Pemphigus foliaceus with neutrophilic spongiosis evolving to an atypical pemphigus phenotype

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A 46-year-old Brazilian man, with initial pustular lesions, neutrophilic spongiosis and subcorneal cleavage evolved to an atypical pemphigus phenotype, with suprabasal acantholysis. Interestingly, his autoantibody profile, tested by immunofluorescence, immunoblotting, enzyme-linked immunosorbent assay, and immunoprecipitation revealed exclusive IgG anti-desmoglein 1 antibodies in all phases of the disease.

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A 46-year-old Brazilian man had a pustular form of pemphigus foliaceus (Fig 1, A). Initial investigation revealed a neutrophilic spongiosis, subcorneal cleavage (Fig 1, B), and intraepidermal, intercellular IgG deposits by direct and indirect immunofluorescence (Fig 1, C). After 3 months of oral prednisone, clinical remission was obtained, but new pustules appeared on soles and ankles (Fig 1, E). Surprisingly, laboratory evaluation revealed suprabasal acantholytic cleavage and the same immunofluorescence pattern (Fig 1, F and G).
Enzyme-linked immunosorbent assay utilizing recombinant desmogleins 1 (rDsg1) and 3 (rDsg3) generated in a baculovirus system revealed reactivity with rDsg1 and absence of response against rDsg3 in all phases of the disease. Cold immunoprecipitation coupled to immunoblotting utilizing rDsg1 and rDsg3 and 2 chimeric ectodomain segments of Dsg1 (EC1-4 and EC-5) showed (1) clear reactivity against EC1-4 and EC-5 during the initial pustular phase (Fig 1, D); (2) a maintenance of EC-5 and reduction of EC1-4 response during clinical remission; and (3) reactivity against EC1-4 and EC-5 segments during clinical relapse (Fig 1, H). These findings corroborate the hypothesis of intramolecular epitope spreading of desmoglein 1 in pemphigus foliaceus. In our case, it is possible that epitope spreading may be leading to a pemphigus phenotype shift once a suprabasilar cleavage was not observed with characteristic clinical or immunohistochemical findings of pemphigus vulgaris. A long-term follow-up is required to evaluate future mucosal involvement, anti-Dsg3 antibodies, or antibodies directed against a possible novel autoantigen.

REFERENCES