

CICATRICAL PEMPHIGOID (The First Case Report From India)

J. S. PASRICHA RAMJI GUPTA AND U. N. BHUYAN

Summary

This is the first case report of cicatricial pemphigoid from India. The patient was a 58-year-old male who developed tense, dome-shaped vesicles in the oral mucosa, followed 18 months later by blisters on the skin which healed by scarring. Symblypharon developed in both the eyes 23 months after the onset. Histologically, the split dermal, and indirect immunofluorescent test on the patient's serum for IgG antibodies was negative.

KEY WORDS: Cicatricial pemphigoid; Benign mucous membrane pemphigoid; Ocular cicatricial pemphigoid.

Introduction

Cicatricial pemphigoid, also described as benign mucous membrane pemphigus, ocular pemphigus, pemphigus conjunctivae, benign mucous membrane pemphigoid and ocular cicatricial pemphigoid is a well-known entity¹⁻⁶, but no case has so far been reported from India, creating an impression that this disease probably does not occur in India. We are reporting the first case confirmed by us.

Case Report

Since June, 1979, a 58-year-old male was getting recurrent vesicles and ulcers in the oral cavity. The vesicles were tense and varied from 2 to 5 mm in size. They were usually located on the soft and the hard palate, buccal mucosa, under surface of the tongue and mucosal surfaces of both the lips. Some of the vesicles were haemorrhagic.

Within 3-4 days, the vesicles would rupture producing painful deep ulcers. Treatment with 20-80 mg prednisolone a day resulted in only slight improvement. In December 1980, he started getting diffusely scattered, tense, dome-shaped vesiculo-bullous lesions on the skin of various parts of his body. The skin lesions would heal in 3-4 weeks leaving behind atrophic scars (Fig. 1). In May 1981, he developed ulcers on the lateral side of the lower conjunctivae in both the eyes which healed with the formation of symblepharon (Fig. 2).

Some of earlier biopsies from the oral mucosa and skin were inconclusive, but two skin biopsies done in May 1981, showed subepidermal splits (Fig 3).

Indirect immunofluorescent test on his serum for skin reactive antibodies using rat lip and anti IgG serum showed no staining on two occasions.

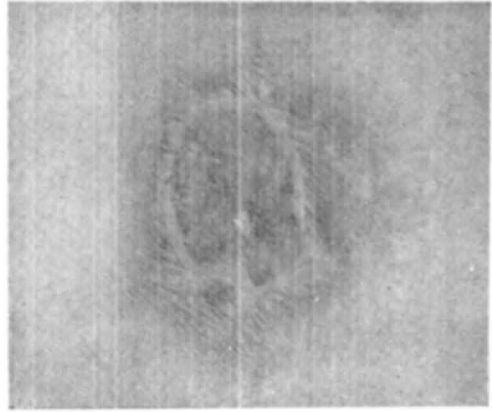
Discussion

In India, pemphigus has been observed in all parts of the country^{7,12}

Departments of Dermato-Venereology and Pathology,
All India Institute of Medical Sciences,
New Delhi-110021, India

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Fig. 1 A healed skin lesion showing an atrophic scar.



except the Rajasthan State where it is believed to be very rare. The reasons for this rarity are not known. Pemphigoid, dermatitis herpetiformis and benign bullous dermatosis of childhood are also frequently seen. Cicatricial pemphigoid, however, has never been reported. Presumably, it has not been diagnosed so far.

The patient being reported by us, showed sufficiently distinctive features to point towards the diagnosis. The oral vesicles were small and dome-shaped and persisted for several days before rupturing, in contrast to the pemphigus lesions which are almost never seen in the vesicular and the bullous stage. The skin lesions in our patient, as a rule healed with scarring

and never with hyperkeratosis and hyperpigmentation which is the classical pattern of healing in pemphigus. Histopathological localisations of the split at the dermo-epidermal level and negative staining for immunofluorescence further corroborated the diagnosis of cicatricial pemphigoid. Appearance of symblepharon completed the clinical picture still further.

It is important to emphasize that in spite of the old terms ocular pemphigus/pemphigoid for this disease, involvement of the conjunctiva is neither the most important sign of this disease, nor it is seen in every case. Cicatricial pemphigoid, therefore, seems to be the most appropriate term for this disease.

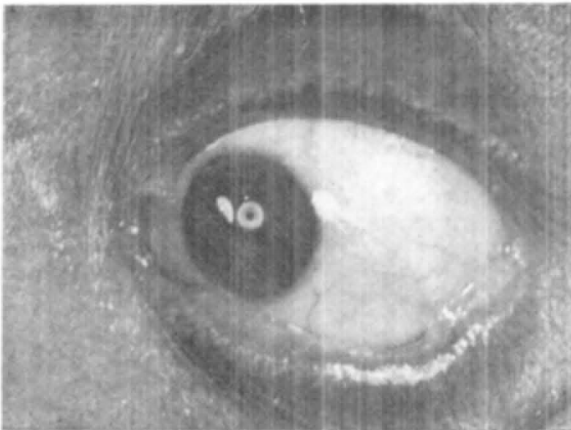


Fig. 2 Symblepharon in the lower eyelid.

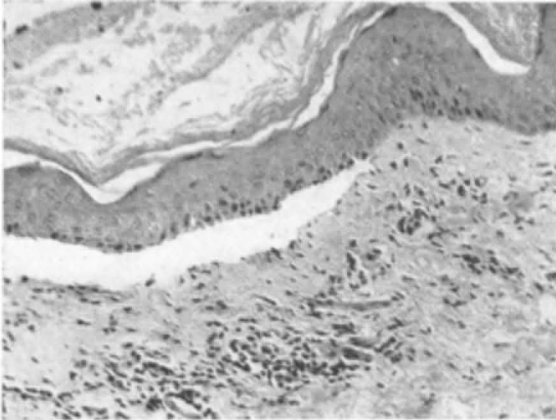


Fig. 3 Subepidermal split on histopathology.

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