietal scalp and forehead. Less commonly, skin lesions may extend to the nose, cheek and upper lip. Lesions extending down to the chin and neck are extremely rare. Here in, the authors are reporting a case of 'en coup de sabre' where the lesions extend from scalp to the neck.

Key words
En coup de sabre, frontoparietal scalp, morphea, sclerotic lesions.

Introduction

'En coup de sabre' is a type of linear morphea characterized by sclerotic lesions present in a linear fashion on the frontoparietal scalp and forehead. Sometimes the skin lesions extend to the nose, cheek, upper lip and chin. Very rarely lesions approaching neck have been reported.[1] In this article, we describe an extremely rare case of 'en coup de sabre' in a 35-year-old female where the skin lesions extend from frontoparietal scalp to the neck.

Case Report

A 35-year-old female, presented with chief complaint of thickening and tightening of skin over left side of her forehead and face for one year. The disease started from the left frontoparietal area of scalp and then progressed downwards slowly on the same side involving forehead, infraorbital area, cheek, maxillary area, upper lip, chin and submandibular area of neck sparing eyelids over a span of 5-6 months. There is no history of trauma or weakness of face. There is no history of seizures, headache or weakness of any other body part. The patient denied any systemic complaints.

Physical examination revealed ill-defined, brownish hyper pigmented, indurated plaques over the left frontoparietal region of the scalp and forehead reaching to the left one thirds of supraorbital region, infra orbital area, cheek, maxillary area, upper lip, chin and submandibular area of neck (**Figure 1**). There were areas of atrophy at some places (**Figure 2**). Neurological examination did not reveal any abnormality. Ocular and otolaryngological examinations were unremarkable. ANA was negative. Brain MRI did not show any abnormal findings. Skin biopsy was not done. Based on history and suggestive clinical findings, a diagnosis of 'en coup de sabre' was made and she was prescribed topical tacrolimus 0.1% ointment to be applied twice daily along with broad spectrum sunscreens. After one year of follow up, she has neither reported any further progression of skin lesions nor development of any such lesions on any other body parts while the existing skin lesions have improved with regard to thickening and hyperpigmentation. Fortunately, there have been no signs of

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Figure 1 En coup de sabre extending from frontoparietal scalp to neck in a 35-year-old female.

Figure 2 Close view of lesions of en coup de sabre showing linear depressed atrophic plaque over forehead.

neurological involvement during the follow up period.

Discussion

In 1854, Addison coined the term 'en coup de sabre' as the lesion resembled the scar resulting from a saber (sword) cut.[1] Morphea en coup de sabre presents clinically in a band-like fashion over the frontoparietal region of scalp. In certain cases, skin lesions extend downwards to the nose, cheek, upper lip, and involve the mouth and gums. However, in very rare cases when the disease is severe, lesions extending to the chin and neck have been reported.[2] The disease usually has an active stage lasting 2 to 5 years, which explains the need of follow-up for such cases.

The etiology of morphea is still unknown. However several triggering factors have been Implicated which include trauma, injections of vitamin K, immobilization, Bacille Calmette–Guérin (BCG) vaccination, mechanical compression from clothing, previous radiotherapy, electronic slim belt for obesity, etc.[3] In our case, triggering factor could not be identified.

A number of neurological abnormalities have been associated with 'en coup de sabre' and are usually preceded by the development of cutaneous lesions months to years. However involvement of CNS usually doesn't correlate with the cutaneous disease activity and may present years after the development of skin lesions which stresses upon the need for regular follow up of these cases. Various neurological manifestations reported include epilepsy, headache, movement disorders, focal neurological deficits and intellectual deterioration.[4] However, in our case there was no neurological involvement seen clinically as well radiologically.

The treatment of morphea has been updated. A variety of treatment options have been tried with variable success. These include calcipotriol combined with betamethasone dipropionate, phototherapy, topical immunomodulators like tacrolimus; Imiquimod, systemic steroids combined with methotrexate, cyclosporine, mycophenolate mofetil, D-penicillamine, photopheresis etc.[5,6] In the present case, topical tacrolimus 0.1% ointment was prescribed twice daily along with broad spectrum sunscreens.

'En coup de sabre' can remain quiescent for a long time and disease activity may return after years of clinical inactivity.[1] In addition, authors have reported a case of a 17-year-old female where 'en coup de sabre' was followed by development of plaque type morphea on her left thigh.[7] Hence, a regular follow up to screen for progression of lesions of 'en coup de sabre', development of some other type of morphea at distant body site; and signs and symptoms of neurological involvement is mandatory.

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