



## Advanced Localized Primary Conjunctival Amyloidosis: Surgical Rehabilitation Using Mucous Membrane Graft Implantation

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Amyloidosis is an extracellular accumulation of heterogeneous, amorphous, proteinaceous material in various organ tissues. The ocular adnexa can be involved primarily or secondarily in the disease process. Amyloid may accumulate in the eyelid, conjunctiva, and anterior orbit. Primary localized conjunctival amyloidosis is a rare form of the disorder. The abnormal protein is confined to the substantia propria and around blood vessels of the conjunctiva. The lesions are usually seen as solitary or multiple, often bilateral, well-vascularized, fusiform or polypoid, painless subconjunctival elevations [1]. The disease may present as spontaneous subconjunctival hemorrhage, yellow subconjunctival or orbital mass, thickened lid and blepharoptosis. Ptosis in ocular amyloidosis can be attributed to several mechanisms. The recurrent swelling of the eyelid due to frequent hemorrhages causes stretching and dehiscence of the levator aponeurosis from the tarsal plate [2]. A large amyloid mass can add considerable weight to the lid and cause mechanical ptosis. Direct infiltration of the muscle by the amyloid material may lead to myogenic ptosis [3].

Ocular surface irritation may arise due to continuous rubbing of the tarsal subconjunctival mass against the cornea, as well as shortening of the fornices, symblepharons, insufficient tear lake, and dryness. We report two patients with primary localized conjunctival amyloidosis, and describe their clinical

presentation and surgical treatment, with emphasis on the role of mucous membrane grafts in rehabilitation of the anterior segment.

### Patient Descriptions

#### Patient 1

A 63 year old woman presented with a 2 month history of right eye 2 mm ptosis. On examination, the upper lid showed a high crease and good levator excursion, consistent with aponeurotic dehiscence. When the lid was everted an irregular infiltrative yellow mass was noted, measuring 35x15 mm, that involved the tarsal conjunctival surface, and extending to the superior fornix and plica semilunaris [Figure A]. Ocular examination including visual acuity, extraocular movements, pupils, intraocular pressure and funduscopy was unremarkable.

Incisional biopsy was made. Histopathologically, the conjunctival epithelium was normal. The substantia propria was infiltrated by amorphous eosinophilic material. Congo red stain was positive, and a green birefringence of the stained material, which was observed by polarizing microscopy, confirmed the diagnosis of amyloidosis. Evaluation for systemic amyloidosis was negative. Due to continued complaints of foreign-body sensation, despite intensive lubrication, and because of restriction of superior visual field in her right eye, surgery could no longer be deferred. At surgery the lesion was removed carefully from the bulbar, palpebral

and forniceal conjunctiva. There was no clinical invasion of the tarsus. A buccal mucous membrane graft was implanted to reconstruct the superior fornix.

The ptosis was repaired primarily through an anterior lid crease approach. The skin, orbicularis and levator muscles were found to be clinically spared of amyloid infiltration. The levator aponeurosis was advanced and secured to the superior tarsal plate.

The patient's ocular irritation and inconvenience were relieved almost immediately in the postoperative period. At 2 years follow-up the patient retained good eyelid level and contour; no recurrence of lesions was observed.

#### Case 2

A 25 year old woman presented with a lower left eye forniceal subconjunctival lesion [Figure B]. The rest of the ocular examination was within normal limits. Incisional biopsy of the lesion, and similar stainings as in patient 1, showed amyloidosis. Evaluation for systemic disease was negative. Three months after initial presentation she developed cicatricial entropion of the lower eyelid, with rubbing of the lashes against the cornea, due to shortening of the posterior lamella.

At surgery we carefully dissected the rest of the lesion off the surrounding tissues. Entropion was repaired with an implantation of a hard palate graft. The patient recovered quickly from the foreign body sensation. At 9 years follow-up the



**[A]** Amyloid infiltration involving the upper tarsal conjunctival surface and extending to the superior fornix and plica semilunaris, in patient 1.



**[B]** Amyloid infiltration involving the lower forniceal conjunctiva, in patient 2.

patient kept a 5 mm deep inferior fornix and the lower lid remained stable, without any symptoms.

## Comment

Some patients with primary localized conjunctival amyloidosis may remain stable for long periods and require only medical treatment, such as topical steroids and lubricants [1]. Others may experience continuous growth of the lesions, recurrent hemorrhages, functional and cosmetic deformity of the lids, and require repeated surgical intervention [4]. In the past, surgery was made difficult by the tendency of the lesions to recur and bleed during the operation.

Data published in recent years show that careful debulking of deposits with a spooned curette, preservation of anatomic planes, avoidance from normal lid tissue sacrifice [5], and careful dissection with the 'Colorado' microdiathermy needle [3] can minimize the surgical complication rate.

When bare sclera or tarsus are left after curettage of the conjunctiva, intensive lubricant use and bandage contact lens are needed to relieve foreign-body sensation [1]. These measures might not suffice to completely prevent formation of symblepharons during the healing process due to adherence of apposed surfaces of raw epithelium. Delayed epithelialization of the ocular surface may require a prolonged rehabilitation period.

Our present report, like a previous one [4], demonstrates that primary implantation of a mucous membrane graft to reconstruct the superior fornix (patient 1), and secondary hard palate implant to deepen the inferior fornix and repair lower lid malposition (patient 2), can shorten the rehabilitation period. Our two patients reported immediate relief in the early postoperative period. This technique may also enable the surgical removal of large areas of tissue involved in the advanced form of the disease. Reconstruction of the fornices is especially important in light of the chronic nature of the disease and

the tendency of some patients to require additional surgery.

In the presented patients surgical intervention brought immediate and long-standing relief. It is likely that conservative management would have been less effective, eventually requiring surgery. We suggest that surgical intervention using mucous membrane graft implantation in advanced cases of primary conjunctival amyloidosis will provide patients with longer symptom-free periods and they will benefit more rapidly and to a greater degree from ocular surface rehabilitation.

## References

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