The Neumann Type of Pemphigus Vegetans

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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 Hallopeau 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
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