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. (J Am Acad Dermatol 2004;51:1012-3.)

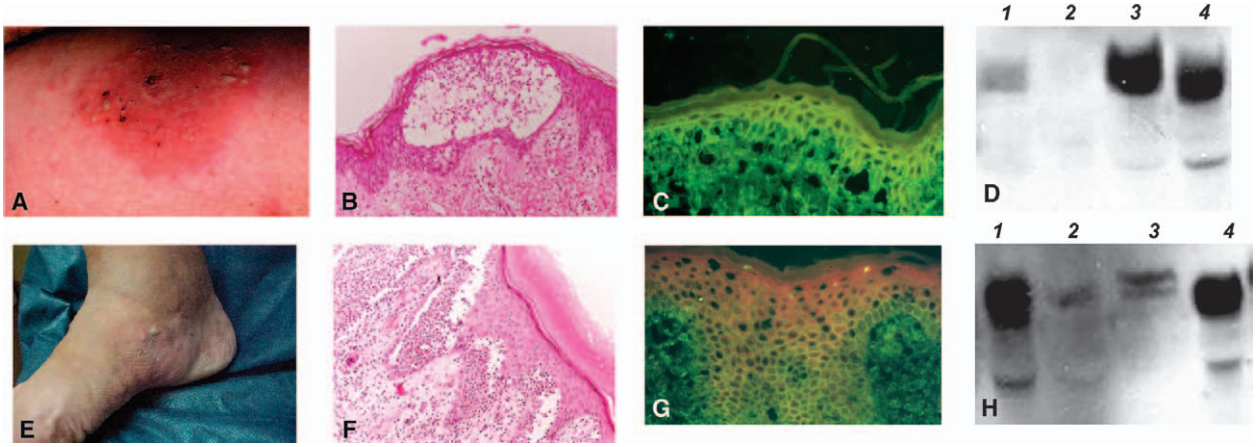

Fig 1. **A**, Initial pemphigus foliaceus. **B**, Subcorneal cleavage with neutrophilic spongiosis. **C**, IgG intercellular deposits. **D**, Immunoprecipitation, 10% SDS-PAGE. Lane 1, rDsg1; lane 2, rDsg3; lane 3, EC1-4, rDsg1; lane 4, EC-5, rDsg1. **E**, Clinical relapse. **F**, Suprabasilar cleavage filled with eosinophils and neutrophils. **G**, IgG intercellular deposits. **H**, Immunoprecipitation, 10% SDS-PAGE. Lane 1, rDsg1; lane 2, rDsg3; lane 3, EC1-4, rDsg1; lane 4, EC-5, rDsg1. (**B** and **F**, Hematoxylin-eosin stain; **C** and **G**, direct immunofluoresence; original magnifications: **B**, ×200; **F**, ×400.)

A 46-year-old Brazilian man had a pustular form of pemphigus foliaceus (Fig 1, *A*). Initial investigation revealed a neutrophilic spongiosis,[1] 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 suprabasilar acantholytic cleavage and the same immunofluorescence pattern (Fig 1, *F* and *G*).

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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 maintainance 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*).[2] These findings corroborate the hypothesis of intramolecular epitope spreading of desmoglein 1 in pemphigus foliaceus.[2] 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 immunochemical 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.

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