

Nail Involvement in Pemphigus Vulgaris: Case Report

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Abbreviations: PV: Pemphigus Vulgaris

Introduction

Pemphigus = Term derived from the Greek word "pemphix" = "bubble" first described by Willan in 1808 (Pemphigus vulgaris). Chronic bullous dermatosis affecting skin + mucous membranes histologically characterized by intraepidermal cleavage secondary to loss of intra-keratinocyte adhesion. It is a specific autoimmune disease characterized immunologically by production of IgG-type auto-pathogens directed against desmosomal proteins or desmosomal antigens 1&3. Few studies in the literature discuss nail involvement during

pemphigus vulgaris, although it is chronic? Disease progression depends on treatment [1,2]. We describe a patient with widespread, severe pemphigus vulgar who had nail involvement of all 4 extremities, which occurred at the beginning of the disease.

Case Report

A 65-year-old woman presented with 5 months history of lesions with fluid content with extremely painful oral lesions. Histology highlights an intraepidermal blister formed by the process of acantholysis with direct immunofluorescence presence of IgG deposits on the surface of keratinocytes. The diagnostic of pemphigus vulgar was made. The characteristic of this patient was the presence of onychomadesis and proximal haemorrhage affecting 13 nails (Figure 1).



Figure 1: Onychomadesis and proximal haemorrhage affecting 13 nails.

The patient received oral corticosteroid therapy at a dose of 1 mg / kg / day and a pretreatment assessment including pre azathioprine and Rituximab at a dose of 1g/15days was performed. Then the patient received her

first injection of rituximab with good evolution and early healing of her lesions with a Nikolsky that is negative (Figure 2).



Figure 2: Disappearance of lesions after treatment with rituximab.

Discussion

We present this case primarily to describe an example of the relatively rare occurrence of nail involvement in pemphigus vulgaris. Nail attack during pemphigus is rare. It may be the consequence of the proximity of the cutaneous lesions which is not the case of our patient or related to peri and/or subungual bubbles, or maybe an autoimmune mechanism. These alterations cause a fragility of the matrix, responsible for the formation of a dystrophic nail and its detachment from the bed. There seen in case of severe pemphigus vulgaris[3].

Nail changes most often found are onychomadesis, onycholysis, cross-ridging, pitting, Beau's lines, subungual hemorrhage, paronychia, nail dystrophy, and nail plate discoloration [4,5]. Stone & Mullins, had studied 21 female patients who had cutaneous lesions and mucous membrane associated paronychia affecting several finger nails, toenails were not affected [6]. Parameswara & Naik who studies 28 patients had presented also cutaneous and mucous lesions, and had onychomadesis of fingers and toes (7). De Berker et al, published a study of 71 patients, having cutaneous and mucous lesions and presenting onycholysis and longitudinal ridging [8].

Onychomadesis like the case of our patient, can occur in pemphigus. It may be because of two possible causes. First, impairment of the nail matrix may prevent new nail formation and thus loosen the existing nail plate. Second, a subungual blistering process may detach the nail plate from the underlying nail bed [9]. The known association of Pemphigus Vulgaris with nail disease, led to the

working assumption that the nail pathology was related to the underlying disease. Complete and permanent nail recovery after successful treatment of the PV supports this assumption. Nail involvement in patients with pemphigus usually occurs when active pemphigus lesions are present in other locations and thus is not normally difficult to diagnose. However, chronic paronychia and onychomadesis have been reported as the initial manifestation of Pemphigus Vulgaris [10].

Conclusion

The nail attack during pemphigus is said to be rare. It is related to peri and / or subungual bubbles or may be the consequence of the proximity of the cutaneous lesions. These alterations cause a fragility of the matrix, responsible for the formation of a dystrophic nail and its detachment from the bed [11]. All constituents of the nail can be affected. The aspects encountered are very polymorphous. Nail biopsy is not necessary if the diagnosis is already demonstrated during the skin biopsy. They must be searched systematically. They can be a criterion of severity or recurrence of a pemphigus under treatment.

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