Primary localised conjunctival amyloidosis presenting as unilateral ptosis

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Abstract
A 23 years old female presented with watery discharge from right eye and mass in the medial aspect of eye with drooping of right upper lid. On examination, the patient had moderate ptosis of right upper lid with a pinkish red multi-lobulated, friable mass, bleeding to touch involving the medial canthus. CECT showed ill-defined mild enhancing soft tissue attenuating lesion involving and encircling the anterior aspect of the right globe. Patient was taken up for excision biopsy which revealed it to be amyloidosis. Systemic investigations turned out to be normal. This case is being reported here for its rarity.

Keywords: Amyloidosis, conjunctiva, ptosis.

Introduction
Amyloidosis is an accumulation of heterogenous, amorphous, proteinaceous material in extra ocular space or any tissue.\(^{12}\) Amyloidosis has mainly two forms, systemic and localized. Systemic amyloidosis is a serious and life-threatening condition because of destruction of tissue and their function due to accumulation of amyloid material while localised amyloidosis has very good prognosis which frequently include head and neck area without systemic manifestation. Primary localized conjunctival amyloidosis is a rare subtype in which abnormal amyloid material accumulates in substantia propria and around conjunctival vessels, it presents as painless, well- vascularised subconjunctival lesion which can be solitary or multiple and usually bilateral. Sometimes it may present with subconjunctival haemorrhage, yellow subconjunctival mass, orbital mass, lid thickening and blepharoptosis. Several factors which attribute to ptosis are frequent episodes of haemorrhage causing recurrent eyelid swelling resulting in levator aponeurosis dehiscence from tarsal plate. Other causes may be a large amyloid mass causing mechanical ptosis or muscle infiltration with abnormal amyloid causing mechanical ptosis. Continuous rubbing of conjunctival mass may cause ocular surface irritation.\(^{18}\)

Here, we present a rare case of primary localised amyloidosis presenting as unilateral ptosis with histopathological finding suggestive of extracellular amorphous and eosinophilic hyaline deposits underneath the conjunctival epithelium in substantia propria. The tissue stained pink with Congo red staining and showed apple green bi-refringences under polarised light, thus confirming the diagnosis of amyloidosis.

Case Report
A 23 years old female presented to our eye OPD with watery discharge from right eye and mass in the medial aspect of upper lid, medial canthus and inferior fornix. On examination, the patient had moderate ptosis of right upper lid with a pinkish – red multi-lobulated, friable mass, bleeding to touch involving the medial canthus. On evertting the upper lid the mass was seen to be involving medial half of the right upper lid till the superior tarsal edge. Ptosis was moderate with good LPS action (Figure 1 and 2).

Fig.1: Right eye showing ptosis

Fig.2: Pinkish lobulated mass in medial canthus
Ocular movements were full in all directions. A probable diagnosis of conjunctival papilloma/conjunctival squamous carcinoma was kept and CECT orbit was advised. CECT showed ill-defined mild enhancing, iso to hypodense soft tissue attenuating lesion involving and encircling the anterior aspect of the right globe with mild oedematous changes in the preseptal segment of right upper lid(Figure 3). Tiny calcific foci were seen in right upper lid with a probable diagnosis of neoplasm. Patient was taken up for excision biopsy. The lesion was removed carefully from the bulbar, palpebral and forniceal conjunctiva(Figure 4).

![CT scan showing soft tissue hypodense mass medially in right eye.](image1)

Fig.3: CT scan showing soft tissue hypodense mass medially in right eye.

![Right eye after excision of the mass](image2)

Fig.4: Right eye after excision of the mass
It was found not to be involving the deeper tissues (deeper to conjunctiva) of the upper lid. Histopathological examination showed extracellular amorphous and eosinophilic hyaline deposits underneath the conjunctival epithelium in substantia propria (Figure 5).

![Histopathology of conjunctival tissue showing eosinophilic hyaline deposits and amorphous material](image3)

Fig.5(a): Histopathology of conjunctival tissue showing eosinophilic hyaline deposits and amorphous material

![Higher magnification of biopsy specimen](image4)

Fig.5(b): Higher magnification of biopsy specimen lesion was composed of a few foci of calcifications. The tissue stained pink with congo red staining and showed characteristic apple green bi-refringences under polarised light suggestive of amyloid deposition. To rule out systemic amyloidosis, certain specific tests were done like urine routine analysis, CBC, abdomen and pelvis ultrasound scan and ECG, which all were normal.

**Discussion**
Localized amyloidosis is a rare disorder with orbit involvement seen in 4% of all cases involving head and neck region. Primary amyloidosis can involve all ocular structures.

Usually focal orbital amyloidosis has deposition in eyelid or conjunctiva and in superior portion of the orbit. Typically, patient presents with unilateral or bilateral solitary or multiple firm, rubbery, painless bleeding mass. It is usually found in young and middle age group and rarely in old age. Most common non familial ophthalmic manifestation of amyloidosis is the localised form in conjunctiva. Condition is usually unilateral as in our case. Our patient had ptosis which does not seem to be due to neurological cause because there is no restriction of movement in any gaze. The amyloid deposition in levator palpabrae superiosis probably attributes to the ptosis in the patient. Amyloidosis must be considered as a differential diagnosis of conjunctival neoplasm.

Various treatment modalities are available for amyloidosis including conservative local excision, debulking, cryotherapy, and superficial cobalt therapy. Surgical complications have been minimized due to careful debulking of deposits with a spooned curette, preservation of anatomic planes, avoidance of normal lid tissue sacrifice and careful dissection with diathermy needle. Mass excision is the
gold standard but it should be as conservative as possible. Total excision of the lesion is usually impossible and surgery should be performed to excise main part of the lesion with preservation of the palpebral lobe of the lacrimal gland, the levator palpabreae superioris and the extraocular muscles.\(^ {13-15}\) Congo red staining is the best method in histopathological examination with findings of a green birefringence under a polarizing light. Histopathological findings in the case were consistent with amyloidosis. Calcification was probably due to chronicity. Systemic examinations and laboratory tests were normal which excludes systemic amyloidosis.

References


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