Intraepidermal IgA Pustulosis Associated with Monoclonal IgA Gammopathy in an HIV-Infected Patient

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Key Words

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Intraepidermal IgA pustulosis (IEAP) is a term coined by Wallach [1] characterized by a pustular eruption similar to the subcorneal pustulosis of Sneddon and Wilkinson, but it differs from this entity in that the direct immunofluorescence test shows deposits of IgA in the epidermis, at the intercellular level in most of the cases. Clinicopathologically there seem to be two distinct types: (1) resembling subcorneal pustular dermatosis (SPD type) and (2) intraepidermal neutrophilic IgA dermatosis (IEN type) [2]. As this entity has recently been associated with HIV infection [2], we would like to report another case of an HIV-infected patient who developed an IEAP and who presented an associated IgA λ paraprotein.

In November 1994, a 35-year-old-male came to our Department because of a generalized vesiculopustular eruption of 1 month of duration. The patient had a history of HIV antibody positivity diagnosed in April 1993 in a routine blood test and had been following therapy with zidovudine since July 1993.

Physical examination revealed the presence of papules, vesicles and pustules arranged in a circinate pattern that were spreading centrifugally. They involved the trunk and, upper limbs, with a predominant involvement of the axillae and groins (fig. 1).

A bacteriological culture revealed the presence of *Staphylococcus aureus* and *Streptococcus* group A, both sensitive to ciprofloxacin. Systemic therapy with ciprofloxacin (500 mg b.i.d.) was initiated and topical antibiotics (neomycin, bacitracin and polymyxin B) were applied. Although some improvement was observed, control of the disorder 1.5 months later was only partial. Topical betamethasone dipropionate 0.05% associated with topical gentamycin 0.1% was then given, and the disorder completely resolved in 3 weeks.

The histopathological study showed a subcorneal pustule containing some isolated acantholytic cells. A direct immunofluorescence test using perilesional skin revealed intercellular IgA in the

upper layers of the epidermis. An indirect immunofluorescence test did not reveal circulating anti-intercellular-substance of epidermis antibodies.

Routine laboratory investigations showed the following abnormalities: total proteins 83.4 g/l, albumin 36.9 g/l and gammaglobulins 29.15 g/l without any homogeneous component. An immunoelectrophoresis test revealed the presence of a monoclonal gammopathy with IgA λ . We report the case of a patient presenting the clinical, histopathological and direct immunofluorescence features reported in IEAP [1]. The fact that the pustules were not sterile raised the question whether the patient suffered from bullous impetigo or not. We consider it a superinfection of the previous vesiculopustular eruption for several reasons: firstly, the lesions only disappeared when topical cortico-

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Fig. 1. Clinical aspect of the skin lesions.

steroids were applied. Secondly, IgA deposits were observed in the intercellular spaces of the upper epidermis. Thirdly, a IgA λ para-protein was detected in the patient's serum. An IgA paraprotein has been reported in 9 patients with IEAP [1, 3-5]. Of these, 2 patients had myelomas, 1 B-cell lymphoma [1] and 1 chronic lymphocytic leukemia [5]. Although dapsone is the first-choice drug for IEAP, our patient did not require systemic therapy.

To our knowledge, only 1 other HIV-infected patient has been reported as suffering from IEAP [6]. We consider that this association might be explained by the presence of the IgA paraproteinemia. HIV-infected patients frequently develop hypergammaglobulinemia with paraproteinemia [7].

The exact nosologic position of the IEAP is not clear. It has been suggested that the antibodies of IEN-type patients react with the pemphigus vulgaris antigen [8, 9]. On the other hand, Ebihara et al. [2] detected antibodies that bind to a doublet of 105- and 115-kD proteins in the sera of 3 patients of the SPD-like type, which appeared to be identical to bovine desmocollins I and II. A reaction of the sera of patients of the same SPD type with human desmocollin I has recently been reported, too [10].

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Remission of Lichen Amyloidosis after Treatment with Acitretin

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Key Words

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In September 1993, a 72-year-old woman presented at our Department with an eruption of reddish brown, coalescent, hyperkeratotic, extremely pruritic papules of her thighs (fig. 1), the extensor surfaces of both forearms and the lumbosacral area. These lesions had been present for almost 2 years, previous treatment with topical cortico-steroids had proven ineffective. The skin was otherwise unaffected. Physical examination and laboratory tests including blood count, liver and kidney function tests were within the normal range.

Histology revealed a papillomatous epidermis and globular eo-sinophilic deposits of the papillary dermis which showed green birefringence under polarized light in Congo red sections and which were also positive for thioflavin T. By direct immunofluorescence, they appeared as aggregated cytooid bodies reacting with anti-IgM and complement C3 antibodies as well as antibodies specific for keratin (Boehringer), vitronectin (Quidel Q) and serum amyloid P (Atlantic Antibodies).

Acitretin treatment was initiated at a dose of 30 mg daily (0.7 mg/kg body weight). Pruritus greatly improved during the subsequent weeks, the lesions of the forearms became flattened, those of the rest of the body cleared almost completely. Acitretin was therefore tapered after 6 weeks and withdrawn after 3 months. As a side effect, moderate dryness of eyes and lips was noted. Laboratory tests including blood lipids remained normal. During an almost 3-year clinical follow-up, no relapse was observed. A control biopsy, taken 6 months after the end of therapy from the previously affected thigh, revealed normal skin. By direct immunofluorescence, only scarce cytooid bodies were detected in the papillary dermis.

Localized amyloidosis is notoriously resistant to therapy. Macular and papular amyloidosis responds poorly to topical steroid. Derm-abrasion may relieve pruritus and result in flattening of the lesions, but the cosmetic outcome is not always satisfactory [1]. Reported beneficial effects of dimethyl sulfoxide [2, 3] or UVB irradiation [4] appear doubtful. Nodular amyloidosis may require surgical or carbon dioxide laser treatment [5]. In the late 1980s, retinoids were tried in lichen amyloidosis with equivocal results in a total of 9 cases described in

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