

## Childhood Cicatricial Pemphigoid with Linear IgA Deposits: A Case Report

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Langeland T. Childhood cicatricial pemphigoid with linear IgA deposits: A case report. *Acta Derm Venereol (Stockh)* 1985; 65: 354-355.

A 16-year-old boy with a bullous eruption from the age of thirteen, also presented symptoms of scarring conjunctivitis and involvement of oral mucosal membranes. Linear IgA deposits were demonstrated in the basement membrane zone in skin and conjunctiva. The eruptions improved during therapy with aldesulfonsodium, but complete remission was not obtained. Gluten-free diet did not significantly influence the activity of the disease. The described patient seems to fit in a recently described entity of chronic bullous diseases. *Key words: Conjunctival scarring; Aldesulfonsodium.* (Received December 18, 1984.)

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Recently, four patients were described, presenting a unique combination of immunopathological and clinical findings, dominated by linear IgA deposits in the dermo epidermal junction, scarring conjunctivitis and bullous eruptions of the skin (1).

In this paper we report a case, presenting the immunological and clinical findings characterizing this apparently distinct subclass of chronic bullous diseases.

### CASE REPORT

The patient was a 16-year-old boy, who developed an itching, vesico-bullous eruption at the age of thirteen. Apart from allergic rhinitis, the boy was healthy. In particular, there had been no signs of gastrointestinal malabsorption.

The skin lesions, being predominantly localized to the extensor surfaces of the extremities, axillae, and to a lesser degree to trunk and face, consisted of erythematous papules, plaque and vesicles and small bullae. In conjunctiva and in oral mucosa vesicles and small erosions were seen.

Histological examination of the skin lesions revealed papillary microabscesses typical of dermatitis herpetiformis. Direct immunofluorescence of biopsies from involved skin, peri-lesional skin and conjunctiva all showed a linear deposition of IgA at the dermo-epidermal junction zone. Faint linear depositions of IgG were also seen. Jejunal mucosa biopsies were macroscopically and microscopically normal, whereas scanning electronmicroscopy revealed possibly slight alterations of the villous architecture. There was no haematological evidence indicating gluten-sensitive enteropathy. Total IgE and IgG in serum were slightly elevated. Urine analysis revealed intermittent microscopical haematuria. Other laboratory tests, including those for malabsorption and anti-basement membrane zone antibodies were negative. The HLA-type was A<sub>2</sub>, B<sub>8</sub>, B<sub>40</sub>, CW<sub>3</sub>.

*Clinical course.* The skin lesions responded well to aldesulfonsodium, whereas elimination of gluten from the diet did not significantly reduce the activity of the disease. The involvement of the conjunctivae has not been controlled by the therapy, and have resulted in continuous complaints and increasing scarring of the eyelids (Fig. 1). This has required surgical correction in order to avoid chronic irritation of cornea due to trichiasis.

### DISCUSSION

The case reported here presents clinical and immunological findings similar to those recently described in 4 female patients by Wojnarowska et al. (1). These authors suggest that this combination of immunopathological and clinical findings represents an unrecognized entity of the chronic bullous disorders, and propose the designation "childhood cicatricial pemphigoid with linear IgA-deposits" (1).

We agree that our patient does not fit in with any of the previously described bullous disorders such as chronic bullous disorders of childhood (2), linear IgA disease of the adults (3), dermatitis herpetiformis, or classical cicatricial pemphigoid.



*Fig. 1.* Scarring conjunctivitis with trichiasis.

In contrast to the patients described by Wojnarowska et al., our patient did not spontaneously improve with respect to the skin involvement. Four years after the onset, continuous administration of aldesulfonsodium was still required, in order to control the skin eruptions. We experience the conjunctival involvement as the most severe aspect of this disease, possibly leading to impaired visus in the future. Therefore, a firm cooperation should be established with an ophthalmologist as soon as the diagnosis is verified.

## REFERENCES

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