A peculiar case of linear IgA bullous dermatosis.

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A Peculiar Case of Linear IgA Bullous Dermatosis

To the Editor.—We observed an adult patient with sequential development of mucosal lesions typical of cicatricial pemphigoid (CP) and skin lesions clinically suggestive of early, urticarioid bullous pemphigoid (BP). Direct immunofluorescence showed a linear IgA deposition at the dermoepidermal junction in both lesions. The patient did not respond either to the classic combined treatment with diamino-dimethyl-sulfone (DDS) and steroids or to immunosuppressive agents. Interestingly, on the contrary, the mucosal and cutaneous lesions showed a specifically different response to different treatments, ie, the lesions were satisfactorily cured by low doses of tetracyclines and oral steroids, respectively.

Report of a Case.—A 61-year-old man was first seen in 1986 with erythematous and erosive lesions of the conjunctival, oral (Fig 1), prepucial, and epiglottal mucosa for 8 months. Major symptoms consisted of burning and clear reduction of smell and taste sensitivity. Histologic examination, performed on a prepucial biopsy specimen, showed a subepithelial bulla and a perivascular infiltrate of mononuclear cells and neutrophils in the lamina propria. Direct immunofluorescence showed a linear deposition of IgA and C3 at the dermoepidermal junction. Indirect immunofluorescence on normal human skin was negative. The diagnosis of linear IgA-CP was made, and the patient was treated with prednisone (50 mg/d orally) and diamino-dimethyl-sulfone (DDS) (50 to 75 mg/d orally); the above treatment was discontinued after 10 months because of minimal improvement of the disease. A new treatment with prednisone (50 mg/d orally), and immunosuppressive agents (azatioprine, 0.5 to 1 mg/kg per day orally) did not produce any significant improvement. This treatment was thus discontinued after 2 months. The patient was left untreated. Seven months later (19 months from the first examination), the patient progressively developed small, erythematous and edematous, itching papules on the limbs (Fig 2), clinically suggesting BP in its early, urticarioid phase. Histologic examination of a skin biopsy specimen showed edema and perivascular lymphohistiocytic infiltrate in the upper dermis; the epidermis and dermoepidermal junction were histologically normal. Direct immunofluorescence showed a linear deposition of IgA and C3 at the dermoepidermal junction. The cutaneous lesions were rapidly cured by low doses of oral steroids (triamcinolone, 8 mg/d orally, then gradually reduced to 2 mg/d). Due to

Fig 1.—Erosive lesions of the vestibular labial mucosa.

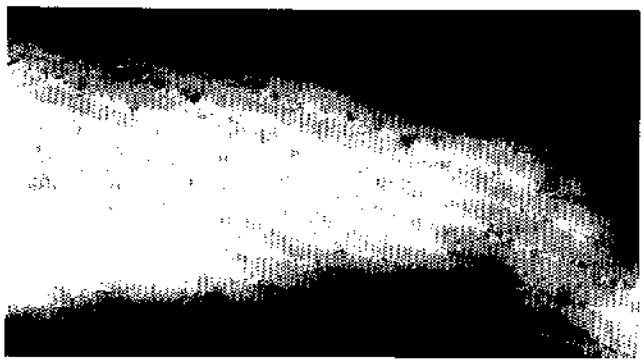

Fig 2.—Erythematous, edematous, urticarioid papules on the right lower limb.

the recent emphasis in the use of tetracyclines in the treatment of bullous pemphigoid,[1] we decided to combine the steroid treatment with tetracycline hydrochloride (250 mg orally two times a day). Surprisingly, the mucosal lesions clearly improved, with progressive reduction of erythema and edema, followed by re-epithelialization, and cicatricial evolution (more evident in the genital area). This objective improvement was accompanied by a gradual and complete normalization of smell and taste sensitivity. At present (43 months follow-up), the skin manifestations are controlled by very low doses (2 mg/d) of triamcinolone. Every effort to discontinue the treatment has invariably lead to a relapse. Mucosal lesions reoccur rapidly when low-dose tetracycline treatment (250 mg/d) is discontinued.

Comment.—Two problems arise based on critical analysis of this case: (1) interpretation and classification, and (2) the peculiarity of the response to treatment. Concerning the first point, we cannot, of course, exclude that CP and urticarioid BP represent two distinct diseases in close temporal sequence. Despite the different clinical and histologic features, however, both mucosal and skin lesions showed a linear IgA deposition at the dermoepidermal junction, suggesting the overlap of different clinical conditions with common immunopathologic features. The association of CP and BP lesions in the same patient may be explained with the assumption that CP and BP represent a continuum of the same disease process, with variable clinical manifestations that often overlap.[2,3] The immunopathologic finding of isolated IgA linear deposition at the dermoepidermal junction in both mucosal and cutaneous lesions reinforces such an interpretation, and justifies the classification of the reported case as linear IgA bullous dermatosis.[4] With this in mind, it is mandatory to emphasize the peculiarity of the cutaneous lesions observed in the present case (urticarioid papules), compared with the majority of cases reported in the literature (vesicular and bullous lesions).[4]

Concerning the second point, the association of diaminodimethyl-sulfone (DDS) and corticosteroids is at present considered the most effective regimen for the treatment of linear IgA bullous dermatosis (and of CP, independent of the immunopathologic findings).[4] This classic therapy was virtually uneffective in the reported case; on the contrary, the combined treatment with steroid and tetracyclines (recently emphasized in the treatment of BP[1]) was a determinant in the control of the evolution of the disease. It is extremely difficult even to speculate why and how this type of treatment works; however, it is important to consider this therapeutic choice when treating cases of linear IgA bullous dermatosis (or CP, independent of the immunopathologic profile).

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