LETTERS TO THE EDITOR

Unilateral ocular cicatrical pemphigoid with circulating IgA and IgG autoantibodies reactive with the 180 kD bullous pemphigoid antigen

EDITOR.—Cicatrical pemphigoid (CP) is a rare, chronic, vesiculobullous disease that primarily affects the mucous membranes, particularly of the mouth and eyes.1 This disease is attributed to a subepidermal autoimmune phenomenon, and is characterised by in vivo deposition of antiepithelial basement membrane zone (BMZ) antibodies.2 Fibrosis in ocular cicatrical pemphigoid (OCP) produces shrinkage of the conjunctiva followed by shortening of the fornices, symblepharon, and cicatricial entropion.3 The disease usually affects both eyes.

We present a patient with OCP in one eye, who exhibited circulating IgA and IgG autoantibodies. These antibodies bound to a 180 kD antigen resembling that recognised by sera from patients with bullous pemphigoid (BP). Circulating IgA antibodies against the 180 kD BP antigen have not been reported in a patient with CP.

CASE REPORT

A 60-year-old Japanese man was referred to our hospital with redness, discharge, and irritation of the right eye that had been present for about 1 year. Physical examination revealed conjunctival injection, shortening of the fornix, symblepharon, and superficial punctate keratitis due to entropion of his right eye, while the left eye appeared normal. Impression cytology indicated a scarcity of goblet cells in the right conjunctiva but a normal amount in the left conjunctiva. Erosions were observed on the oral and nasal mucosa but there were no skin lesions.

A direct immunofluorescence was performed on a biopsy specimen of the right conjunctiva and revealed linear deposits of IgG, IgA, and C3 along the epithelial BMZ (Fig 1). An indirect immunofluorescence for identifying IgG and IgA antibodies in the patient's serum was performed using 1 M sodium chloride-split skin, described by Gammon et al.4 Both IgG and IgA antibodies in the serum reacted with the epidermal side of the split (Fig 2).

To identify the BMZ antigens, western blotting was performed as described previously.4 IgG in the patient's serum reacted relatively weakly but clearly with antigens with molecular weights of approximately 230 kD and 180 kD, and IgA reacted only with the 180 kD antigen (Fig 3). The 230 kD antigen co-migrated with the 230 BP antigen, while the 180 kD antigen co-migrated with the 180 BP antigen. Clinical manifestations and immunopathological findings led to a diagnosis of OCP.

COMMENT

Although patients with OCP usually exhibit bilateral ocular involvement, the disease is occasionally asymmetric as in the present case.2 Approximately 20%-30% of the patients with CP demonstrate serum anti-BMZ antibodies detected by indirect immunofluorescence test.1 Previous immunohistological studies suggested that the major CP target antigen is a 180 kD protein that demonstrates immunological cross-reactivities with the 180 BP antigen.5,6 IgG is the most common immunoglobulin observed. Smith et al.7 found circulating IgA antibodies against a 45 kD unknown protein in all seven OCP patients with exclusive ocular involvement.

We found that the IgA antibodies in our patient's serum reacted exclusively with the 180 kD BP antigen, while IgG antibodies reacted with both 180 kD and 230 kD BP antigens. Because IgA anti-BMZ antibodies are rarely detected in BP, the presence of IgA antibodies against the 180 kD BP antigen may account for the difference of clinical features between CP and BP. An alternative explanation is that the 230 kD and 180 kD BP antigens have several epitopes, and that circulating antibodies in patients with CP and BP, respectively, react with different epitopes. However, the possibility cannot be excluded that the 180 kD proteins which reacted with CP sera may be different proteins with similar molecular weights. Further investigations using cDNAs for these antigens will be needed to clarify the molecular structure of the antigens for CP.

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Conjunctival epithelial inclusion cyst arising from a pterygium

EDITOR.— Conjunctival epithelial inclusion cysts are not uncommon and may occur spontaneously or following ocular surgery, inflammatory conditions, or trauma.8 A patient who had rapidly developed a large conjunctival mass over his pterygium is described. Because

Figure 1 Direct immunofluorescence demonstrates linear deposition of IgA along the epithelial basement membrane zone.

Figure 2 Indirect immunofluorescence using 1 M sodium chloride-split skin showed that IgA antibodies in this patient's serum reacted with the epidermal side of the split.

Figure 3 Western blot analysis using human epidermal extracts. Two protein bands were specifically recognised by bullous pemphigoid (BP) sera: the 230 kD and 180 kD BP antigens (lane 1). Both BP antigens were recognised weakly by the IgG antibodies in the patient's serum (lane 2). Only the 180 kD BP antigen was recognised by IgA antibodies in the patient's serum (lane 3).
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