

What Is Your Diagnosis?



A 9-year-old Bolivian boy presented with progressive facial changes that had developed at 2 years of age. The lesions were entirely asymptomatic and nonresponsive to any form of topical treatment. The patient's mother reported that the child was dropped on his head prior to onset, which she believed was the cause. Family history was noncontributory.

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Casey Bowen, MD; Louisiana State University Health Sciences Center, Shreveport.

Todd T. Kobayashi, MD; US Air Force Academy, Colorado Springs, Colorado.

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The Diagnosis: Segmental Vitiligo and En Coup de Sabre



Vitiligo is a relatively common skin disease characterized by the progressive development of white macules and patches. Our patient demonstrated a classic depigmented patch along his hairline. Two commonly classified subtypes of vitiligo exist: generalized and localized. Localized lesions that follow a dermatomal distribution and do not cross the midline can be further classified as segmental vitiligo.^{1,2} Absence of epidermal melanocytes is the predominant histologic finding in areas of depigmentation. While the precise cause of this melanocyte loss is unclear, the observation of circulating antimelanocytic antibodies and lymphocytic infiltrations at the margins of lesions in a preponderance of cases supports the theory of autoimmune-mediated destruction.³⁻⁷

First described by Addison⁸ in 1854, en coup de sabre is a rare form of morphea/localized scleroderma classified as linear morphea. Morphea is a relatively benign, well-circumscribed variant of scleroderma characterized by the presence of thickened and sclerotic collagen bundles in an indurated

cutaneous lesion. Homogenization of the collagen bundles occurs in the dermis and may involve the panniculus and fascia. Atrophy of skin appendages occurs as collagen increases, and perivascular infiltrates (ie, histiocytes, eosinophils, lymphocytes, plasma cells) are commonly present. En coup de sabre is manifested as an indurated or depressed segment of skin on the frontoparietal region of the face and/or scalp that respects the midline and resembles the stroke of a sword.⁸⁻¹¹ In time the lesion may progress to involve atrophy of one side of the face, giving the appearance that the stroke of the sword removed some of the tissue, which may then be considered facial hemiatrophy or Parry-Romberg syndrome. En coup de sabre and Parry-Romberg syndrome frequently coexist; although the precise relationship is unclear at this time, it is likely they are both variants of morphea.^{12,13} Our patient demonstrated features of both variants. Clinical features of en coup de sabre included sclerotic dermal tissue on the cheeks and perioral area that was easily

palpable but difficult to appreciate by photograph, hyperpigmentation, and normal ocular and bony growth. Although Parry-Romberg syndrome classically does not have notable cutaneous involvement, the patient did exhibit facial hemiatrophy.

Vitiligo and morphea have a similar epidemiology, and while the etiology of both is unknown, they are both postulated to be autoimmune in nature. Both vitiligo and morphea have been reported in association with many organ-specific autoimmune diseases: vitiligo with Hashimoto thyroiditis, pernicious anemia, Addison disease, hypoparathyroidism, myasthenia gravis, autoimmune hemolytic anemia, and regional enteritis,¹⁴⁻¹⁷ and morphea with Hashimoto thyroiditis, thyrotoxicosis, hypothyroidism, and pernicious anemia.^{16,18}

While vitiligo and morphea are relatively common, their simultaneous occurrence rarely has been reported.^{14-17,19-21} Although it is possible that the occurrence of each lesion is unrelated, the pattern reported in this case suggests otherwise. The simultaneous clinical presentation and codistribution of both vitiligo and morphea in this case seem to fortify the theory that these 2 separate clinical entities are autoimmune in nature and may be related. The history of head trauma in our patient may provide further insight, as linear morphea (scleroderma) and vitiligo have both been associated with trauma, nerve injury, and sympathetic dysfunction.^{1,17,22}

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