DOI: 10.23937/2474-3682/1510101

Volume 4 | Issue 4 Open Access



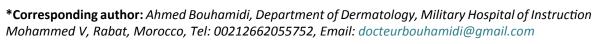
Clinical Medical Image Library

IMAGE ARTICLE

The Neumann Type of Pemphigus Vegetans

Ahmed Bouhamidi^{1*}, Youssef Zemmez¹, Hafsa Chahdi², Mohamed Oukabli² and Naoufal Hjira¹

¹Department of Dermatology, Military Hospital of Instruction Mohammed V, Rabat, Morocco ²Department of Pathology, Military Hospital of Instruction Mohammed V, Rabat, Morocco





Abstract

Pemphigus vegetans is a variant of pemphigus vulgaris and is the rarest form of pemphigus. It is classified based on the clinical picture and evolution as the Neumman type or Hallopeau type. We report a 50-year-old woman with a Neumann type of pemphigus vegetans who was successfully treated with corticosteroids.

Keywords

Neumann subtype, Pemphigus vegetans, Corticosteroids



Figure 1: Erosive vesicles on the lip (A) and hypertrophic verrucous vegetative plaques on the inguinal folds (B).

Introduction

Pemphigus vegetans is a rare clinical form of pemphigus, two clinical subtypes of pemphigus vegetans exist, which are initially characterized by bullae and erosions (the Neumann subtype) or pustules (the Hal-

lopeau subtype). We report a 50-year-old woman with a Neumann type of pemphigus vegetans who was successfully treated with corticosteroids.

Clinical Case

A 50-year-old woman presented with a 2-months



Citation: Bouhamidi A, Zemmez Y, Chahdi H, Oukabli M, Hjira N (2018) The Neumann Type of Pemphigus Vegetans. Clin Med Img Lib 4:101. doi.org/10.23937/2474-3682/1510101

Accepted: August 07, 2018; Published: August 09, 2018

Copyright: © 2018 Bouhamidi A, et al. This is an open-access content distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

DOI: 10.23937/2474-3682/1510101 ISSN: 2474-3682

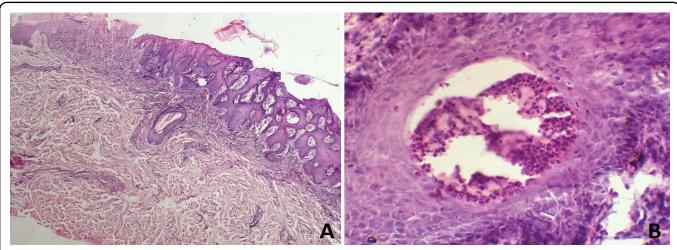


Figure 2: A biopsy of the vegetating plaque from the inguinal folds showed a hyperplastic epidermis, eosinophilic spongiosis, and intraepithelial abscesses packed with eosinophils (A: H&E, ×10 - B: H&E, ×40).



Figure 3: Evolution of lesions after 2 weeks of treatment.

history of vegetating plaques on the inframammary, axillary and inguinal folds with erosions of the oral mucosa, and perioral area. There was no history of other skin disease. She had no family history of a blistering disorder. On physical examination, there were oozing, erosive vesicles on the lip and hypertrophic verrucous vegetative plaques on the inframammary, axillary and inguinal folds (Figure 1). Biopsy specimens were obtained from the verrucous plaque on the inguinal folds. The histologic findings showed massive papillomatosis and spongiosis, as well as eosinophilic granulocytes throughout the entire thickness of the epithelium. In addition to eosinophilic spongiosis, we also found suprabasal acantholy-

sis (Figure 2). Although direct immunofluorescence was negative, Indirect immunofluorescence reveals circulating anti-epithelial cell-surface IgG which confirmed the diagnosis of pemphigus vegetans. The patient was treated with oral Prednisone 60 mg daily, The erosive vesicular and verrucous lesions were healed 2 weeks. She has not had any recurrence of the lesions since (Figure 3).

Comments

Pemphigus vegetans is a rare variant of pemphigus that is characterized by vegetative lesions mainly in flexions, although lesions can occur at any site, this disorder affects middle-aged adults [1]. Two types of Pemphigus

vegetans are recognized (Neumann and Hallopeau subtype), which are differentiated according to their clinical presentation, their evolution and their response to treatment [2]. Clinical findings, histopathological, direct and indirect immunofluorescence studies allow the clinician to make the correct diagnosis [3]. Treatment of pemphigus vegetans is similar to that for pemphigus vulgaris, and this is normally accomplished with systemic steroids [4]. The addition of immunosuppressants, such as azathioprine, mycophenolate mofetil and cyclosporine, can improve remission rates and allow steroid sparing effect [5].

Conclusion

In these uncommon diseases, careful analysis of the clinical and histological results allowed us to establish a diagnosis and treat the patient successfully.

Conflicts of Interest

The authors do not declare any conflict of interest for this work.

Author's Collaboration

All authors were actively involved in the development of this work.

References

- Cozzani E, Christana K, Mastrogiacomo A, Rampini P, Drosera M, et al. (2007) Pemphigus vegetans Neumann type with anti-desmoglein and anti-periplakin autoantibodies. Eur J Dermatol 17: 530-533.
- 2. Almeida HL Jr, Neugebauer MG, Guarenti IM, Aoki V (2006) Pemphigus vegetans associated with verrucous lesions: expanding a phenotype. Clinics (Sao Paulo) 61: 279-282.
- 3. Monshi B, Marker M, Feichtinger H, Schmid G, Kriehuber E, et al. (2010) Pemphigus vegetans-immunopathological findings in a rare variant of pemphigus vulgaris. J Dtsch Dermatol Ges 8: 179-183.
- 4. Ichimiya M, Yamamoto K, Muto M (1998) Successful treatment of pemphigus vegetans by addition of etretinate to systemic steroids. Clin Exp Dermatol 23: 178-180.
- Werth VP, Fivenson D, Pandya AG, Chen D, Rico MJ, et al. (2008) Multicenter randomized, double-blind,placebo-controlled, clinical trial of dapsone as a glucocorticoid-sparing agent in maintenance-phase pemphigus vulgaris. Arch Dermatol 144: 25-32.

