LETTERS TO THE EDITOR

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

EDITOR.—Cicatricial 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.¹ Fibrosis in ocular cicatricial pemphigoid (OCP) produces shrinkage of the conjunctiva followed by shortening of the fornices, symblepharon, and cicatricial entropion.² 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 have 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.*³ 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 kD BP antigen, while the 180 kD antigen co-migrated with the 180 kD 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 immunochemical studies suggested that the major CP target antigen is a 180 kD protein that demonstrates immunological cross reactivities with the 180 kD 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 the 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 antigen proteins are needed to clarify the molecular structure of the antigens for CP.

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Accepted for publication 19 April 1996

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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.¹ 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).

of this particular association, the mass also involved the cornea and caused significant astigmatism.

CASE REPORT

A 66-year-old male farmer noticed a red mass that grew rapidly within 2 weeks on his left eye. He also reported that his left visual acuity decreased progressively during the same period. He did not have any previous ocular surgery and did not recall any prior complaints related to his eyes.

On examination, the best corrected visual acuity was 20/40 in the right eye and 20/100 in the left. He had moderate nuclear sclerotic cataracts in both eyes. The right eye was otherwise normal. There was a non-tender, soft, reddish cystic mass on the nasal conjunctiva of the left eye (Fig 1). The surface of the mass was minimally keratinised due to exposure. There were dilated and distended blood vessels over the tumour. The mass measured 20 × 18 × 22 mm and protruded through the palpebral fissure preventing the closure of the eyelids. The tumour was attached to an underlying pterygium and extended about 4 mm onto the cornea. Transillumination confirmed the cystic nature of the tumour. Keratometry gave measurements of 50D at 35 degrees and 41D at 135 degrees.

During the excisional biopsy, the cyst could be separated easily from the pterygium that formed its base. The pterygium was also excised. The conjunctiva was then primarily closed. Postoperatively, his best corrected visual acuity rose to 20/40 in the left eye and the irregular corneal astigmatism disappeared.

Histopathologically there was one large cystic cavity lined by single or more layered flattened epithelium in which rare goblet cells were observed (Fig 2). The squamous nature of the lining epithelium could be observed focally. Few chronic inflammatory cells were scattered under the lining epithelium and substantia propria. The surface epithelium displayed hyperplastic changes. A clinicopathological diagnosis of conjunctival epithelial inclusion cyst spontaneously arising from a pterygium was made.

COMMENT

Epithelial inclusion cysts are frequently encountered benign conjunctival lesions. However, they can, rarely, involve the limbus or cornea.² In one large survey of biopsy proved lesions of the conjunctiva in an adult population, epithelial cysts comprised 22.5% of all acquired epithelial lesions and they constituted 80% of all cystic lesions.³ They occurred with almost equal frequency in men as in women and at an average age of 47 years.³

It is believed that conjunctival epithelial cysts can occur in the conjunctiva following

the stagnation of mucus in epithelial spaces formed by amalgamations of mucosal folds that result from irregularly elevated surface epithelium in inflammatory conditions.⁴ This may explain the development of an inclusion cyst over a pterygium as was the case in our patient. These cysts have also been reported to arise from dislocated epithelium below the surface of the conjunctiva or cornea secondary to trauma⁵ or scleral buckling surgery.⁶ Most cysts remain small and asymptomatic but occasionally they may reach large proportions and cause significant corneal astigmatism and visual impairment.⁷

Treatment consists of excisional biopsy, cryotherapy, or cauterisation with trichloroacetic acid.⁸ Nd-YAG laser has also been shown to be effective for acquired inclusion cysts of the conjunctiva.⁹

Conjunctival epithelial inclusion cysts can occur spontaneously over a pterygium and thus involve secondarily the cornea causing significant functional impairment.

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Accepted for publication 15 April 1996

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Figure 2 Histopathological section of the cyst showing the surface covered by multilayered squamous epithelium (short arrow). There is marked oedema in the stroma with congested veins and a few scattered inflammatory cells (long arrow). The cavity is lined by flattened squamous epithelium (double arrows) (haematoxylin and eosin, × 10).

Figure 1 A smooth, cystic mass on the nasal bulbar conjunctiva extending on to the cornea of the left eye. The surface vessels are engorged.

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Racemose haemangioma of the iris

EDITOR,—Benign vascular tumors are well known to occur in the retina and choroid where they can be classified into capillary, cavernous, and racemose types.^{1,2} Vascular neoplasms of the iris are very rare and most reported cases have been of the cavernous type. We report our observations of a patient with an arteriovenous lesion in the iris compatible with a racemose haemangioma.

CASE REPORT

An asymptomatic 40-year-old man was found on routine examination to have a vascular lesion in the iris of his left eye. His past medical history, family history, and systemic findings were entirely normal. Specifically he had no history or findings compatible with Wyburn-Mason syndrome, the von Hippel-Lindau syndrome, or other systemic vascular entities. There was no history of systemic or ocular vascular problems.

Slit-lamp biomicroscopy revealed three blood vessels that extended from the anterior chamber angle temporally to terminate in a small complex of intertwining vessels on the temporal aspect of the pupil. Although the vessels could be easily visualised with slit-lamp biomicroscopy, they were difficult to document photographically because they were in the iris stroma. However, fluorescein angiography demonstrated the vascular lesion more clearly. The central vessel was a distinct feeding artery that passed from the peripheral portion of the iris at the 3 o'clock location (Fig 1) and rapidly fed the complex mass of vessels near the pupillary margin. Two distinct veins, one at 2 o'clock and one at 4.30 o'clock, appeared to drain from the lesion and exit into the anterior chamber angle (Fig 2). There was no detectable leakage of fluorescein from the blood vessels. The diagnosis was racemose haemangioma of the iris. The lesion has remained stable for about 12 months since it was initially observed.

COMMENT

Intraocular haemangiomas usually occur in the retina or the uveal tract. Retinal haemangiomas can be subdivided into capillary, cavernous, and racemose, and acquired non-familial types, each of which has distinctive ophthalmoscopic features and systemic associations.¹ Uveal haemangiomas occur almost exclusively in the posterior choroid and are mostly of the cavernous type.²

Haemangiomas of the iris are generally considered to be quite rare. In 1972, Ferry³ obtained and reviewed histopathological sections of reported cases of iris haemangioma and found that virtually all of them had been misdiagnosed histopathologically. Many of the reported cases were found to be highly vascularised melanomas, juvenile xanthogranuloma,