AMYLOIDOSIS is usually categorized as a degenerative process. Nevertheless, the nature and pathogenesis of this disorder remain most uncertain. Reimann, Koucky, and Eklund (1935) have classified amyloidosis in four groups: primary, secondary, tumour-forming, and associated with multiple myeloma.

Tumour-forming amyloidosis is known to occur as a primary process in the tissues of the eye, urinary bladder, tongue, and upper respiratory passages. Instances of localized amyloidosis involving ocular tissues after chronic inflammation are rare, and only a few have been recorded in the recent literature (Elles, 1945; Chinaglia, 1952; Handousa, 1954; Oppel, 1956).

A review of the literature suggests that amyloidosis as such is uncommon in India. Mathur and Bhende (1957) could trace only eighteen reported cases and added two cases of their own. In none of these cases was involvement of the ocular tissues described and to our knowledge no case of tumour-forming amyloidosis has been reported from India, although chronic inflammatory conditions of the eye are common.

Case Report

A woman aged 30 years complained of a gradually increasing swelling of the upper and lower lids of both eyes for the last 12 months and of difficulty in seeing. The swelling of the lids started from the inner angles and had involved the right side more than the left.

Examination.—The right side showed a uniform thickening of the upper and lower lids with a well-defined firm round nodule nearly 0.5 cm. in diameter on the medial third of the upper lid. The left eye also showed uniform thickening of both the lids and a similar nodule occupying the medial third of the upper lid (Fig. 1).

Fig. 1.—Bilateral amyloid tumour of the eyelids.

The nodules were not tender and were not adherent to the skin. A detailed examination of the eyes was not possible owing to the narrowed palpebral fissures. The palpebral conjunctiva could not be examined as it was not possible to evert the lids. Absence of

* Received for publication July 18, 1957.
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Epiphora suggested that the naso-lacrimal ducts were not involved. The patient denied having applied or injected any irritant to the eyes, and there was no history of any chronic infection in the eye or elsewhere.

General examination revealed no abnormality. The blood picture and urine were normal and serological tests for syphilis were negative.

A clinical diagnosis of "trachomatous folliculoma" or "fibromatous tumour" was made, the nodules were excised, and the lids were corrected.

Pathology.—The tissues consisted of four irregular pieces, each measuring nearly 0.5 cm. in largest dimension. They were uniformly yellowish in colour and of firm consistency. There was no obvious capsule around them. One of the cut pieces was treated with iodine which stained it brown and the colour changed to blue on addition of sulphuric acid thus suggesting the presence of amyloid in the tissue.

Histologically, the nodules were composed of large masses of a homogeneous, non-fibrillar, eosinophilic hyaline material lying beneath the conjunctival epithelium (Fig. 2).

The deposition was diffuse but predominated around the blood vessels (Fig. 3, opposite). The lumen of the blood vessels was markedly narrowed and often occluded because of the infiltration of the hyaline material into the vessel walls. The acini of the glands of Moll were separated by the infiltration of similar substance into the interstitial tissue around them (Fig. 3).

The adipose tissue of the lids also showed the hyaline substance around the walls of the vacuolated fat cells (Fig. 4, opposite). The epithelium of the palpebral conjunctiva showed thickening and squamous metaplasia in some places while in others it was thinned out. The epithelium also showed gland-like invaginations. Chronic inflammatory cells in which lymphocytes and plasma cells were prominent were seen lying in groups below the conjunctival epithelium, round about the blood vessels and within the diffuse hyaline mass of amyloid tissue. There was no cholesterol deposition or calcification, and no giant cell reaction. The hyaline masses showed the typical metachromatic reaction with Congo red which stained it pinkish-red.
Comment

The histological appearance and specific tinctorial reactions for amyloid show that the nodules were of localized amyloidosis of the eyelids.

The aetiology of amyloid degeneration of the ocular tissues is obscure. According to Oppel (1956), these tumour-like degenerative conditions develop on the basis of chronic conjunctivitis, and trachoma is present in 60–70 per cent. of such cases. In the cases of amyloidosis described by other
authors inflammatory conditions of the eye also preceded the degenerative change.

Lubarsch (1929) first clearly defined the difference between primary and secondary amyloidosis, the following being characteristic of primary amyloidosis: absence of antecedent or co-existent disease, involvement of mesodermal tissues (such as smooth and skeletal muscles, cardiovascular system and skin rather than liver, spleen, kidneys and adrenals), variability in the staining reactions of amyloid as opposed to their constancy in the secondary form, and a tendency to nodular deposition of amyloid.

It has been noted that primary and secondary amyloidosis are not sharply defined terms and considerable overlapping of the characteristic features described for each of them occurs in some cases (Koletsky and Stecher, 1939; King, 1948).

In the present case there seems to be an overlapping of these features. The localized nature of the tumour and the involvement of the mesenchymal tissues is in favour of primary amyloidosis, while the well-marked specific staining reaction for amyloid, association of chronic inflammatory cells, thickening of the lids, and absence of giant cell reaction around the masses of amyloid (Iverson and Morrison, 1948) point towards secondary amyloidosis. The primary inflammatory process in this case was probably trachoma—a condition frequently seen in India.

We are grateful to Mr. S. P. Sookraj for supplying the photograph of the patient.

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doi: 10.1136/bjo.42.7.433

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