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Case Report

Nail Involvement in Pemphigus: Brief Report

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Abstract

Background: The objective of our study is to describe the different nail involvement at the beginning of the disease according to the type of pemphigus and according to the severity of the pemphigus.

Material and methods: We carried out a prospective study over a 3-year period from 2019 to 2022 that included 21 hospitalized patients out of 80 with polydactyly nail involvement 13 pemphigus vulgaris, 4 pemphigus vegetans, and 3 superficial pemphigus in our dermatology department Centre Hospitalier Universitaire (CHU) Hassan II Fez.

Results: Paronychia was the most frequent finding overall, observed in 11 (52.38%) cases, followed by onychomadesis and Beau's lines in 8 cases (38%) and subungual hemorrhage in 6 cases (28.57%). Nail involvement was correlated with the severity of the pemphigus, in particular with severe mucosal involvement (17 patients or 80.9%).

Conclusion: Nail involvement is a sign of pemphigus severity and may herald an exacerbation of the underlying disease.

Keywords: Immunofluorescence; Autoimmune Bullous Dermatosis; Nails

Introduction

Pemphigus is a chronic autoimmune bullous dermatosis whose nail involvement is often underestimated. It seems to be an important clinical sign reflecting the severity and evolution of pemphigus considering the presence of target antigens in the proximal nail fold, nail matrix, and hyponychium. The objective of our study is to describe the different nail involvement at the beginning of the disease according to the type of pemphigus and according to the severity of the pemphigus.

Material and Method

We carried out a prospective study over a 3-year period from 2019 to 2022 including all patients hospitalized for pemphigus in our dermatology department CHU Hassan II Fez. The diagnosis of pemphigus was based on clinical and immunopathological data. Clinical data were collected through an information sheet before the initiation of treatment. The diagnosis of pemphigus was retained by the confrontation of the clinical data: flaccid and fragile bubbles leading to dragging and painful erosions with bright red surface usually superinfected bordered by an epithelial collar, histological: intraepidermal bullae with suprabasal acantholysis and immunohistochemistry: Direct Immunofluorescence (DIF) showing deposits of IGG on a mosaic substrate. Indirect immunofluorescence has been performed in all patients showing the presence of anti-intercellular substance antibodies, which is a marker of disease progression. The following variables were recorded: age, gender, type of pemphigus, duration of evolution, a Japanese pemphigus disease severity score, and clinical aspects. The clinical signs analyzed were: paronychia, onychomadesis, Beau's lines, subungual hemorrhage, periungual bullae, trachyonychia, anonychia, and onycholysis. Consent and authorization from the local ethics committee were required. Data were entered into Excel and analyzed using Epi info version 7 software.

Case Report

Over a 3-year period, 80 cases of pemphigus were collected. Of these, 21 patients had polydactyly nail involvement (26.25%), with 13 patients suffering from pemphigus vulgaris, 4 from pemphigus vegetans, and 3 from superficial pemphigus. The duration of the disease ranged from 1.5 months to 5 years. Of the 39 patients with Pemphigus Vulgaris (PV), 13 (33.33%) had nail changes. Paronychia and onychomadesis were present in 7 patients each (53.8%), Beau's lines were present in 6 patients (46.15%), subungual hemorrhage was present in 5 patients (38.4%), trachyonychia in 3 patients (23.07%), and periungual bullae in 1 patient (7.69%). Distal onycholysis, periungual bullae, and melanonychia bands were observed in 1 patient each. Of the 8 patients with pemphigus vegetans, 4 patients had nail involvement. Paronychia was present in all 4 patients, Beau's line, onychomadesis, and subungual hemorrhage in 1 patient each. On the other hand, of the 33 cases of superficial pemphigus (SP), only 3 had nail involvement, i.e., 9% of the cases, of the type anonychia, distal onycholysis, and Beau's line. Paronychia was the most frequent finding overall, observed in 11 (52.38%) cases, followed by onychomadesis and Beau's lines in 8 cases (38%) and subungual hemorrhage in 6 cases (28.57%). Other nail manifestations such as distal onycholysis and anonychia were observed in one patient each. Nail involvement was correlated with the severity of the pemphigus, in particular with severe mucosal involvement (17 patients or 80.9%). The types of nail involvement found in severe pemphigus are dominated by paronychia and onychomadesis (Fig. 1).

Figure 1: Images showing onychomadesis, Beau's lines, paronychia, and periungual bullae in pemphigus patients.

Discussion

Nail changes in autoimmune blistering disorders, pemphigus, in particular, are usually underestimated, and very few studies have looked at them. Sinclair, et al., showed that all antigens of normal skin are also normally expressed in the proximal nail fold, nail matrix, and hyponychium including desmoglein 1 and 3 explaining nail involvement occurring in pemphigus [1]. According to the literature, the prevalence of nail changes in pemphigus vulgaris is ranged from 31.6% to 47%, which is similar to our results of 33.33% [2].

Even if nail involvement has often been reported with pemphigus vulgaris, our study showed that it could occur in other types such as pemphigus vegetans and superficial pemphigus. In our study, the fingernails were the most affected (66.67%), and paronychia and onychomadesis were the most common nail changes (46.15%) followed by Beau's line (38%). The most common nail aspects in PV were paronychia and onychomadesis 53.8% each. We found only 3 patients with nail changes in superficial pemphigus, anonychia, distal onycholysis, and Beau's line in each patient. However, in pemphigus vegetans, paronychia was the most common and was found in 50% of patients. The fingernails in PV are usually more affected than toenails [3].

Paronychia and onychomadesis are the most common manifestations reported in pemphigus vulgaris [4]. Paronychia occurs as a result of acantholysis of the lateral nail fold epidermis [5], and onychomadesis is a result of a severe underlying inflammatory process that causes complete arrest of nail matrix activity. The distance from the proximal nail fold in Beau's line reflects the time from disease onset and multiple lines correspond to recurrent exacerbations [6]. They are no studies in the literature regarding nail involvement in superficial pemphigus.

However, although poorly reported, pemphigus vegetans were responsible for the complete destruction of the nail unit with an unaesthetic aspect [7]. In our study, nail involvement was correlated with the severity of pemphigus, in particular with severe mucosal involvement (80.9%). All patients had a severe Japanese pemphigus disease severity score with moderate to severe mucosal involvement. The most frequent nail changes in severe pemphigus are paronychia and onychomadesis. As other studies have shown, nail involvement is taken as an indicator of the severity of the pemphigus [8,9]. Laffitte, et al., suggested that expression of Dsg1 and Dsg3 in the periungual skin is disease-specific, and autoantibodies against Dsg3 alone may have a role in the development of paronychia in PV which explains why paronychia is the most frequent nail involvement in pemphigus vulgaris [10-12].

Conclusion

Nail involvement is a sign of pemphigus severity and may herald an exacerbation of the underlying disease. This involvement can be explained by the high titers of anti-DSG1/anti-DSG3 or anti-DSG3 autoantibodies in severe cases and their specific presence in periungual skin and also in the matrix and nail bed.

Conflict of Interest

The authors have no conflict of interest to declare.

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