

Case Report

A case of localized amyloidosis of the eyelid misdiagnosed as recurrent chalazion



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Abstract

Localized amyloidosis of the eyelid is uncommon and is classically associated with systemic manifestations. We present an interesting case of a localized eyelid mass misdiagnosed as a recurrent chalazion presenting in an 85-year-old Saudi gentleman with no definite associate findings suggestive of an underlying systemic amyloid disease. Debulking surgery was subsequently performed. Proper diagnosis was reached based on the histopathologic examination of the excised tissue, which demonstrated the typical Congo red staining of the amyloid deposits.

Keywords: Eyelid, Amyloid, Chalazion, Dermatochalasis

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Introduction

Primary amyloidosis is a rare disease entity, with the global yearly incidence of amyloidosis estimated to be five to nine cases per million patients. Amyloidosis is characterized by deposition of insoluble amyloid fibrils into body tissues leading to impairment of function. It can be either systemic with deposits throughout the body (most commonly renal and cardiac) or localized to a single site, which is infrequent in the periocular area.¹

We are reporting this case of primary localized amyloidosis of the eyelid as an uncommon presentation at King Abdulaziz University Hospital in Riyadh, Saudi Arabia.

Case report

An 85-year-old hypertensive gentleman was referred from a screening clinic with a right upper eyelid mass. The patient was complaining of worsening vision in both eyes for the past 4 months. He had longstanding history of misdirected

eyelashes that were epilated at home and a previous history of chalazion excision in the same eyelid 3 months earlier.

His past medical history included hypertension and high cholesterol. He had undergone aortic valve replacement 2 years earlier and was stable on oral anticoagulant.

On ocular examination, the vision was poor in both eyes (counting fingers near the face, OD and counting fingers at 8 feet, OS). Intraocular pressure was normal in both eyes. External examination revealed a firm mass in the right upper eyelid along with dermatochalasis, bilateral upper lids entropion and rubbing lashes (Fig. 1a and b). He demonstrated a poor tear meniscus. Slit lamp examination showed superficial punctate keratopathies (SPKs) bilaterally, mature cataract in the right eye and early cataractous changes in the left eye. His fundus examination was not possible in the right eye and showed normal findings in the other eye.

Based on his presentation, a provisional diagnosis of chalazion was made. The lesion was excised and sent for histopathology which showed a soft tissue mass consisting mostly of adipose tissue infiltrated by amorphous deposits.

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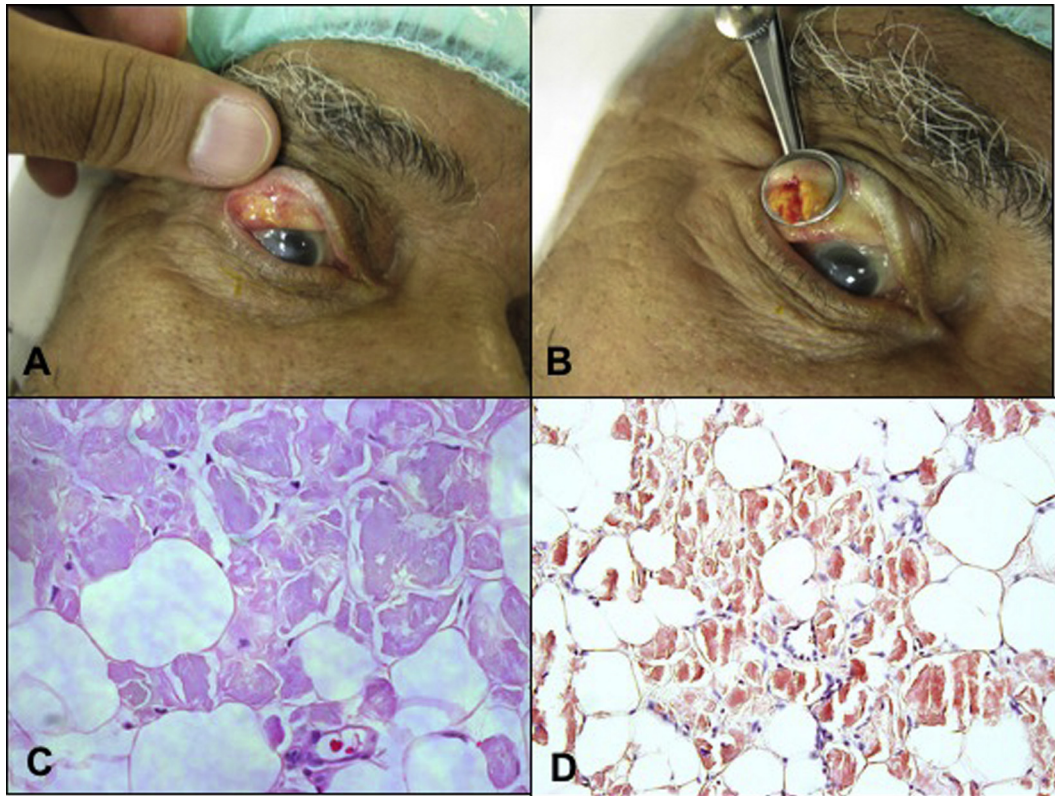


Fig. 1. (A): The clinical appearance of the chalazion-like mass through the conjunctival side of the right upper lid; (B): Intraoperative appearance of the mass using the eyelid clamp with yellowish contents; (C): The histopathology of the mass soft tissue excised showing amorphous material between fat cells (Original magnification $\times 400$ Hematoxylin and Eosin); (D): Confirmation of the Amyloid amorphous material between fat cells (Original magnification $\times 200$ Congo red).

The deposits stained positive with Congo red stain and showed birefringence with polarizing light, typical of amyloid deposits (Fig. 1c and d).

He subsequently underwent bilateral upper eyelid entropion repair because of his other symptoms. He was referred to his local hospital for systemic evaluation, which ruled out the presence of systemic amyloidosis, however the details of which were not available to us at the time of this report. In the one week follow-up period, his vision remained mostly the same, he demonstrated good upper lids position and the sutures were removed. There was no recurrence of his right upper lid mass and no other symptoms or findings suggestive of systemic amyloidosis for a total follow-up period of 6 months. The patient elected to have his cataract surgery in his local hospital.

Discussion

Although amyloidosis is an uncommon occurrence, ocular structures can be involved as either part of a systemic disorder or localized amyloidosis. Periocular involvement has been infrequently reported in the literature. The most common presenting symptoms in these cases are visible mass and ptosis. Other signs include the following: pain and discomfort, subcutaneous hemorrhages, dry eye, restricted motility, pupillary abnormalities, proptosis and globe displacement². The course is often prolonged, with a mean duration of 31 months at presentation.³

Due to its diverse symptomatology, diagnosis can be challenging and a high clinical index of suspicion is needed.

Definitive diagnosis is based on tissue biopsy using Congo red staining. The red-green dichroism under unidirectional polarized light is pathognomonic for amyloidosis. In cases of eyelid skin involvement, work-up for systemic amyloidosis is warranted as it has a high affinity for this particular site.² Even with advancing methodology for the identification of the type of amyloid, Congo red stain remains the gold standard for initial identification of the deposits such as in our case.⁴

Management options are diverse and should be individualized. Since the general disease behavior in cases with amyloidosis is slow progression of the accumulated material, conservative management and observation can be an option for many patients depending on the site involved. More progressive cases especially in the periocular area are better managed by surgical debulking such as in our patient.⁵ Besides controlling the underlying disease if any, the main goal of surgical therapy is to preserve function and might be for cosmetic indications.³ One of the reported observations related to conjunctival amyloidosis surgical excision is the tendency for bleeding, which hasn't been a constant encounter.⁶ More invasive therapies such as radiation have been also described to reduce the risk of recurrence.⁵

Primary dermal amyloidosis of the eyelid is not a common occurrence and should be entertained as a differential diagnosis in cases of eyelid and/or periocular masses. Due to its rarity and in the absence of typical features in this case (such as ptosis and irritation), histopathology was the only clue.⁵

Unlike other periocular sites, systemic amyloidosis has a unique predilection for the eyelid skin.² This does not seem

to be the case in our patient, since an underlying systemic amyloidosis was ruled out in his local hospital. The other interesting findings in our case were the initial misdiagnosis as chalazion and the associated dermatochalasis, which has been previously reported in association with eyelid amyloidosis.⁷

In conclusion, this is one of the relatively rare cases of presumed primary dermal amyloidosis of the eyelid, which was clinically unsuspected until tissue diagnosis was obtained and was associated with dermatochalasis. Ophthalmologists should add amyloidosis in the differential diagnosis of an eyelid mass.

Conflict of interest

The authors declared that there is no conflict of interest.

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