Pemphigus – an Atavism?

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Regarding pemphigus, there have been remarkable discoveries during the last 50 years: dermatopathology; immunofluorescence; antigen isolation; genetic background. Originally described by Auspitz in the 1880s, acantholysis and suprabasilar cleft formation were recognized in their diagnostic importance only more than half a century later, by the Paris School [1] and by Lever [2, 3]. Immunofluorescence phenomena were first described in the mid-sixties [4, 5]; pemphigus antigens, first isolated in the early seventies, could be characterized more precisely within the last decade [6, 7]. The discovery of a certain genotype, conferring the propensity for the disease onto carrier individuals, could be unraveled recently [8].

Per se, pemphigus is an 'old' disease. The clinical entity was developed during the last century, when Cazenave in 1844, Hebra in 1860 and Isidor Neumann in 1886 coined the now familiar terms of p.foliaceus, p.vulgaris, p.vegetans (later to be supplemented, in this century, by Senear and Usher, p. erythematosus, and Jablonska et al, p.herpetiformis) [9].

As a term, pemphygos, later pemphigus, i.e. the latinized version of an originally Greek designation, was introduced by Boissier de Sauvages in the 1750s [9]. Case reports under the designation febres pemphigodes, however, are much older. Semantically, febris pemphigodes, or ἡνπετολ ἡμφοιγώδεε; > date back to the corpus hippocraticum and the dawn of scientific medicine.

Now, more exciting news may just be around the corner. Grando [10], in this issue of the journal relates the shedding of snake skin (Matrix natrix) to a pemphigus-like process. Contrary to earlier studies [11], the author traces intercellular antigens in shedding snake epidermis, by means of human pemphigus autoantibodies. The key question remains unanswered, yet: the author does not report on the existence of snake autoantibodies in the animals' serum and their fixation to the respective epidermal antigens, during shedding, despite our urging to check this important point. We felt, nevertheless, that the paper should not be held back. From the conceptual point of view, these hints as to the possible existence of a natural model, in animals, for what pemphigus as a disease is in man, are very stimulating and other investigators may be attracted.

Is pemphigus an atavism then?

Definitions of atavism vary. The detection of a certain genotype, which confers a high risk to carriers, to develop the disease [8], may permit to suggest a combination type of atavism.

Another step forward, actually backward, in the elucidation of the pathogenesis of pemphigus may have been achieved, in case these results prove to be reproducible elsewhere.

To relate natural shedding of epidermis, in animals, to 'shedding' of epidermal layers, in pemphigus patients, by mediation of autoantibodies to intercellular epidermal antigens, would fit almost too neatly into our perception of the significance of the underlying autoimmune mechanism in man.

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