Pterygium in bullous pemphigoid: An unusual complication

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Key words: bullous pemphigoid; nail involvement; pterygium unguis.

INTRODUCTION

Bullous pemphigoid (BP) is the most frequent autoimmune subepidermal blistering disease of the skin.[1] BP is associated with an autoantibody response against BP180 and BP230, 2 components of junctional adhesion complexes called *hemidesmosomes* in basal keratinocytes. BP predominantly affects elderly patients and manifests with a broad spectrum of clinical features.[1] Although lesions may be widespread, nail involvement is rarely described.[2] We report on a BP patient with lesions involving the nail unit, which resulted in a peculiar and as yet rarely recognized complication of BP, pterygium unguis.

Abbreviation used:

BP: Bullous pemphigoid

CASE REPORT

A 77-year-old man was admitted for evaluation and management of a generalized pruritic bullous eruption of 2 months' duration. He showed eczematous and urticarial-like lesions with vesicles and tense blisters on the trunk, abdomen, and upper and lower limbs as well as postbullous erosions and crusting but no lichenoid papules. Bullae were also present on the proximal nail fold of several fingers. Close inspection found that his right thumbnail was severely atrophic with a wide band of smooth skin spanning from the proximal nail fold to the nail bed (Fig 1). No longitudinal ridges, furrows, or splits typical for nail lichen planus were present. The patient reported having had large blisters involving the periungual region of the thumbs, which ultimately resulted in loss of these fingernails. While erosions and crusting persisted, the right thumbnail only grew 2 narrow lateral nail fragments. Light microscopy studies of a biopsy specimen obtained from the thigh found subepidermal blister formation with a dermal inflammatory cell infiltrate very rich in eosinophils but without hydropic degeneration of basal cells. Direct immunofluorescence of perilesional skin found linear deposits of IgG and C3 along the epidermal basement membrane zone. By enzyme-linked immunosorbent assay, the patient had circulating autoantibodies directed against BP180 (37.5 U/mL; normal, < 12 U/mL). The patient was given topical clobetasol propionate cream, once daily for 1 month and subsequently once every other day. This treatment induced a complete control and regression of the skin lesions and pruritus.

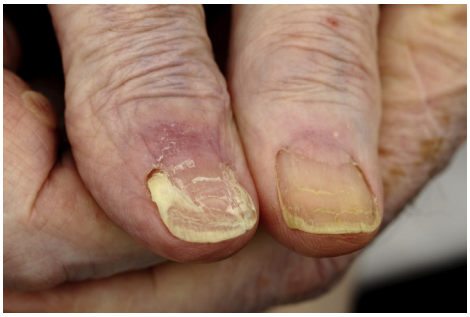

Fig 1. Pterygium in BP. The right thumbnail shows loss of the nail in its proximal median part with the skin of the proximal nail fold bridging to the nail bed causing a wide pterygium. The left thumbnail shows 2 transverse lines of horizontal splitting, which are analogous to Beau lines.

DISCUSSION

The immune response in autoimmune blistering diseases of the skin and mucosae potentially also targets structural components of the nail unit[1-3] and

From the Department of Dermatology, Inselspital, University Hospital of Berne.
Funding sources: None.
Conflicts of interest: None disclosed.
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JAAD Case Reports 2020;6:737-9.

2352-5126

https://doi.org/10.1016/j.jdcr.2020.05.026

Table I. Diseases observed or assumed to cause dorsal nail pterygium

Frequent
- Lichen planus
- Mucous membrane (cicatricial) pemphigoid
- Epidermolysis bullosa acquisita
- Trauma
  - Burns
  - Surgery
  - Onychotillomania, onychoteiromania
- Vasculopathy, diabetic angiopathy, Raynaud's disease

Rare
- Bullous pemphigoid*
- Chronic graft-versus-host disease
- Pemphigus foliaceus
- Systemic lupus erythematosus
- Toxic epidermal necrolysis
- Sarcoidosis
- Idiopathic
- Congenital
- Clouston syndrome
- Porokeratosis of Mibelli
- Linear porokeratosis
- Marfan syndrome
- Dyschromatosis universalis hereditaria

*Current case.

may result in tissue damage and nail alterations, such as blisters and erosions of the proximal nail fold, paronychia, and onychomadesis with involvement of the nail bed and matrix. Permanent nail loss may even occur.[2,4-8] In pemphigus, epidermolysis bullosa acquisita, in distinct forms of mucous membrane pemphigoid (previously reported as cicatricial pemphigoid) as well as in anti-p200 pemphigoid, nail alterations and dystrophy with scarring and nail loss have been reported at variable frequency, whereas such changes have only been anecdotally described in BP.[5-8] An important differential diagnosis of BP is lichen planus pemphigoides. Its peak incidence is in middle-age persons (significantly younger than in BP), and it usually shows blisters plus lichenoid papules. Lesions are predominantly found on the extremities, whereas those of BP are more generalized, often occur on erythematous and edematous skin, and are associated with pruritus.[9]

Pterygium formation, which is characteristically observed in ungual lichen planus and rarely in other conditions (Table I), represents another uncommon complication of autoimmune bullous diseases. Pterygium in nail lichen planus is likely caused by the cellular infiltrate invading the apical matrix and the proximal aspect of the ventral surface of the proximal nail fold where the nail stem cells are presumed to be located.[10] Its formation in autoimmune bullous diseases most likely results from extensive postbullous erosions and ulcers[1,4-8] that do not re-epithelialize rapidly enough to prevent adhesion of the base and the roof of the nail pocket (Fig 2). Pterygium has not been observed in bullous lichen planus of the nail,[11] but complete anonychia of involved nails was seen in a case of bullous-erosive nail lichen planus.[12] To our knowledge, pterygium formation has not yet been described in BP. Its presence should prompt one to consider not only lichen planus, but also BP, epidermolysis bullosa acquisita, cicatricial pemphigoid, as well as the group of hereditary epidermolysis bullosa.[9] In these cases, proper classification of affected patients depends on light microscopy studies, direct immunofluorescence microscopy, and the characterization of the targeted autoantigens. Our observation hence further extends the spectrum of nail alterations observed in the course of BP and identifies BP as a new cause for pterygium unguis formation.

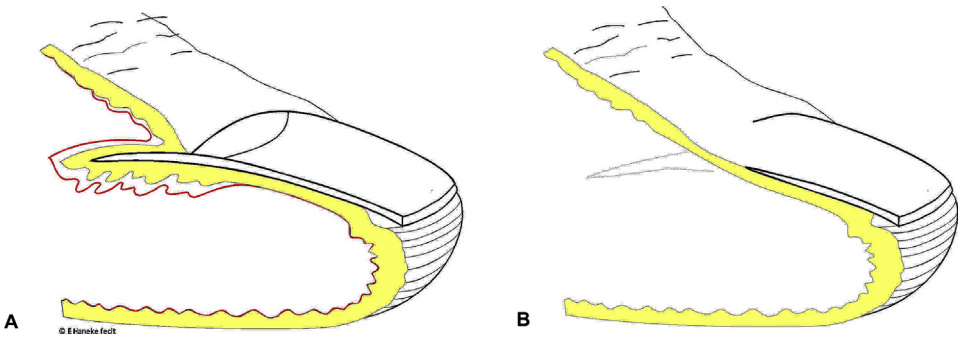

Fig 2. Pterygium in BP. Schematic illustration of the potential pathomechanism of pterygium formation in BP. **A**, There is extensive blister formation with detachment of the epidermis of the ventral surface of the proximal nail fold and the matrix epithelium. When healing of the denuded areas is faster than re-epithelialization, they grow together and cause the pterygium unguis. Further, the nail stem cells are located in the dorsal portion of the apical matrix and are thus potentially lost with such an extensive damage. **B**, End stage with obstruction of the nail pocket (*dashed line*) resulting in a cicatricial pterygium.

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