Can lesions of 'en coup de sabre' progress after being quiescent for a decade?

Dear Editor,

'En coup de sabre' is a form of localized scleroderma (also known as morphea) characterized by sclerotic lesions distributed in a linear, band-like fashion on the frontoparietal scalp and forehead. In some cases, skin lesions may extend to the nose, cheek, upper lip, chin, and neck. Herein, we describe a unique case of en coup de sabre in a 32-year-old female which has not been reported before to the best of our knowledge.

Medical history of the patient can be traced 12 years back when she suffered blunt trauma to the forehead. Few months after the trauma, she developed a brownish indurated skin lesion affecting her forehead at the site of trauma that progressed downwards for 3 to 4 months up to the medial part of the left supra orbital region after which it stopped on its own. During the next 12 years, the skin lesion remained localized to the forehead and there was no further disease progression clinically. However, in the last 4 months, the patient complained of new lesions progressing down the forehead involving the medial portion of left orbit, nasal bridge, ala of the nose, skin over the left maxilla, and chin in a vertical fashion in that order. There was no history of seizures, headache, weakness of face, or weakness of any other body part. The patient denied any systemic complaints. Physical examination revealed well-defined, brownish hyper pigmented, indurated plaques over the frontal region of the scalp and forehead reaching to the glabellar region in the left paramedian location, medial wall of left orbit, bridge of the nose, left side of the nose, left nasal ala, skin over the left maxillary region, and chin in a vertical fashion (Figure 1). There were areas of scarring alopecia in the frontal region of the scalp. Neurological examination did not reveal any abnormalities. Ocular and otolaryngological examinations were unremarkable. ANA was negative. Brain MRI did not show any abnormal findings. Histopathological examination of the skin biopsy revealed epidermal atrophy, perivascular infiltrate of lymphocytes and plasma cells in the dermis and subcutaneous tissue (Figure 2). Thick bundles of densely packed collagen with sparse adnexal structures were seen in the dermis and subcutaneous tissue (Figure 3). Based on physical examination and supported by histopathology, a diagnosis of en coup de sabre was made and she was prescribed topical tacrolimus.

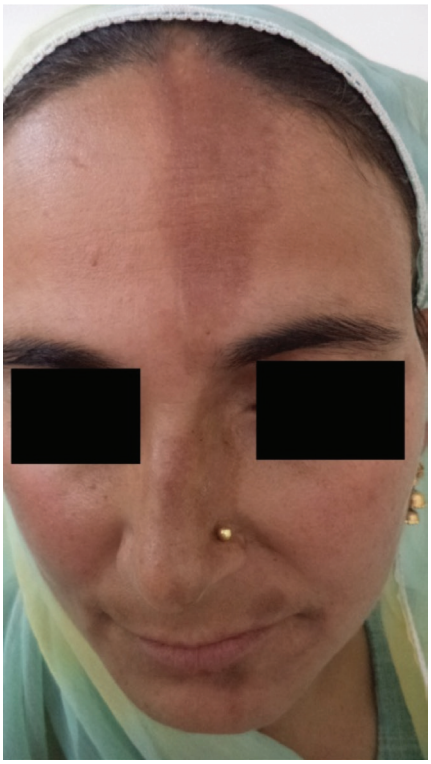

Figure 1. 'En coup de sabre'. Well defined, brownish hyper pigmented, indurated plaques over the frontal region of the scalp, forehead, and face in a vertical fashion.

Scleroderma is a disease of unknown etiology characterized by increased collagen deposition in the skin and other tissues. It can be localized (localized scleroderma) to the skin or diffuse (systemic sclerosis). The localized form, also called morphea, is characterized by predominant involvement of the skin; it occasionally involves the underlying muscles and usually spares the

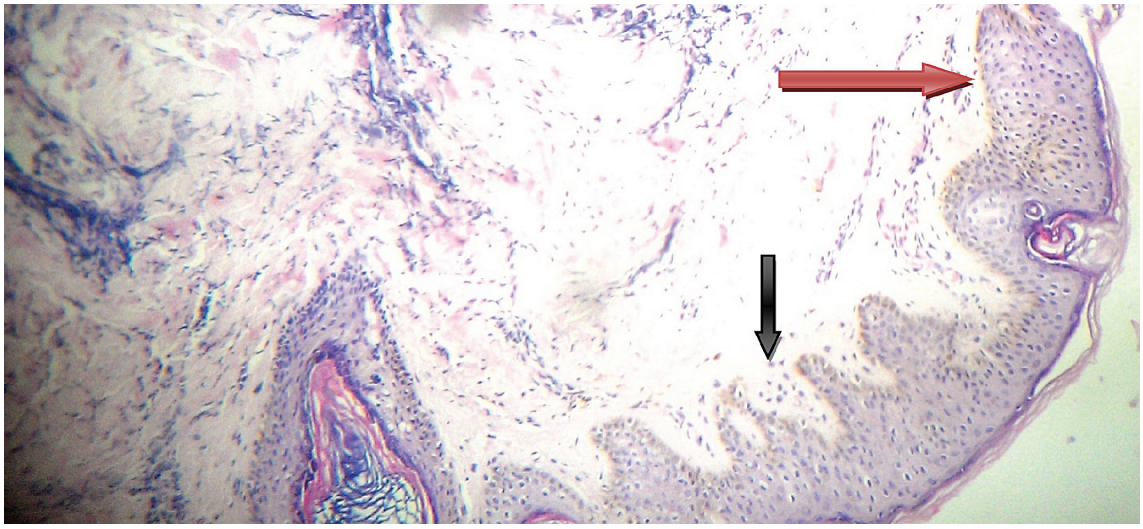

Figure 2. Morphea showing epidermal atrophy with loss of rete ridges (brown arrow) and a dermal mononuclear cell infiltrate. Note this biopsy has been taken from the edge of the lesion to keep relatively normal skin with normal rete ridges (black arrow) as control (H and E, × 100).

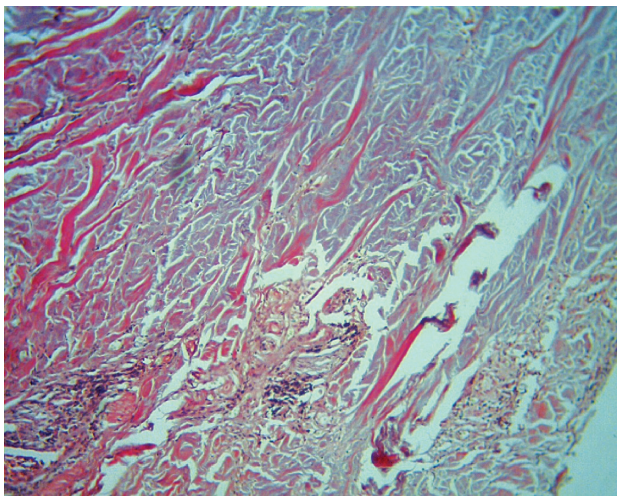

Figure 3. Morphea showing thick collagen bundles arranged parallel to each other in the reticular dermis (H and E, × 400)

internal organs. Morphologically, morphea has been divided into five types: plaque, linear, en coup de sabre, generalized, and pansclerotic [1,2]. It was Addison in 1854 who first coined the term 'en coup de sabre' as the lesion resembles the scar resulting from a saber sword cut [3]. En coup de sabre presents in a band-like fashion on the frontoparietal region, extending into the scalp. The skin lesions may extend downwards to the nose, cheek, upper lip, and involve the mouth and gums. However, in severe cases, lesions extending to the chin and neck have been reported [4]. The disease usually has an active stage lasting 2 to 5 years. Relapse of the disease activity after it has been quiescent for decades has not been reported previously in the literature to the best of our knowledge, which makes our case a unique one.

Although the specific etiology of morphea is unknown, several triggering factors have been incriminated, including trauma, immobilization, Bacille Calmette–Guérin (BCG) vaccination, injections of vitamin K, mechanical compression from clothing, previous radiotherapy, etc. [5]. In our case, trauma was the triggering factor.

Many neurological abnormalities may be associated with en coup de sabre, including headache, movement disorders, epilepsy, focal neurological deficits, and intellectual deterioration [6]. However, in our case, there were no associated neurological abnormalities.

The treatment options for morphea include topical tacrolimus, phototherapy, calcipotriol in combination with betamethasone dipropionate, imiquimod, methotrexate in combination with systemic steroids, photophoresis, D-penicillamine, etc [7]. Our patient was treated with topical tacrolimus 0.1% ointment applied twice daily. After a follow-up of two months, no new skin lesions appeared and the existing lesions showed reduction in skin thickening, induration, and hyperpigmentation.

The clinical course of en coup de sabre is very uncertain and can progress even after decades of disease inactivity. Therefore, proper follow-up is recommended in such patients.

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