Letter to the Editor

A pediatric case of scleroderma en coup de sabre and segmental vitiligo

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Sir,

Scleroderma *en coup de sabre* is a rare subtype of localized scleroderma in children that affects the face. Localized scleroderma is an autoimmune inflammatory sclerosing disorder, and thus the co-existence of secondary autoimmune disorders has been reported [1]. Herein, we report a rare pediatric case of the co-existence of scleroderma *en coup de sabre* and segmental vitiligo.

A 14-year-old female was referred to our department complaining of a facial lesion without a family history of connective tissue disease. A physical examination showed slightly reddish and slightly depressed longitudinal plaques on the forehead extending onto the scalp (Fig. 1). However, sclerotic lesions were not observed on the trunk and extremities. Hemiatrophy was not observed on the face. The patient had no medical history of seizures or epilepsy. Further examination revealed segmental vitiligo involving the left lower extremity (Fig. 2), but no vitiliginous lesions other than on the left leg. A biopsy specimen from the forehead revealed thickened collagen fibers in the lower dermis, and a specimen from the thigh revealed a loss of epidermal melanocytes. A laboratory examination detected antinuclear antibodies (1:80) but no anti-centromere or anti-Scl-70. Oral prednisolone 20 mg/day was administered, which was gradually tapered and ceased over the next three months. The facial erythematous lesion was sufficiently improved, whereas the depigmentation on the lower extremity remained unaffected.

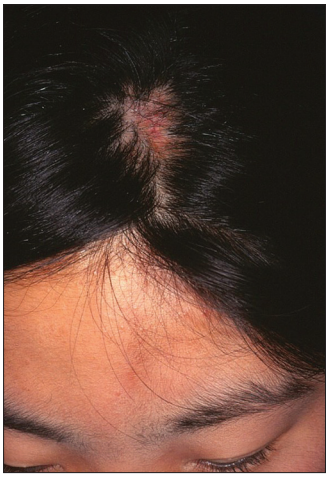

Figure 1: An en *coup de sabre* morphea on the forehead involving the scalp.

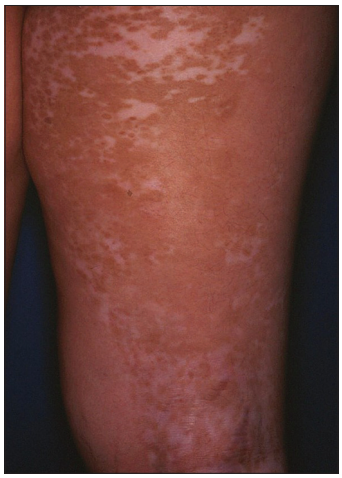

Figure 2: A depigmentation on the left lower extremity.

The co-existence of localized scleroderma and vitiligo is extremely rare, and the association of linear scleroderma with homolateral segmental vitiligo has rarely been reported [2]. Among 44 patients with linear scleroderma, vitiligo was noted in only one [3]. On the other hand, among 701 patients with childhood vitiligo, there were no cases of connective tissue disease [4]. Scleroderma *en coup de sabre* is a rare type of linear scleroderma affecting

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the forehead and sometimes the scalp in young people and impairing the patient's quality of life. Occasionally, the tissues deeper than the subcutis are involved, leading to functional limitations [5]. In a case reported by Ubaldo and Castro, a cleft lip was also observed on the same side with the lesions on the face, which may have been caused by embryological cell division, suggesting mosaicism [2]. In our case, linear scleroderma appeared on the face, whereas segmental vitiligo involved the lower extremity. Several pathogenic hypotheses of segmental vitiligo have been proposed, including a neuronal mechanism, somatic mosaicism, and microvascular skin homing, although without certainty as to whether or not leading to autoimmune destruction. In addition to trauma, neurological and infectious agents and immunological abnormalities have been postulated as the causative agents of morphea. Various pathogeneses have been proposed for these two entities, with the autoimmune mechanism common to both, although cutaneous mosaicism in linear scleroderma may not be confirmed on a molecular basis [1]. In conclusion, although the co-occurrence of scleroderma *en coup de sabre* and segmental vitiligo can be caused by sheer chance, the immunological, developmental, and neurological factors may have played a major role in our patient.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients have given consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

REFERENCES

1. Torok KS, Li SC, Jacobe HM, Taber SF, Stevens AM, Zulian F, et al. Immunopathogenesis of pediatric localized scleroderma. Front Immunol. 2019;10:908.
2. Ubaldo HDC, Castro CCS. Coexistence of segmental vitiligo, scleroderma en coup de sabre and cleft lip on the same hemiface: association with mosaicism? An Bras Dermatol. 2019;94:248-50.
3. Christen-Zaech S, Hakim MD, Afsar FS, Paller AS. Pediatric morphea (localized scleroderma): review of 136 patients. J Am Acad Dermatol. 2008;59:385-96.
4. Martins CPDS, Hertz A, Luzio P, Paludo P, Azulay-Abulafia L. Clinical and epidemiological characteristics of childhood vitiligo: a study of 701 patients from Brazil. Int J Dermatol. 2020;59:236-44.
5. Kunzler E, Florez-Pollack S, Teske N, O'Brien Jack, Prasad S, Jacobe H. Linear morphea: clinical characteristics, disease course, and treatment of the morphea in adults and children cohort. J Am Acad Dermatol. 2019;80:1664-70.

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