CASE REPORTS

CICATRICIAL PEMPHIGOID
OCULAR RISK

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Two cases of cicatricial pemphigoid are reported. In one patient (case 1) the disease started as early as at 27 years of age. In both the cases oral mucosa was first affected and circulating anti BMZ antibodies were negative.

Key Words : Blistering disease, Pemphigoid, Cicatricial, Ocular pemphigus

Introduction

Cicatricial pemphigoid, also known as ocular pemphigus is a rare chronic blistering disease of skin and mucosae which results in permanent scarring of the affected area, particularly the conjunctiva.[1] It may thus produce a host of ocular complications e.g., trichiasis, symblepharon and entropion which may result in impairment of vision and even blindness.[2] Though it is called benign mucosal pemphigoid, its outcome turns out to be malignant in conjunctival mucosa. This could be the reason for naming it "Ocular pemphigus", the word 'pemphigus' connoting severe nature of the disease.[3] All cases of cicatricial pemphigoid do not present with ocular lesions at a given time, conjunctiva is eventually affected in approximately 75% of cases.[4]

We report herein 2 cases of cicatricial pemphigoid recently seen by us.

Case Report

Case I. A 31-year-old female presented in 1989 with generalized vesicobullous lesions and hoarseness of voice of 7 years duration. The disease started as painful erosions of oral mucosa and hoarseness of voice. Subsequently she developed vesicobullous lesions all over the body, and redness in both the eyes associated with burning sensation and mucoid discharge. She was diagnosed as a case of cicatricial pemphigoid. Tzanck smear was negative for acantholytic cells. Skin biopsy revealed subepidermal bulla with inflammatory infiltrate rich in eosinophils. Direct immunofluorescene (DIF) test showed linear deposit of IgG and C_3 along the dermoepidermal junction.

She was put on oral prednisolone, initially 60 mg daily increasing thereafter to 120 mg till the development of blisters was controlled and redness of the eyes subsided. Subsequently she received gradually tapering dose of prednisolone upto 25 mg daily for a period of 2½ years when she was lost to follow up. However, while on treatment, she had recurrent attacks of redness and discharge from eyes with gradual diminution of vision. This time (April, 1993) she presented with generalized bullous lesions with erosions over oral and nasal mucosae. Ocular examination revealed bilateral conjunctival scarring, symblepharon (Fig.1), entropion and trichiasis. Schirmer's test was 10 mm and 5 mm in right and left eyes

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respectively after 5 minutes. Direct laryngoscopy revealed laryngeal stenosis. She was given dexamethasone pulse therapy for 3 consecutive days. With pulse therapy

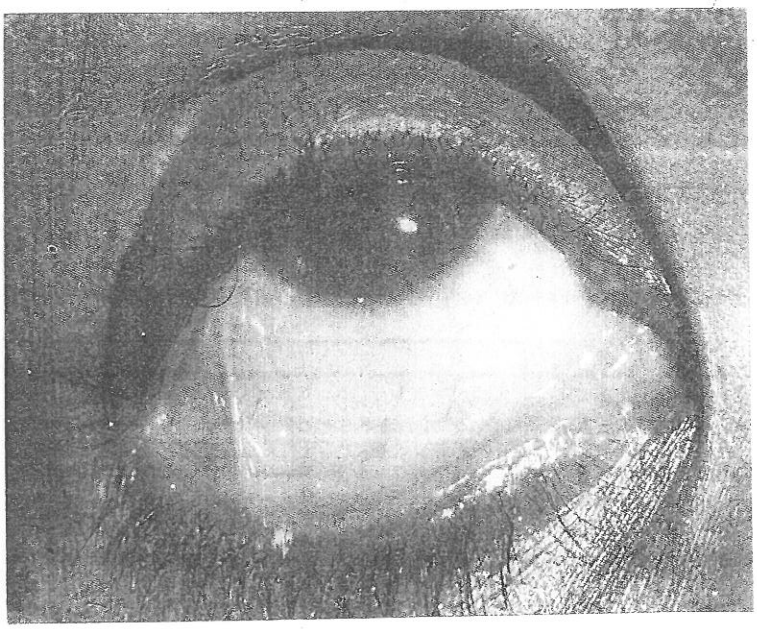

Fig. 1. (Close up) Conjunctival 'Cicatrix' over outer canthus of left eye

new blisters stopped coming and old bullae started healing. For conjunctival xerosis methyl cellulose eye drop (0.7%) was prescribed 6-8 times daily.

Case II. A 50-year-old male presented with ulceration over oral mucosa of 4 years, generalized vesicobullous lesions, conjunctival scarring and impairment of vision in left eye with mucoid discharge for last 4 years. Rest of the features were same as in case I. Examination revealed conjunctival scarring, trichiasis, symblepharon and entropion in the left eye, the right eye being normal. Schirmer's test was 5 mm and 15 mm after 5 minutes in left and right eyes respectively. Histopathology of skin was same as case I. Direct immunofluorescene (DIF) test showed deposition of IgG, IgA and C_3 at the dermoepidermal junction. He was put on daily 60 mg oral prednisolone and there was improvement of cutaneous lesions. However, oral erosions and conjunctival scarring persisted. Presently he is on follow-up and free of skin lesions. However, oral erosion and conjunctival scarring have persisted.

Discussion

Although cicatricial pemphigoid has been described as a disease of late middle age[2], one of our patients (Case 1) presented as early as at 27 years. In both the patients, the disease started in the oral mucosa and the early ocular changes were just like mucopurulent conjunctivitis.

The disease in general and ocular changes in particular progressed inspite of treatment. This is a unique feature of "Ocular pemphigus" as observed by others also.[2,5] In a study of 62 patients of cicatricial pemphigoid with eye involvement, 21 eventually became blind inspite of treatment, 17 in both eyes.[2] Both of our patients have been subjected to repeated opthalmologic check ups and only long term follow-up will reveal the ultimately outcome.

Contrary to ocular changes, vocal stenosis observed in Case I was nonprogressive and arrested by treatment.

Circulating antibasement membrane zone (BMZ) antibodies were negative in both of our cases as seen by others.[6]

In the first case, we had to resort to aggressive dexamethasone pulse therapy as she did not respond to oral corticosteroids. Though many more treatment modalities[1] have been tried, none has so far been able to prevent the serious ocular outcome of the disease.

References

1. Pye RJ. Bullous eruption. In: Textbook of Dermatology (Champion RH, Burton JL, Ebling FJG, eds), 5th edn. Oxford Blackwell scientific Publications, 1992 1652-5.

2. Hardy KM, Perry HO, Pingree GC. Benign mucous membrane pemphigoid. Arch Dermatol, 1971; 104 : 467-75.

3. Caroll JM, Kuwabara T. Ocular pemphigus. Arch Opthalmol 1968; 80 : 683-95.

4. Knee B. Ocular Pemphigus wjth scarring of the skin and mucous membranes. Arch Dermatol Syphilol, 1947, 55 : 37-41.

5. Person JR, Rogers RS III. Bullous and cicatricial pemphigoid : Clinical, histopathologic and immunopathologic correlations. Mayo Clin Proc, 1977; 52 : 54-66.

6. Griffith MR, Fukuyama K, Tuffanelli D, et al. Immunofluorescence studies in mucous membrane pemphigoid. Arch Dermatol, 1974; 109 : 195-9.