MUCOUS MEMBRANE PEMPHIGUS

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PEMPHIGUS affecting the mucous membrane of the mouth and larynx is a disease rarely encountered in otolaryngological practice. Associated lesions of the conjunctival mucous membranes and the skin render the condition more commonly the province of the ophthalmologist and dermatologist.

Case Report

C.J., a female, aged 59.

The patient was first seen in December, 1963. She had complained of recurrent sore throats and had undergone tonsillectomy four years previously at another hospital. She admitted to fewer sore throats since. For the previous year she had had intermittent epistaxis with marked crusting in the nose and had been under treatment for conjunctivitis for four months. Recurrent sores on the hard palate had healed without scarring. Blisters repeatedly occurred on the skin of her nose; these healed spontaneously. She had lost almost two stones in weight in the previous three years.

Clinical examination of the eyes showed:

(*a*) inflammation of the conjunctivae, particularly peripherally,
(*b*) thickening of the eyelids,
(*c*) narrowing of the palpebral fissures,
(*d*) bands of adhesion extending from the lid margins to the limbus (Fig. 1),
(*e*) partial obliteration of the cul-de-sac,
(*f*) trichiasis, particularly of the right upper lid.

Anterior rhinoscopy revealed marked crusting of the nasal mucous membrane; attempts at removal of the crusts produced epistaxis. There was a circumscribed zone of superficial ulceration in the centre of the hard palate which was apparently healing. Indirect laryngoscopy showed a thin pale membrane covering the epiglottis which itself was overhanging obscuring the vocal cords and ventricular bands. No skin lesions were present at the first attendance.

Investigations included a full blood count which showed a microcytic anæmia with normal white cell and differential count. E.S.R. (Wintrobe) was 27 mm. in the first hour. W.R. and Kahn tests were negative.

A clinical diagnosis of pemphigus was made and it was decided to biopsy the epiglottis. Direct laryngoscopy was carried out under general anæsthesia and inflammation of all the laryngeal mucosa was noted; suction applied to the epiglottis tended to strip off the pale superficial membrane. A biopsy was taken

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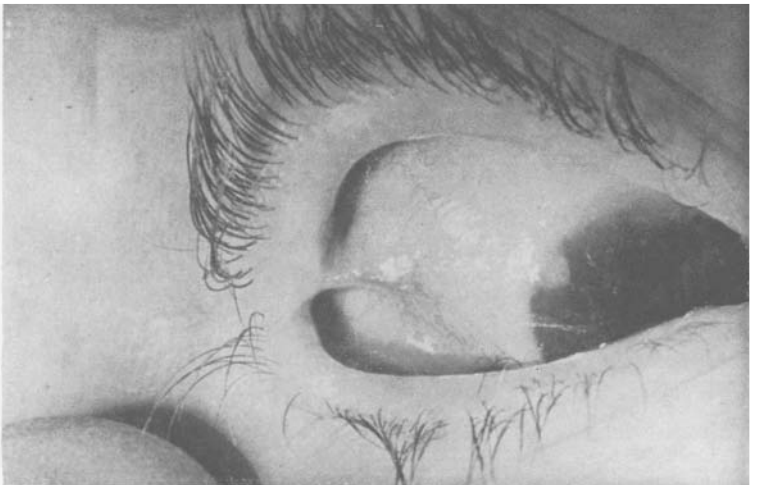


FIG. 1.
Conjunctival adhesion between lid margin and sclera.

from an untraumatized area on the upper edge of the epiglottis. The squamous epithelium, on microscopic examination, showed intra-epidermal bullae and the dermis was densely infiltrated with lymphocytes. The appearances were consistent with pemphigus (Fig. 2).

Treatment with Prednisolone 5 mg. t.d.s. was begun in January, 1964, and the patient immediately showed an improvement. Two weeks later the

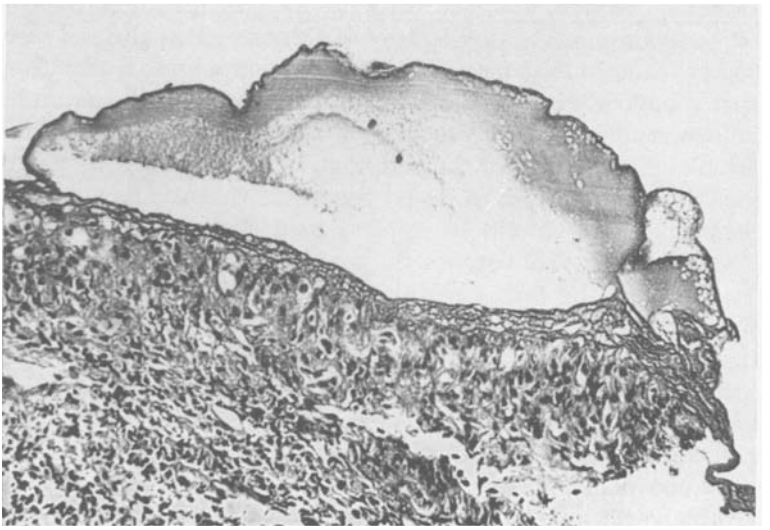


FIG. 2.
Biopsy of epiglottis showing intra-epidermal bulla (× 100).

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eyes were less sore and the inflammation had diminished. The dose of Prednisolone was increased by 5 mg. daily, however, when skin lesions appeared on the right ala nasi and abdominal wall adjoining the umbilicus. Both these lesions had begun as "water blisters" which had burst leaving ulcerated areas.

The patient was last seen in May, 1964, and up to that time no new lesions had appeared. The nasal mucosa was normal, no epistaxis having occurred since treatment began. No active inflammation was present in the eyes; the epiglottis was normal on examination. The patient has gained seven pounds in weight since treatment with steroids was started; the dosage is now being reduced until a satisfactory maintenance dose is arrived at.

Comment

The causative factor in pemphigus is unknown. The case here presented shows all the characteristics of so-called benign mucous membrane pemphigoid or the pemphigus conjunctivae of Lever (1954). Bullous lesions and denuded areas are present on the mucous membranes. In about half the cases lesions are present also on the skin. Scarring commonly develops on the conjuctivae, on the mucous membranes and occasionally on the skin. Scarring does not occur in any other form of pemphigus except very rarely, after severe secondary infection of the skin. The bullae are, according to Civatte (1943), subepidermal in the benign mucous membrane type but inter-epidermal in pemphigus vulgaris and its subdivisions, vegetans and foliaceus. The histologic feature of intra-epidermal bullae in this one case, at any rate, is at variance with his findings.

"Essential shrinkage of the conjuctiva" was first described by von Kries in 1878 and is a striking feature of this case. It is the end result of a pathological process that may be caused by pemphigus; it may, however, accompany bullous diseases other than pemphigus (erythema multiforme and bullous eruptions after vaccination); or it may be the end result of any violent inflammation of the conjunctiva accompanied by destruction of tissue. Klauder and Cowan (1942) suggested that the term "pemphigus of the conjunctiva" should be applied to a slow progressive shrinkage of the conjunctiva that occurs as part of pemphigus of the mucous membranes while the term "shrinkage" or "shrivelling" should be used to designate a similar process but one produced by a disease other than pemphigus.

In their review of 12 cases and in Franke's (1900) series of 88 cases compiled from the literature, the disease of ocular pemphigus was commoner in females, usually occurred in later life and had an incidence of one in 20,000 ocular cases. They further state that, although there is considerable variation in the course of the disease, progress may be rapid and blindness due to corneal involvement may ensue within two years.

Pemphigus vulgaris had a very high mortality before the introduction

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of steroid therapy. Lever and White (1963) have reported excellent results in so treating 46 patients between April, 1950 and October, 1959. They stress the importance of early diagnosis and prompt treatment with high initial doses of corticosteroids. Ocular pemphigus also responds satisfactorily to steroid therapy; treatment is warranted in view of the progressive nature and uncertain course of the disease and the hazard of further eye damage.

Summary

A case of mucous membrane pemphigus in which the conjuctiva, nose, mouth and larynx and skin were involved is presented and discussed. A satisfactory response to treatment with corticosteroids is recorded.

Acknowledgments

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