A Rare Cause of Unilateral Upper and Lower Eyelid Swelling: Isolated Conjunctival Amyloidosis

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Amyloidosis is a term encompassing a group of disorders characterized by the extracellular deposition of a substance called amyloid in various tissues. A very rare cause of chronic ocular discomfort is amyloidosis. We treated a patient with the above complaint in whom both eyelids were diffusely swollen with no other chronic ophthalmic complaints. Transconjunctival biopsy revealed amyloidosis and was confirmed on congo-red staining. Since we are aware that surgical excision may cause eyelid cicatrization or secondary xerophthalmia, we explained the pros and cons of surgery to the patient. In view of the 12-year history and lack of other symptoms, the patient preferred to be kept under observation. Follow-up at two years did not reveal any significant increase in the size of the swelling. Therefore, we recommend that in a patient with amyloidosis with no visible/ocular impairment, the condition should be left alone without surgical intervention.

Key words: amyloidosis, cryotherapy, eyelid, radiotherapy, surgery

INTRODUCTION

Amyloidosis of the palpebral conjunctiva is a rare condition that results in chronic ocular discomfort. Various treatment modalities including conservative surgical excision, cryotherapy and superficial radiotherapy have been attempted with partial success. Amyloidosis results from the deposition of insoluble, fibrous amyloid proteins, nearly always in the extracellular spaces of organs and tissues. Due to extensive deposits, the organ attained tumourous weight. We present our experience with this rare case of a patient in whom both the upper and lower palpebral conjunctivae were involved with no systemic amyloidosis.

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CASE REPORT

A 43-year-old man presented with a diffuse swelling of the left lower eyelid for 12 years which, in the last six months, also involved the upper eyelid. Both palpebral conjunctivae were congested and the disease was entirely subconjunctival with sparing of the bulbar conjunctiva (Fig. 1). The disease was predominantly in the tarsal plates and the overlying skin was stretched. Careful history taking did not reveal any preceding chronic ophthalmic complaints. The patient had already undergone multiple biopsies elsewhere with inconclusive pathology reports. Transconjunctival biopsy at our hospital revealed the deposition of acellular, pale, eosinophilic, hyaline material beneath the conjunctival epithelium (Fig. 2). There was a sprinkling of mature plasma cells and lymphocytes. On congo red
staining, the material exhibited congophilia, which on polarisation showed an apple green birefringence suggestive of amyloid deposition. The birefringence was sensitive to potassium permanganate pretreatment, signifying secondary amyloid deposition. Immunoelectrophoresis revealed a normal electrophoretic pattern, the absence of a monoclonal band, normal immunoglobulin levels (IgA, IgM, IgG) and the absence of Bence-Jones proteins in the urine. Computerized tomography of the eye showed a soft tissue mass in both left eyelids with no intraorbital extension. The extraocular muscles, eyeball, retrobulbar area and optic nerve were normal. To rule out the systemic involvement of amyloidosis, urine protein, blood counts, an ultrasound scan of the abdomen and pelvis and an echocardiogram were performed and were found to be normal. The patient was advised of surgery, however after having the benign nature of the disease explained, and in view of the twelve-year history, he insisted to be kept under observation. On follow up, the lesion appears to be static.

**DISCUSSION**

Amyloidosis of the conjunctiva is extremely rare.1-3,6,7 This has previously been reported by few authors as an unusual cause of ptosis1 and equally uncommon manifestation of trachoma.4 The term “amyloidosis” was coined by Virchow in 1854 on the basis of color after staining with iodine and sulfuric acid.5 In 1968, Macoul and Winter reported a case of primary ocular manifestation in systemic amyloidosis. The first case of localized ocular amyloidosis was reported in 1871. The prevalent classification of amyloidosis has been based on clinical features, using the nomenclature suggested at the Sixth International Symposium on Amyloidosis in Oslo, 1990.8

Primary amyloidosis is known to involve all ocular structures. There is a thinning of the tear film with decreased lacrimation with the involvement of the lacrimal gland. It usually affects the central cornea and the vision decreases as the deposits increase in size and density.9

Localized conjunctival amyloidosis is apparently the most common non-familial ophthalmological manifestation of amyloidosis. The disease is usually unilateral as in our patient, but may involve both eyes. It usually begins in the fornix and extends into the bulbar and palpebral conjunctiva.9 Patients typically have a long history of slowly enlarging tumefaction of the eyelids, decreased tearing with multiple biopsies and surgeries performed.2 In our patient as well, he gave a past history of multiple biopsies. A review of the literature showed that treatments varied from conservative local excision,1,2 debulking9 cryotherapy to superficial cobalt therapy,4 but with partial success.

The disease is known to recur locally2 with persistent ocular complaints that could perhaps be attributed to conservative local excision and persistent disease. Systemic examination in all these patients was normal.4 The etiology and pathogenesis of the disease is still unclear. Surgery is still the gold standard of management that should be as con-
servative as possible, although infrequent recurrences have been reported.1,2

REFERENCES