Pemphigus and Intraepidermal IgA

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Sir,

Over the last decade, chronic vesiculo-pustular eruptions with intraepidermal deposits of IgA have been repeatedly reported and tentatively organized in two entities, IgA pemphigus foliaceus and IgA pemphigus of the intraepidermal neutrophilic type [1]. Lately, a third group has been added, in which IgA deposits have a linear subcorneal configuration [2]. Recently, Wallach [3] observed that for such cases the term 'IgA pemphigus' may be misleading and proposed the unified concept of 'intraepidermal IgA pustulosis' with definite clinical, histological, immunological and therapeutic criteria.

To add to this conundrum, it should be noted that intraepidermal deposits of IgA had been observed also as an additional finding in classical forms of pemphigus [4]. Such patients, like the one described herein, may be of some interest.

A 65-year-old otherwise healthy man presented with a 3-month history of mildly itching lesions on his back. No history of photosensitivity, allergy or drug intake was obtained. He had been treated with topical antibiotics and oral corticosteroids with transient benefit. Lesions had recurred and gradually extended to the chest, arms and neck.

Physical examination revealed erythematous scaly crusted lesions and superficial erosions on his upper chest and back and bilaterally in the retroauricular folds. Small grouped hyperkeratotic lesions were also present on his anterolateral cervical regions and on his arms. Pustules, seborrheic scales and bullae were absent. Mucous membranes were spared.

Routine laboratory tests were normal or negative, including fluorescent antinuclear antibodies. A skin biopsy from a lesion on the

back disclosed a subcorneal cleft with acantholytic cells and no neutrophilic infiltrate. Direct immunofluorescence performed on perilesional skin showed intraepidermal suprabasilar deposits of IgA, IgG and C3. Indirect immunofluorescence performed on monkey esophagus showed only IgG directed to the intercellular substance of the epidermis at the final titer of 1/40. Circulating antiinter-cellular substance IgA antibodies were not found.

Diagnosis of pemphigus erythematosus was made and a treatment with 100 mg/day dapsone started. Three weeks later the patient had not improved and was given 37.5 mg prednisone daily that cleared his lesions within 15 days.

Apparently, the cutaneous disorders characterized by intraepidermal deposits of IgA do not form a homogeneous group. Clinically, the published cases have been described as pemphigus foliaceus [5], subcorneal pustular dermatosis [6], dermatosis herpetiformis [7] and even erythema multiforme [8]. Two main types may be distinguished. In true IgA pemphigus, a mild

acantholysis is associated with deposits of IgA in the upper stratum spinosum while in other cases, in which vesiculopustules are the main clinical features, acantholysis is minor and the intercellular IgA are located only in the subcorneal region. Indirect immunofluorescence is usually negative.

Our patient did not fit any of such types of the disorder. Clinically, he had a typical pemphigus erythematosus, with no trace of pustules, and histologically, an intraepidermal acantholytic cleft with no neutrophils in the infiltrate. With direct immunofluorescence, he disclosed IgG and C3 as well as IgA in a suprabasilar location and had circulating low

titers of IgG and no IgA. Finally, his response to dapsone was poor. In brief, he was a classical case of pemphigus erythematosus in which the IgA deposit was only an additional direct immunofluorescence feature.

Our patient resembles the second of the cases described by Nishikawa et al. [7], which had also in vivo intraepidermal IgG deposits and annular herpetiform features. Such patients may be numerous and may go unmentioned. In fact, as early as in 1968, IgA antibodies directed to the intercellular substance have been noted in the sera [9, 10], but also in the blister fluid [10], of many pemphigus patients. In 1971, Jordon et al. [4] found intraepidermal IgA associated with IgG in 1 of 4 cases of pemphigus and gave them no practical importance. Judd and Lever [5], in 1979, found that 46% of IgG-positive pemphigus cases had also deposits of IgA.

In fact, our experience is different, as we found intraepidermal deposits of IgA only in 1 of 43 otherwise ordinary cases of pemphigus. Investigating the real prevalence and significance of intraepidermal IgA in IgG pemphigus would clarify whether patients like ours represent a further subgroup of IgA pemphigus or just a common meaningless coincidence.

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