En Coup de Sabre

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PRACTICE **POINTS**

- En coup de sabre (ECDS) is a rare subtype of linear scleroderma that is limited to the hemiface in a unilateral distribution.
- Neurologic involvement is common and should prompt a comprehensive neurologic workup in patients suspected to have ECDS or progressive hemiface atrophy.
- Corticosteroids remain the treatment of choice, but other modalities such as methotrexate, excimer laser, and grafting have been used with varying success.

En coup de sabre (ECDS) is a rare form of localized scleroderma that typically manifests in children and women. It presents as a fibrous pansclerotic plaque extending in a bandlike distribution on the frontoparietal scalp with surrounding scarring alopecia. Many patients have comorbid central nervous system involvement in addition to the cutaneous findings. En coup de sabre is a rare entity that should be delineated from Parry-Romberg syndrome, as both entities share some common features and may coexist. Corticosteroids remain the treatment of choice, but other modalities such as methotrexate, excimer laser, and grafting have been used with varying success. We report a case of an elderly woman who presented with an asymptomatic alopecic plaque consistent with ECDS.

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En coup de sabre (ECDS) is a rare subtype of linear scleroderma that is limited to the hemiface in a unilateral distribution. The lesional skin first exhibits contraction and stiffness that lead to characteristic fibrotic plaques with associated linear alopecia.[1] The pansclerotic plaques are ivory in color with hyperpigmented to violaceous borders extending as a paramedian band on the frontoparietal scalp.[2,3] The skin lesions bear resemblance to the stroke of the sabre sword, giving the condition its unique name. Many patients initially present with concerns of frontal scalp alopecia.[3] Linear morphea, including the ECDS subtype, is predominantly seen in children and women, usually presenting within the first 2 decades of life.[1,4]

The differential diagnoses of ECDS include focal dermal hypoplasia, steroid atrophy, localized morphea, and lupus profundus.[5] En coup de sabre should be distinguished from progressive hemifacial atrophy (PHA) (also known as Parry-Romberg syndrome).[6] Progressive hemifacial atrophy presents as unilateral atrophy of the face involving skin, subcutaneous tissue, muscle, and underlying bone in the distribution of the trigeminal nerve.[1] Both PHA and ECDS exist on a spectrum of linear scleroderma and may coexist in the same patient.[6]

There is a strong association with extracutaneous neurologic involvement, including seizures, ocular abnormalities, trigeminal neuralgia, and headache.[7-10] One study examining ECDS and PHA demonstrated that 44% (19/43) of patients who underwent central nervous system imaging had abnormal findings.[11] The majority of patients had magnetic resonance imaging with or without contrast, computed tomography, or both. The most common findings on T2-weighted images were white matter hyperintensities, mostly in subcortical and periventricular regions. The findings were bilateral in 61% (11/18) of patients and ipsilateral to the lesion in 33% (6/18) of patients.[11] We present a case of ECDS masquerading as alopecia in a 77-year-old woman.

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The authors report no conflict of interest.

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Case Report

A 77-year-old white woman presented with a chief concern of hair loss on the scalp that had been present since 12 years of age. During her adult life, the scalp lesion remained unchanged with no associated symptoms. Her medical history was remarkable for hypertension and non–insulin-dependent diabetes mellitus. The patient denied any history of seizure disorders, facial paralysis, or neurologic deficits. Physical examination revealed a 13.6-cm linear, alopecic, hyperpigmented plaque extending from the left forehead and temporal scalp to the posterior occipital scalp with notable loss of underlying subcutaneous tissue (Figure). The left temporal scalp and forehead demonstrated pronounced atrophy with overlying telangiectases. Minimal depression was observed in the temporal scalp and forehead. The left cranial bone was easily palpable with no underlying subcutaneous tissue present. Computed tomography of the brain revealed no underlying skull or soft tissue abnormalities. No treatment was initiated, as the lesion was of minimal concern to the patient.

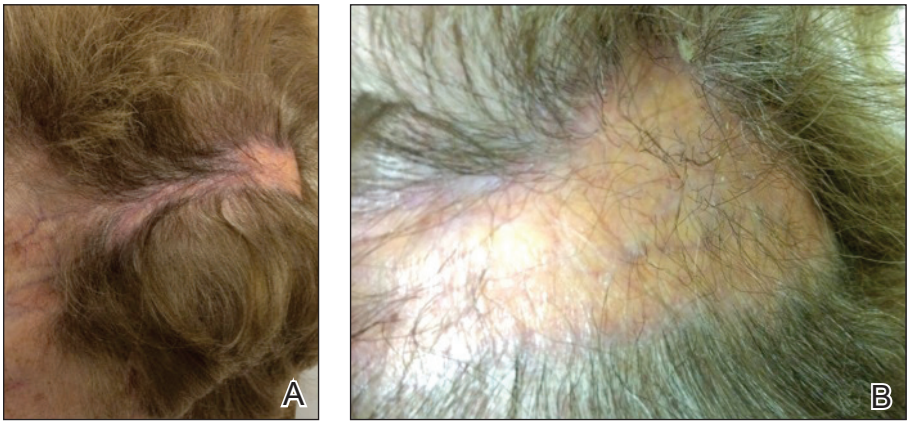
Comment

Etiology and Presentation—En coup de sabre is a rare subtype of linear morphea that involves the frontoparietal scalp and forehead.[7,12,13] It manifests as a solitary, linear, fibrous plaque that involves the skin, underlying muscle, and bone.[7] Although most cases present with a single lesion, multiple lesions can occur.[8] The exact etiology of this disease remains to be determined but is characterized by thickening and hardening of the skin secondary to increased collagen production.[7] The incidence of linear morphea ranges from 0.4 to 2.7 cases per 100,000 individuals and is more prevalent in white patients and women.[14] Linear morphea is commonly found in children. Children are more likely to have linear morphea on the face, which can lead to lifelong disfigurement.[2] Although the disease peaks in the fourth decade of life for adults, most pediatric cases are diagnosed between 2 and 14 years of age.[14-16]

Pathogenesis—Clinical and histopathological data suggest that a complex interaction among the vasculature, extracellular matrix, and immune system plays a role in the pathogenesis of the disease. Similar to scleroderma, the CD4 helper T cell may be involved in the fibrotic changes that occur within these lesions.[17] Early in the disease process, T_H1 and T_H17 inflammatory pathways predominate. The late fibrotic changes seen in scleroderma are more associated with a shift to the T_H2 inflammatory pathway.[17] Infection with *Borrelia burgdorferi* has been implicated abroad, but a large-scale study confirming *Borrelia* as a pathologic factor within morphea lesions has not been completed to date.[18-20] Some authors believe early lesions of ECDS mimic erythema chronica migrans, with the late lesions resembling acrodermatitis chronica atrophicans.[20]

Histopathology—Histopathologic findings of morphea tend to vary depending on the stage of the disease. The 2 stages of morphea can be differentiated by the degree of inflammation present histologically.[14,21] The early phase of morphea primarily affects the connective and subcutaneous tissue surrounding eccrine sweat glands.[14,21] A dense dermal and subcutaneous perivascular lymphocytic infiltrate with a mixture of lymphocytes, plasma cells, and histiocytes is commonly observed.[5] Later stages of the disease demonstrate densely packed homogenous collagen with minimal inflammation and loss of eccrine glands and blood vessels.[14,21] The adipose tissue is generally replaced by sclerotic collagen, giving the biopsy a squared-off appearance.[5,14]

Management—En coup de sabre presents a treatment challenge. In active lesions, topical or intralesional corticosteroids are considered treatment of choice.[5] Methotrexate has proven useful in the treatment of acute and deep forms of linear morphea. A study examining methotrexate in juvenile localized scleroderma, with the majority of patients having the linear subtype, revealed that methotrexate is both efficacious and well tolerated.[22] Other reports in the literature reveal efficacy with the use of intravenous corticosteroids



A and B, Paramedian, linear, alopecic, hyperpigmented plaque extending from the left frontal forehead and temporal scalp to the posterior occipital scalp characteristic of en coup de sabre.

and methotrexate combination therapy for treatment of morphea.[23,24] A longitudinal prospective study examining the use of high-dose methotrexate and oral corticosteroids for the treatment of localized scleroderma yielded positive results, with patients showing clinical improvement within 2 months of initiation of combination therapy.[25] Other treatments include excimer laser; calcipotriene and tacrolimus; and surgical approaches such as autologous fat grafting, grafting with muscle flaps, and tissue inserts.[21,26-31] In addition, patients can choose to forego therapy, as was the case with our patient.

Conclusion

En coup de sabre is a rare subtype of linear scleroderma that is limited to the ipsilateral scalp and face predominately in children and women. Neurologic involvement is common and should prompt a comprehensive neurologic workup in patients suspected to have ECDS or PHA. Current treatment recommendations include topical, intralesional, and oral corticosteroids; methotrexate; and surgical grafts. Although ECDS is a rare entity, more intensive research is needed on the exact pathophysiology and effective treatment options that focus on improving the cosmetic outcome in these patients. Cosmesis is the primary concern in patients with ECDS and should be managed early and appropriately to prevent long-term psychological sequelae.

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