Linear discoid lupus erythematosus mimicking en coup de sabre morphea: A case report

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Abstract
Discoid or chronic lupus erythematosus is an autoimmune disease that produces skin lesions on the face and scalp. Rarely do lesions present with linear configuration, but when they do, the lesions often follow the lines of embryologic migration. A 24-year-old man presented with a slowly progressing asymptomatic violaceous linear patch running from the root of his frontal scalp to the nasal tip. A Doppler ultrasound and skin biopsy were performed and the histological findings demonstrated characteristic findings of discoid lupus erythematosus. A full physical examination, review of systems and laboratory investigations showed no indication of systemic lupus. High potency topical steroids and calcineurin inhibitors were prescribed along with photoprotection. At 4-month follow-up, all his lesions had mostly cleared. We report here the first case, to our knowledge, of discoid lupus erythematosus with en coup de sabre presentation mimicking morphea.

Keywords
Lupus erythematosus, discoid lupus erythematosus, en coup de saber, linear lupus

Introduction
Lupus erythematosus is an autoimmune disease that can be skin limited or systemic. Discoid lupus erythematosus (DLE) is the most common variant of cutaneous lupus. While it has a lower chance (5%–10% life time) compared to other subtypes to progress to systemic forms, it has significant morbidity because of its tendency to involve cosmetically important areas such as the face and scalp and resulting disfiguring scarring alopecia.¹ Similar to other subtypes of lupus, DLE is most commonly seen in women and in the third to fourth decade of life.² DLE typically presents as erythematos to violaceous papules or plaques that eventually develop scales, follicular plugging, central hypopigmentation and scarring in photo-distributed areas.³ A localized form of the condition occurs in 80% of the patients with lesions developing solely on the face, ears, and/or scalp and 20% of patients present a disseminated form of the disease defined by the presence of lesions beyond the head and neck area.² Rarely, discoid and other forms of cutaneous lupus may present in linear fashion following the lines of embryologic migration (lines of Blaschko).⁴ We report, the first to our knowledge, case of linear lupus with en coup de sabre presentation mimicking acute morphea.

Case report
A 24-year-old non-smoking middle eastern male presented to dermatology clinic with a history of slowly progressive asymptomatic linear skin eruption. On clinical examination, a sharply demarcated pink to violaceous linear patch was noted extending from the root of his frontal scalp to the nasal tip (Figure 1(a)–(b)). Given the clinical presentation, a Doppler ultrasound and a skin biopsy were performed to rule out vascular etiology and early en coup de sabre morphea. However, the histological picture was diagnostic of discoid lupus showing characteristic features of basal vacuolation, follicular plugging, lichenoid infiltrate and mucin

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deposition (Figure 1(c)). Full body examination, review of systems and laboratory investigations did not reveal any features of systemic lupus. The patient was prescribed an alternative day regimen of high potency topical steroid (clobetasol dipropionate 0.05% cream) and calcineurin inhibitor (tacrolimus 0.1% ointment) and strict photoprotection to halt inflammation and prevent scarring. He was seen in follow-up 4 months later, at which time his lesions had mostly cleared.

Discussion
Linear forms have been described in many morphological subtypes of lupus erythematosus, such as subacute, discoid, panniculitis and profunda. Various attempts have been made at explaining the linearity of these lesions as well as their tendency to follow the lines of Blaschko. For example, linear nevoid and linear inflammatory dermatosis both follow Blaschko lines and both show evidence of genetic mosaicism as a potential cause. Therefore, antigenic effects mediated by T-cell activity targeted at the mosaic cells could produce the observed skin lesions.5 Others have postulated that the linear lesions may be due to microchimerism that specifically targets skin cells in a mechanism like that of graft-versus-hosts disease.5 However, the predominate view is that the presence of the linear lesions is caused by genetically unique keratinocytes that, when exposed to ultraviolet light, produce an inappropriate cytokine response.

Although overlap syndromes that fulfill the diagnostic criteria of both DLE and morphea have been described, the case we present illustrates a case of mimicry. Upon skin biopsy, only classic histological findings of DLE, such as follicular plugging, increased mucin, basement membrane thickening and perivascular lymphocytic inflammation with involvement of adnexal elements were observed.6 This knowledge had clear implications on our choice of treatment and further emphasizes the importance and near-necessity of performing skin biopsies and subsequent histopathological analysis when using other mainstay methods of diagnosing cutaneous maladies.

Cutaneous lupus erythematosus is an autoimmune skin disease favoring sun exposed skin. The most common form is DLE. Similar to other subtypes of lupus, DLE may rarely follow the lines of Blaschko. We report here, for the first time, a case of linear DLE mimicking en coup de sabre morphea.

Figure 1. (a) Erythematous plaque runs from the root of the frontal scalp down to the glabella. Two red and one purple papules are present in the region of the glabella, as well (white arrows). (b) The erythematous plaque appears to continue, after a brief interruption, down the left dorsum of the nose (black arrows). (c) Periodic acid–Schiff (PAS) staining of skin biopsy shows follicular plugging, vacuolar degeneration of the basal layer and lymphocyte predominant periadnexal inflammation.
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Informed consent
Patient consent was obtained for publication of this case report.

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