

A case of pemphigus erythematosus with classical facial butterfly configuration and trunk involvement

Um caso de pêmfigo eritematoso com afeção facial clássica em borboleta e envolvimento do tronco

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A 50-year-old male presented with erythematous and scaly plaques on his face, scalp, chest, and dorsum with 1-year duration. Facial lesions affected particularly the malar region, bilaterally, strikingly sparing the nasolabial fold (Fig. 1). Histopathology of lesional skin (Fig. 2) revealed a subcorneal blister with discrete acantholysis in the granular layer. Direct immunofluorescence (Fig. 3) revealed intercellular deposits of Immunoglobulin G (IgG) and granular deposition of IgG in the basement membrane. These findings supported the diagnosis of pemphigus erythematosus (PE). Blood analysis revealed positive antibodies against desmoglein-1 (407 RU/mL, reference value < 20) and negative ANA and desmoglein-3 antibodies. Blood cell count and renal function were unremarkable. The patient was treated initially with oral prednisolone and rituximab with little control and is now under azathioprine and hydroxychloroquine with an excellent response.

PE, also known as Senear-Usher syndrome, is a localized variant of pemphigus foliaceus with clinical overlap with lupus erythematosus¹, hence the name of the disease. This is a rare autoimmune blistering



Figure 1. Erythematous and scaly dermatosis affecting the face, scalp and upper trunk.

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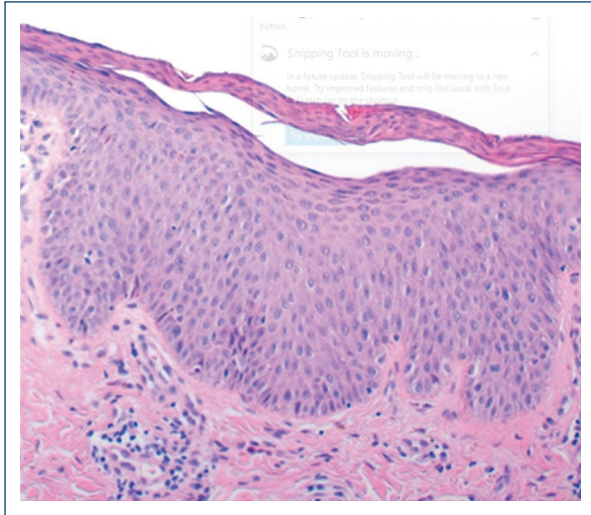


Figure 2. Routine histology of lesional skin, revealing subcorneal blister with discrete acantholysis in the granular layer. HE, x 200.

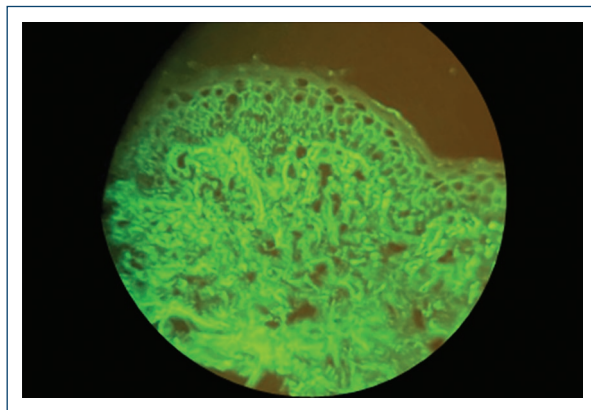


Figure 3. Direct immunofluorescence revealed intercellular deposits of IgG and granular deposition of IgG in the basement layer.

disorder, with little over 200 cases described in literature². Clinically, erythematous scaly patches are found in typical locations of pemphigus foliaceus, with addition of the malar region in a butterfly distribution, simulating

cutaneous lupus erythematosus. However, it is rare for patients to have systemic lupus erythematosus³. Histologically, PE is similar to pemphigus foliaceus, with superficial blisters below the stratum corneum or within the granular layer³. Direct immunofluorescence, however, differs from pemphigus foliaceus due to the granular deposition of IgG along the basement membrane, as in cutaneous lupus erythematosus.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

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