

## A young man with Pemphigus and ungueal changes

Homem jovem com Pênfigo e alterações ungueais

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### Abstract

Nail involvement in people affected by pemphigus vulgaris has been associated with the severity of disease. Paronychia, onychomadesis, Beau's lines and nail hemorrhages, are frequently reported. In the young patient herein described the conspicuous nail changes developed during the convalescent period of disease, after a long-term administration of azathioprine and sulfone. Nail changes can be reversible and these drugs are safe and useful to the pemphigus control. Case reports might contribute to better knowledge about phenomena not well understood yet.

**Key words:** Drug treatment, nail changes, pemphigus vulgaris

### Resumo

O envolvimento ungueal em portadores de pênfigo vulgar tem sido associado com a gravidade da doença. Paroníquia, onicomadese, linhas de Beau e hemorragias nas unhas, são frequentemente relatadas. No paciente jovem aqui descrito as alterações ungueais conspícuas desenvolveram durante o período de convalescença da doença, após administração a longo prazo de azatioprina e sulfona. Alterações das unhas podem ser reversíveis, e essas drogas são seguras e úteis para o controle de pênfigo. Relato de caso pode contribuir para um melhor conhecimento sobre os fenômenos ainda não bem compreendidos.

**Palavras chave:** Tratamento medicamentoso, alterações ungueais, pênfigo vulgar

### Introduction

Pemphigus vulgaris is the most common and life-threatening variety of pemphigus, which is a group of intraepidermal bullous disease, involving

cutaneous and or mucosal manifestations. Early diagnosis and prompt adequate treatment reduce the high morbidity and mortality rates. Systemic corticosteroid associated with immunosuppressive drug is a

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main therapeutic tool.<sup>1-6</sup> Administration of azathioprine and sulfone reduces the steroid dose in the control pemphigus. However, these drugs may have some role in the development of nail disorders in this setting.<sup>7</sup> Nail involvement can precede a flare up of pemphigus or appear later in the course or during convalescence, and the changes may be related to the disease or drug adverse effect.<sup>3,4</sup> We describe nail disorders appearing later in a patient treated with azathioprine and sulfone.

### Case report

FPS, a 32-year-old man, was admitted on the Hospital of West in the city of Barreiras-Bahia, Brazil, during two months of the second semester in 2013. On admission, he presented with generalized bullous and erosive lesions affecting the skin as well as the mucosal surfaces of the eyes, nose and mouth. Worthy of note, his nails had normal features (fig. 1 A and B). Moreover, both the typical signs of Nikolsky and of Asboe-Hansen were found positive. There was neither personal nor familial antecedent of similar mucocutaneous disturbances. With base on clinical features and skin biopsy studies, the diagnosis was further established. Pemphigus vulgaris was confirmed with base on the finding of intraepidermal blisters and suprabasal acantholytic changes, and immunoglobulins of the IgG class in

keratinocytes. Absence of microorganisms in cultures of blood and skin ulcerations, and negative serological tests contributed to the differential diagnosis with infectious and connective tissue diseases. Additionally to accurate nutritional balance and specialized topical care of cutaneous lesions, he was treated with prednisone 60mg/day, azathioprine 150mg/day, and dapsone 100mg/day. After two months of clinical management in hospital environment, the main changes improved well, and he was referred to the outpatient surveillance with no symptoms, and only taking azathioprine. Despite of the good outcome of his severe disease, major changes were observed in his finger and toe nails during convalescence (fig. 1 C and D).

### Discussion

Pemphigus vulgaris is the most frequent and life-threatening variety of pemphigus, a group of intraepidermal bullous diseases with cutaneous and or mucosal manifestations.<sup>1-6</sup> The bullae are typically developed in a suprabasal location due to akantolysis, phenomenon related to autoantibodies against a desmosomal protein complex of skin and mucosae.<sup>1-6</sup> Clinical hypothesis must be confirmed by histopathology, including immunofluorescence.<sup>1-6</sup> Early diagnosis and prompt adequate treatment can reduce the morbidity and mortality.<sup>1</sup> Nail involvement is

scarcely reported in this disease,<sup>3-6</sup> and has been associated with severity.<sup>6</sup> Paronychia, onychomadesis, Beau's lines and nail hemorrhages are more often reported; but cross ridging, discoloration, hyperkeratosis, paronychia, pitting, pterygium, onychodystrophy, onycholysis, onychorrhexis, onychoschizia, and trachyonychia have been described.<sup>1-6</sup>

Proximal subungual hematomas, Beau's lines, onychodystrophy, onychomadesis, transverse leukonychia, and development of pincer nails occurred in the patient herein described. Bacterial, fungal and viral agents can cause local infections and give rise to misdiagnosis. Nail involvement either precede a flare up or can appear later,<sup>3,4</sup> as occurred in this patient.



**Figure 1. A and B:** Normal aspect of hand and toe nails before starting drug-treatment of pemphigus vulgaris; some bullous changes and extensive erosions are showed on abdomen; **C:** All the hand nails show variable detachment of the nail plates (onychomadesis) and transversely-directed furrows (Beau's lines), in addition to some transverse leukonychia, and proximal dark-brown discolorations (subungual hematomas) in two of the fingers; and **D:** The toe nails show mild degree of onychodystrophy, with proximal black to bluish discoloration (subungual hematomas) in five of the ten fingers, and bilateral thickening and transverse overcurvature of the nail plates of the great toes (mimicking pincer nails at an initial stage).

Mechanisms of the changes involve bullous lesions in the nail bed, nail matrix or nail fold.<sup>3</sup> The conspicuous nail changes developed in the young patient herein reported during the convalescent period of disease, after a long-term administration of azathioprine and sulfone. Although may pose eventual concern in relation to the origin of the nail abnormalities,<sup>7</sup> both drugs may help to reduce the steroid dose in the management of pemphigus vulgaris.<sup>1</sup> Benhiba et al. reported typical suprabasal acantholysis in nail biopsies performed in a 45-year-old man with diagnosis of pemphigus vulgaris confirmed by direct immunofluorescence.<sup>2</sup> Cahali et al. described five patients with this disease and nail involvement; 80% were females, the mean age was 38 years and the changes antedated (33%) or were part (47%) of flare up. Interestingly, nail changes were the unique manifestation of disease in 20% of the cases.<sup>3</sup> Carducci et al. reported nail involvement, which appeared after skin and mucosal changes, in three patients with pemphigus vulgaris; two were women, and the mean age was 40 years.<sup>4</sup> Despite of the relative rarity, Cahali et al. and Patsatsi et al., reported individuals presenting with 19 and 20 nails involved, similarly to the findings of the young male herein described.<sup>3,5</sup> Nail changes in our patient are Beau's lines, onychomadesis, leukonychia, subungual hematomas and pincer nail deformities. As

the finger nails are normal appearing at the beginning, the respective changes seem to have developed due to pemphigus itself and/or the medications used to treat this disease. Although the photography of the left first toe nail is showing normal features at the time of diagnosis, it is rather difficult to confirm that all the toe nail changes occurred later in the course of disease due to the involvement of pemphigus. We believe that case studies enhance the suspicion index about scarcely reported conditions.

### Conclusion

In the young patient herein described conspicuous nail changes developed during the convalescent period of disease, after a long-term administration of azathioprine and sulfone. Although may pose some concern about the aesthetic role, the nail changes can be reversible and these drugs are safe and useful in the management of patients with pemphigus vulgaris.

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