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Acquired progressive unilateral blepharoptosis

Case Report

A 37-year-old healthy lady was referred for left eye (OS) upper lid ptosis correction with history of gradually progressive upper lid drooping for 2 years and, an episode of subconjunctival hemorrhage. There was no history of trauma, allergy, diurnal variation or generalized weakness. Best-corrected visual acuity was 20/20 in both eyes (OU). External ocular examination revealed significant blepharoptosis in OS [Fig. 1a]. Eversion of the upper lid revealed a yellowish-pink sub-conjunctival lesion throughout the length of the upper edge of the tarsus [Fig. 1b]. Anterior segment evaluation of right eye (OD) showed a yellowish-pink subconjunctival mass near the caruncle, extending into medial bulbar conjunctiva and inferior fornix [Fig. 1c and 1d]. Fundus evaluation was unremarkable.

What is Your Next Step?

A. Observe with regular follow-up
B. Blepharoptosis correction OS
C. Incision biopsy OU
D. Excision biopsy and cryotherapy OU.

Findings

In the presence of yellowish-pink subconjunctival lesion with an episode of subconjunctival hemorrhage, our clinical suspicion was conjunctival amyloidosis. Complete excision of the lesion was performed in OD followed by cryotherapy. Careful debulking to preserve LPS function and cryotherapy was done in OS. Histopathological examination revealed a positive Congo red staining of the specimen with apple green birefringence in polarized light, which is pathognomic of amyloidosis. Systemic evaluation was within normal limits.

Diagnosis: Bilateral Primary Localized Conjunctival Amyloidosis.

Correct Answer: D

Discussion

This case highlights the importance of bilateral detailed ocular examination in all cases of blepharoptosis. Upper lid eversion in a droopy eyelid aided in the clinical diagnosis.

Amyloidosis results from the localized (one organ) or systemic (multiple organ) deposition of amyloid fibrils in the extracellular spaces. Three major types include: primary (AL: amyloid light chain), secondary (AA-amyloid A), and heredofamilial (ATTR- amyloid transthyretin).[1] Ocular adnexal amyloidosis accounts for 4% of the localized cases. Most common sites involved are eyelids, orbit and conjunctiva presenting as conjunctival mass, ptosis or subconjunctival hemorrhage.[2] Misdiagnosis as ocular surface malignancy is common.[1] Excision or debulking with cryotherapy, depending on extent of the amyloid deposit is the treatment of choice. Systemic amyloidosis can involve heart, liver, kidney, nerves and gastrointestinal tract, causing vital organ dysfunction and mortality. Ocular findings can be the initial manifestation of life threatening systemic involvement in 10% that should be ruled out in all cases.[1,3]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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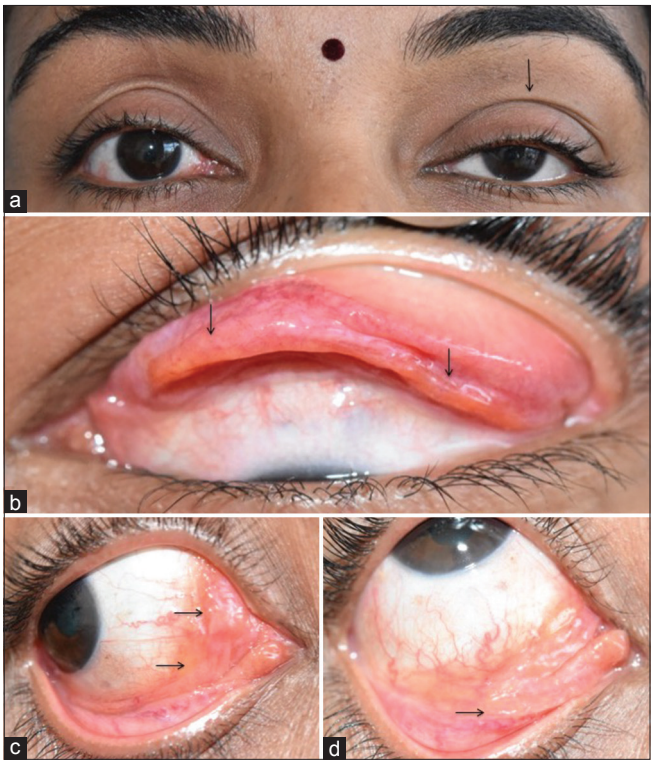

Figure 1: (a) Showing significant blepharoptosis of left eye (OS) in a 37-years-old lady. (b) On eyelid eversion there is yellowish pink sub-conjunctival lesion visible in the upper border of the tarsus extending into the upper fornix. (c and d) Similar appearing lesions are seen in the right eye (OD) involving the caruncle, medial bulbar and inferior forniceal conjunctiva

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Conflicts of interest

There are no conflicts of interest.

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