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

M.A. Barnadas
C. Nadal
G. Verger
C. Gelpí
J.L. Rodriguez
J.M. de Moragas

Departments of aDermatology, bInfectious Diseases and cImmunology, Hospital de la Sta. Creu i St. Pau, Barcelona, Spain

Key Words
Intraepidermal IgA pustulosis
IgA paraproteinemia
HIV infection

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. Clinico-pathologically 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 \( \lambda \) 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 centripetally. 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-

References

Fig. 1. Clinical aspect of the skin lesions.
Dermatolog 1997;194:309-311

Remission of Lichen Amyloidosus after Treatment with Acitretin

N. Reider, N. Sepp, P. Fritsch
Department of Dermatology, University of Innsbruck, Austria

Key Words
Lichen amyloidosus ■ Retinoids · Apoptosis

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 corticosteroids 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 eosinophilic 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 cystoid bodies reacting with anti-IgM and complement C3 antibodies as well as antibodies specific for keratin (Boehringer), vitronecting (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 cystoid 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 amyloidosus with equivocal results in a total of 9 cases described in