

En Coup De Sabre: A Case Report

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Abstract

En coup de sabre is an unusual variant of localized scleroderma defined by its distinct location involving the frontoparietal region of the forehead and scalp. We report a rare case of en coup de sabre in a 16-year-old female with vitamin D deficiency. The workup, management, and relationship between scleroderma and serum vitamin D concentrations will be discussed.

Figure 1



Figure 2

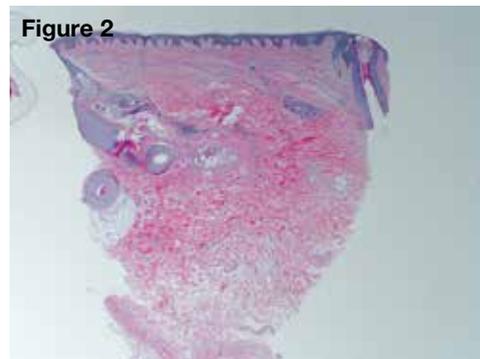
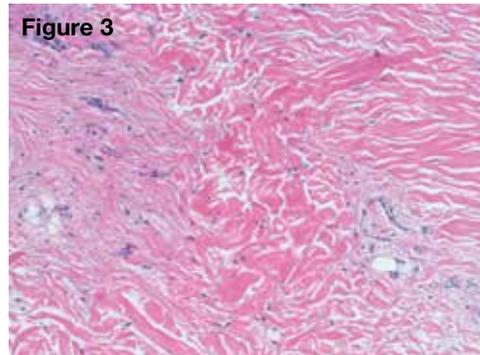


Figure 3



Introduction

Scleroderma is a rare connective-tissue disorder of unknown etiology. It is characterized by increased collagen production resulting in dermal thickening and hardening of the skin. Scleroderma involves a wide range of disease. It can include systemic involvement of the internal organs, referred to as systemic sclerosis, or it can be confined to the skin in a localized form, referred to as morphea or localized scleroderma. En coupe de sabre is a rare craniofacial subtype of localized scleroderma.

Case Report

A 16-year-old female presented to our clinic with a one-year history of hair loss involving the right side of her scalp. She denied any associated symptoms such as headaches, vision changes or trauma to the scalp. The patient had no medical history and no significant family or drug history.

On physical examination, there was a hyperpigmented, atrophic plaque extending from the vertex of her scalp down to the right side of her forehead (Figure 1). The patient's complete blood count (CBC) and comprehensive metabolic panel (CMP) were unremarkable, but the vitamin D level was decreased at 16 ng/ml. Autoimmune serology was performed, and patient was negative for ANA, DSDNA, SSDNA, SCL-70, anti-

centromere, SS-A, SS-B, C3, and C4.

A biopsy was taken from the vertex of the scalp. Histopathology showed thick, dense hyalinized collagen bundles in the dermis (Figure 2). There was a sparse, deep lymphocytic infiltrate that did not extend into the subcutaneous fat (Figure 3). The patient's clinical history, physical examination, laboratory results and histopathologic findings were consistent with linear scleroderma en coupe de sabre, a rare subtype of localized scleroderma.

Our patient received a prednisone taper, which began at 60 mg daily and eventually tapered down to 10 mg daily over a period of one month. Concurrently, the patient began methotrexate

Figure 4



15 mg once a week with folic acid 1 mg every day except the day methotrexate was taken. Vitamin D supplementation was not given to our patient. Our clinic did not have UVA/PUVA or narrowband UVB therapy available, but the patient was referred to a clinic that did have these modalities. The patient refused these therapeutic options. This treatment plan yielded good results, with the patient and physician both noting less induration of the lesion after three months of methotrexate. The patient's lesion did not show much improvement visually (**Figure 4, p. 47**), but on palpation there was significant softening of the plaque.

Discussion

Scleroderma is a rare connective-tissue disorder in which increased collagen production leads to thickening and hardening of the skin. In systemic sclerosis, there is systemic involvement of internal organs; in morphea, or localized scleroderma, the disease is confined to the skin. Both localized and systemic forms are characterized by fibrosis of the skin. Localized scleroderma is further divided into five main subtypes on the basis of clinical appearance and distribution.¹ These include plaque, bullous, generalized, deep, and linear scleroderma.

The incidence of localized scleroderma ranges from 0.4 to 2.7 per 100,000 people.² Though all races may be affected, there is an increased prevalence among Caucasians, accounting for 72% to 82% of patients.² There is a female predominance in all subtypes of localized scleroderma except for linear scleroderma, in which males and females are equally affected.¹ There is a similar distribution among children and adults. In adults, the incidence peaks in the fifth decade of life. In children, 90% of patients are diagnosed between the ages of 2 and 14 years.³⁻⁶

Though the pathophysiology of scleroderma has yet to be fully elucidated, it is postulated that the fibroblasts of patients with scleroderma produce increased levels of extracellular matrix components including collagen, elastin, fibronectin and glycosaminoglycans.⁷ This elevated fibroblast activity can also lead to increased levels of signaling and transcription molecules, including IL-6, pro-IL-1 alpha and ICAM-1.⁷ This culminates in increased deposition of extracellular matrix components, resulting in hardening of skin.

En coup de sabre is an unusual variant of linear scleroderma and is defined by its distinct location involving the frontoparietal region of the forehead and scalp. The term "en coup de sabre" was originally coined by the French to depict the wounds inflicted on foot soldiers who were struck on the head with a sword, which resulted in a thickened scar on one side of the forehead.

Clinically, the lesion presents as a linear, band-like, ivory-colored, sclerotic plaque with violaceous borders. The violaceous border often surrounds the indurated plaque and has been described as a lilac ring. En coup de sabre is also characterized by atrophy and furrowing of the skin. Alopecia

may be present in the area of involved scalp, which can be the presenting complaint. Typically, the lesions are confined to the skin and subcutaneous tissue; occasionally, underlying muscle, cartilage and bone can also be involved, resulting in facial atrophy. When hemifacial atrophy occurs, Parry Romberg syndrome (PRS) should be in the differential diagnosis. It has been reported that PRS coexists in 20% to 37% of patients diagnosed with en coupe de sabre.⁸ There have been many discussions as to whether these are two distinct diseases or clinical variants of the same disease. The ailments have comparable ages of onset and disease progressions. PRS may have dermatologic findings similar to those seen in en coupe de sabre, but they are typically more prominent and do not exhibit cutaneous sclerosis at any stage of the disease.

On rare occasions, localized scleroderma has been associated with multisystemic involvement, including rheumatologic, ophthalmologic and neurologic manifestations, which occur in about 20% of cases.⁹ Onset of cutaneous disease precedes extracutaneous manifestations. Neurologic abnormalities occur most commonly in association with en coupe de sabre, and of these, complex partial seizures occur most frequently.¹⁰ Radiologic anomalies are predominantly ipsilateral to the skin lesion. Neuroradiologic abnormalities can involve white-matter lesions, cerebral atrophy, intraparenchymal calcification, meningocortical variations and skull atrophy.¹¹ CT scan of the brain can show thinning of the skull under the cutaneous lesions. MRI of the brain may show focal cerebral atrophy and blurring of the gray-white matter.⁹ A gadolinium-enhanced MRI of the brain has been recommended for all patients with neurologic symptoms.¹² Amaral et al. suggest that all patients, regardless of symptoms, should be considered for neuroimaging studies at the time of diagnosis given that a subset of patients will have neurological damage without displaying clinical signs.⁹

The pathogenesis of scleroderma is not entirely known. Evidence suggests it is autoimmune in nature and initiated by a provocative event, most commonly local trauma to the skin. Subsequent endothelial-cell damage leads to an increase in fibroblast activity and ischemia secondary to narrowing of the lumen and alteration in collagen production.¹¹ There have been reports of a positive association between *Borrelia* infection and scleroderma, but there has been no evidence of *Borrelia* infection in scleroderma lesions.¹³

Patients may also have elevated autoantibodies, most frequently anti-nuclear antibody and anti-single stranded DNA (ssDNA) antibody. Rheumatoid factor, anti-histone antibody, anti-phospholipid antibody and anti-topoisomerase II antibody may also be elevated, but these are seen more commonly in generalized morphea.¹³ There is no autoantibody that correlates with disease activity.^{14,15}

It has also been noted that patients with scleroderma have lower serum levels of vitamin D. It is not entirely clear whether this is an incidental

finding or reflects a true association between vitamin D levels and clinical manifestations of scleroderma. Recent studies, however, have shown that individuals with vitamin D deficiency are at a higher risk of developing autoimmune diseases.¹⁶ It is postulated that the relationship between vitamin D and autoimmune disease has to do with vitamin D's immunomodulating activity on vitamin D receptors present on antigen-presenting cells and activated T cells.¹⁷ In 2011, a study conducted in Hungary to find a link between scleroderma and serum vitamin D levels concluded that patients with scleroderma do have considerably lower serum concentrations of vitamin D when compared to controls, and that cutaneous fibrosis is inversely related to serum vitamin D concentrations.¹⁸ This relationship between vitamin D and autoimmune disease suggests that vitamin D may be a modifiable environmental factor in patients with scleroderma.

Treatment and Prognosis

En coupe de sabre is typically a self-limited disease in children. There can be softening or regression of skin lesions, but complete resolution seldom occurs and reactivation is always possible.¹⁹ The long-term prognosis of en coupe de sabre in children is generally excellent. In most cases, the cosmetic defect is minimal and can be covered by the patient's hair. Adults tend to have a more variable course, with some patients clinically deteriorating over time. This may result in severe contractures.

Studies have found that early intervention during the active phase of the lesions is most beneficial. For initial management, it has been recommended to use methotrexate and systemic glucocorticoids, followed by UVA1 with or without psoralens, narrow-band UVB, or mycophenolate mofetil.^{20,21} Other treatment options include topical and intralesional glucocorticoids, vitamins E and D₃, D-penicillamine, antimalarials, retinoids, and immunosuppressive agents such as cyclosporine and cyclophosphamide.¹¹

Ultraviolet A light (UVA), with or without psoralens, is an effective therapeutic option. UVA1, which is a specific wavelength range of UVA, offers particularly deep skin penetration and is believed to soften the plaques by two mechanisms: causing systemic immunosuppression, and inducing enzymes that degrade the collagen matrix in the skin.

In cases where the lesions are disfiguring and the patient pursues cosmetic improvement, surgical repair is a corrective option. Narrow lesions can be treated with simple excision followed by primary closure. Wider lesions may necessitate a more elaborate reconstruction, which can include an array of flaps, implants or autologous fat or bone transplantation. Due to the invasiveness of these procedures, the use of fillers, such as hyaluronic acid, have been employed as an alternative.²²

Conclusion

En coup de sabre is a rare connective-tissue

disorder, identified by its distinct location on the frontoparietal scalp and forehead. Early intervention during the active phase of the disease has shown maximum benefit in overall outcomes in these patients. Scleroderma has been associated with low levels of vitamin D. Further studies are needed to elucidate whether there is a causative relationship between vitamin D deficiency and scleroderma. This could give insight into whether normalizing vitamin D levels would modify the disease progression, potentially presenting a new therapeutic focus.

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