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Epidermolysis Bullosa Pruriginosa Associated with Folliculitis Decalvans: Case Report and Review of the Literature

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Dear Editor,

Epidermolysis bullosa (EB) represents a clinically heterogeneous group of genetic diseases characterized by skin fragility leading to blister formation after local trauma. EB pruriginosa (EBP) is a rare clinical variant of dystrophic EB presenting at birth or in early adulthood [1, 2]. Folliculitis decalvans (FD) is a primary neutrophilic scarring alopecia of unclear etiology affecting the scalp of young adults [3]. Even though the clinical aspects of both diseases have been well described, an association has never been reported in the literature.

We describe the case of a 31-year-old Caucasian woman who presented with pruritic blisters after mild trauma on the lower limbs, associated with milia, nail abnormalities, and lichenoid plaques since the age of 1 year. On physical examination, erythematous lichenified plaques with scaling, prurigo-like lesions and scattered milia were distributed on the lower legs, mainly on the extensor aspects (Fig. 1). Also, she presented anonychia on all toes and nail dystrophy on some fingers (Fig. 2). Both her mother and brother had similar skin lesions. When the patient was 20 years old, she developed papules and folliculitis-like lesions affecting the occipital and vertical scalp, which progressed to hair tufting and scarring alopecic areas (Fig. 3). Her teeth, mucosa, and musculoskeletal growth were normal. The histopathological examination of the vertex scalp area showed typical features of FD, such as follicular infiltrate with neutrophils, lymphocytes and plasma cells, hyperkeratosis, dermal fibrosis, and follicular tufts.

The clinical presentation in dystrophic EB can be very heterogeneous, displaying variable trauma-induced skin fragility, nail dystrophy, milia, atrophic scarring, granulation tissue, scalp abnormalities, keratoderma, and extracutaneous involvement [4]. EBP is a recently described form of dystrophic EB characterized by hypertrophic scarring and intense pruritus, a hallmark of the disease [5]. In this case, the mild clinical phenotype, presence of milia, dystrophic nails, and normal teeth and mucosa, besides similar family lesions, suggested the autosomal dominant subtype of EBP.

Fig. 1. **a** Erythematous and scaly patches, lichenified lesions on the limbs, and blisters on the pretibial areas. **b** Scales and scattered milia on the knee.

Work for this case report was conducted at the Department of Dermatology, Universidade de São Paulo, São Paulo, Brazil.

Fig. 2. **a** Anonychia on all toes. **b** Nail dystrophy on some fingers.

Fig. 3. **a** Ivory-white scarring patches, follicular tufting, and pustules on the vertex of the scalp. **b** Prominent follicular tufts, and white fibrotic areas devoid of follicular openings.

Folliculitis-like lesions were reported in one patient with EBP, a 18-year-old boy who developed multiple, painless, red follicular papules and pustules over the scalp. The histopathological examination of the scalp showed hyperkeratosis within the folliculitis-like lesions, with a flattened rete ridge near the orifice of the hair follicles, and a subepidermal cleft. Fibrosis was present, as well as a mild perivascular and perifollicular lymphohistiocytic infiltrate in the papillary dermis. However, no infiltration of neutrophils was observed around the hair follicles [6].

Almaani et al. [7] described a 40-year-old female patient with EBP and associated scalp alopecia. After 15 years of follow-up, she reported hair loss affecting the parietal, temporal, and frontal areas of her scalp. On scalp examination, a patchy scarring alopecia with perifollicular violaceous lesions could be noted in the parietal area. The histopathologic analysis of the scalp revealed perifollicular lymphocytic inflammation, mostly in the infundibuloisthmic regions, and perifollicular fibrosis, suggesting a diagnosis of lichen planopilaris. In this case, no blisters or signs of lichenoid inflammation were seen at the dermoepidermal junction.

In junctional and dystrophic forms of EB such as EBP, blisters usually involve the uppermost dermis, and the resultant cicatricial alopecia occurs due to the inflammatory process in the interfollicular epidermis and upper portion of the hair follicles [8]. Both recessive and dominant forms of dystrophic EB occur as a mutation in the *COL7A1* gene, which is responsible for encoding type VII collagen, a major structural component of anchoring fibrils at the dermoepidermal junction [4].

Different forms of alopecia have been reported in association with autoimmune bullous diseases, including tufted folliculitis [9], which is considered by some authors as a variant of FD. However, the only report associating a hereditary disease with FD was described in a patient with Darier disease, a rare disturbance of keratinization [10]. Although secondary scarring alopecia is very common in different forms of EB, especially in areas of trauma due to rubbing and repetitive blisters [8], to our knowledge, this is the first report of a patient presenting with EBP in association with FD, a primary neutrophilic scarring alopecia.

Statement of Ethics

The authors have no ethical conflicts to disclose. Written informed consent was obtained from the patient to have the details and photographs of her case published.

Disclosure Statement

The authors declare no conflicts of interest.

References

- 1 Schumann H: Epidermolysis bullosa. An Update (in German). *Hautarzt* 2009;60:614–621.
- 2 Smolyannikova VA, Kubanova AA, Albanova VI, Nefedova MA, Karamova AE: Current approaches to the morphologic diagnosis of different types of congenital epidermolysis bullosa (in Russian). *Arkh Patol* 2016;78:9–16.
- 3 Olsen EA, Bergfeld WF, Cotsarelis G, Price VH, Shapiro J, Sinclair R, et al: Summary of North American Hair Research Society (NAHRS)-sponsored Workshop on Cicatricial Alopecia, Duke University Medical Center, February 10 and 11, 2001. *J Am Acad Dermatol* 2003;48:103–110.
- 4 Ee HL, Liu L, Goh CL, McGrath JA: Clinical and molecular dilemmas in the diagnosis of familial epidermolysis bullosa pruriginosa. *J Am Acad Dermatol* 2007;56(suppl):S77–S81.
- 5 Bibi Y, Chuang G, Bush M, Barak O, Scheinman P: The elusive side of mechanobullous disease: epidermolysis bullosa pruriginosa. *J Am Acad Dermatol* 2009;60(suppl 1):AB96.
- 6 Fan YM, Yang YP, Li SF: Medical genetics: sporadic dystrophic epidermolysis bullosa with albopapuloid and prurigo- and folliculitis-like lesions. *Int J Dermatol* 2009;48:855–857.
- 7 Almaani N, Liu L, Perez A, Robson A, Mellerio JE, McGrath JA: Epidermolysis bullosa pruriginosa in association with lichen planopilaris. *Clin Exp Dermatol* 2009;34:e825–e828.
- 8 Tosti A, Duque-Estrada B, Murrell DF: Alopecia in epidermolysis bullosa. *Dermatol Clin* 2010;28:165–169.
- 9 Miteva M, Murrell DF, Tosti A: Hair loss in autoimmune cutaneous bullous disorders. *Dermatol Clin* 2011;29:503–509.
- 10 Choudry K, Charles-Holmes R, Vella EJ, Burge S: Scarring alopecia due to folliculitis decalvans in a patient with Darier's disease. *Clin Exp Dermatol* 2001;26:307–308.