

CASES FOR DIAGNOSIS

Vesicular Eruption Associated With Conjunctival Injection

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Patient History

A 30-year-old man consulted with a 20-day history of cutaneous lesions on the trunk accompanied by conjunctival injection and discomfort. The initial diagnosis was acute conjunctivitis, although topical antibiotics had no effect. He denied having taken medication in the days before the consultation.

Physical Examination

A noticeable eruption was observed with round erythematous-edematous lesions measuring approximately 1 to 2 cm in diameter, with clear vesicular blisters that were predominant at the periphery of the lesions (Figure 1). The eruption was well extended on the back and in isolated areas of the upper limbs and thorax, with sparing of the acral areas. The patient also presented conjunctivitis in the fornix. The ciliary body was not involved and there were no adhesions or leukomas.

Histopathology

A skin biopsy showed subepidermal detachment with predominant accumulation of polymorphonuclear neutrophils on the tip of the papillae, as well as a perivascular infiltrate of lymphocytes in the dermis (Figure 2). Immunofluorescence revealed a linear immunoglobulin (Ig) A deposit in the region of the basement membrane (Figure 3), but was negative for IgG, IgM, and C3.

Additional Tests

A blood analysis (complete blood count, biochemistry) and urine sediment analysis were requested, and the results were normal. The results of the immune workup—



Figure 1.

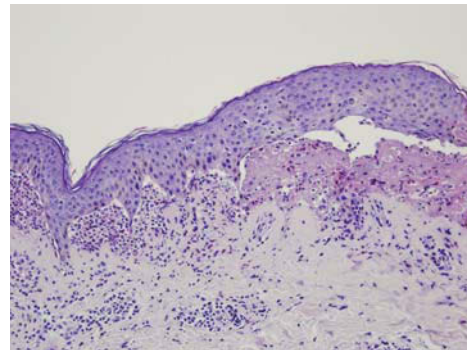


Figure 2.
Hematoxylin-eosin, $\times 40$.

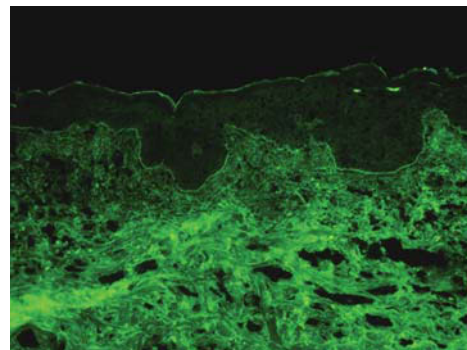


Figure 3.
Direct immunofluorescence for immunoglobulin A, $\times 40$.

antinuclear antibodies, complement components, cryoglobulin level, intercellular antibodies, anti-basement membrane antibodies, anti-tissue transglutaminase antibodies, and antigliadin antibodies—were negative for all tests.

What is your diagnosis?

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Diagnosis

Adult linear IgA dermatosis.

Course and Treatment

A second ophthalmological examination revealed conjunctival pseudomembranes and trichiasis associated with the underlying disease. Initial treatment was with topical corticosteroids, although the patient improved little, with the result that dapsone was started at 100 mg every 24 hours. The lesions on the skin and on the conjunctival mucosa resolved after a few days.

Comment

Adult linear IgA dermatosis is an autoimmune bullous disease that is closely linked with chronic bullous dermatosis of childhood and whose distinguishing feature, as occurs here, is a linear deposit of IgA in the basement membrane.¹

There have been reports of cases associated with digestive processes (celiac disease, inflammatory bowel disease²), collagen diseases (systemic lupus erythematosus, dermatomyositis³), hematologic or solid neoplasms,⁴ infections (varicella-zoster, tetanus), and drugs (especially vancomycin⁵). The pathogenesis is unknown and is associated with the formation of IgA antibodies targeting unknown basement membrane antigens, although collagen XVII (BP180) and its antigen LAD-1, and antigen BP230 and collagen VII (the latter are less common) are thought to play an important role.

The symptoms are varied and can be confused with those of bullous pemphigoid or dermatitis herpetiformis, although a typical pattern has been reported, with vesicles and blisters on an inflammatory base and a characteristic annular distribution (the image is often compared to a pearl necklace). The eruption is more often located on the trunk followed by the limbs. It is slightly pruriginous and has a rapid onset.

Mucosal involvement is common (affecting up to 80% of cases), especially on the conjunctival mucosa, which can occasionally heal leaving scars and synechiae.

Histopathology reveals the presence of subepidermal bullae and neutrophil aggregates (and occasionally eosinophil aggregates), with formation of microabscesses on the tip of the dermal papillae. Direct immunofluorescence reveals

linear IgA deposits on the basement membrane, and indirect immunofluorescence reveals the presence of anti-basement membrane IgA antibodies in 30% of cases.

The differential diagnosis should be made with other bullous conditions, such as dermatitis herpetiformis or bullous pemphigoid. Direct immunofluorescence is the most useful diagnostic tool. It can reveal a linear IgA aggregate in the basement membrane in the case of linear IgA dermatitis, linear C3 and IgG deposits in the basement membrane in bullous pemphigoid, and granular IgA deposits in dermatitis herpetiformis. Cases involving mixed aggregates (IgA, IgG, and IgM) can make diagnosis difficult,⁶ especially when there is predominant involvement of the mucosa, since it is then difficult to carry out a differential diagnosis with cicatricial pemphigoid. It seems likely that there may be overlapping forms for several of these entities.

As for treatment, most cases respond to sulfapyridine or dapsone after a few days, and oral corticosteroids must occasionally be added (especially in cases presenting a mixed IgG-IgA aggregate, probably because they are closer to the spectrum of bullous pemphigoid). Severe forms can be treated with azathioprine and cyclosporine.

Conflicts of Interest

The authors declare no conflicts of interest.

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