CASE REPORT

A CASE REPORT OF PRIMARY LOCALIZED AMYLOIDOSIS OF EYELID WITH EXTENSIVE CALCIFICATION AND OSSIFICATION, CLINICALLY PRESENTING AS A BONE TUMOR

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Abstract- Primary localized amyloidosis of eyelid is a localized type of amyloidosis without evidence of systemic involvement. We report a 75-year-old man suffering of unilateral (left) ptosis due to upper eyelid mass with bony consistency. Eye examination revealed upper lid mass attached to tarsus. No ocular infection or inflammation was found. Pathologist reported diffuse distribution of eosinophilic homogenous deposits of stroma that show apple green birefringence by polarized light compatible with amyloidosis. Primary localized amyloidosis may appear as a bony tumoral mass. It is important to consider secondary localized and systemic amyloidosis in differential diagnosis of the disease.

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Key words: Amyloidosis, eyelid, Congo-red staining

INTRODUCTION

Amyloid is an extracellular eosinophilic homogeneous matrix material that can be deposited in connective tissues and blood vessels anywhere in the body (1-3). This tissue change may have many different causes that are grouped under the term amyloidosis (1). Amyloidosis is classified as systemic and localized (1, 4, 5). In systemic amyloidosis multiple organs may be involved but in localized amyloidosis only one body organ is involved. Primary amyloidosis is an uncommon condition (1, 6, 7) without any systemic or antecedent disorder. Secondary amyloidosis is preceded by chronic inflammatory conditions such as rheumatoid arthritis or is associated with plasma dyscrasia (1).

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Primary localized amyloidosis of eyelid develops slowly in healthy, young or middle age (occasionally elderly) persons on a hereditofamilial basis without evidence of systemic amyloidosis (1, 7). It may be followed by ptosis (1, 7-9). In differential diagnosis, secondary localized amyloidosis due to longstanding chronic inflammation like trachoma and systemic amyloidosis must be considered (1, 10).

CASE REPORT

A 75-year-old man referred to Farabi Eye Hospital suffering from unilateral ptosis due to left upper eyelid mass with palpebral conjunctival (tarsal) attachment. He didn't have any systemic symptoms and signs (Fig. 1). Mass had a bony consistency. Eye examination revealed no ocular/visual infection and inflammation. The patient had no systemic signs and symptoms and all hematologic and biochemistry laboratory tests were within normal limits.

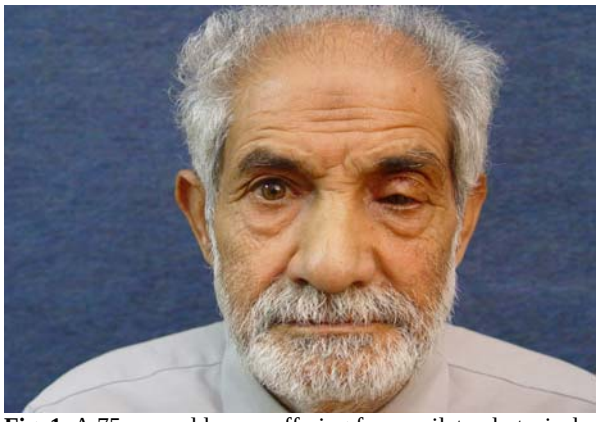

Fig. 1. A 75-year-old man suffering from unilateral ptosis due to left upper eyelid mass.

First, an incisional biopsy of left eye mass was performed for the patient. In this procedure a tumor with very hard consistency on the tarsus was seen. Tumor and attached conjunctival area were biopsied. The specimens were sent to pathology laboratory, then incision was sutured with 6/0 nylon. After ten days, in second surgery, Weise procedure (blepharotomy) was performed.

The specimen received in formalin solution consisted of two creamy yellow colored tissues with hard and bony consistency measuring 0.7×0.4×0.2 and 0.3×0.2×0.1 cm, respectively, totally embedded in one block. Microscopical findings showed amorphous eosinophilic pale hyaline deposits, foci of calcification, extensive foci of ossification near the hyaline area (Fig. 2 and 3) and apple green birefringence in Congo-red staining with polarized light in subepidermal area of eyelid extending to subconjunctival stroma (Fig. 4).

We obtained informed consent to publish details of the patient's history and his picture.

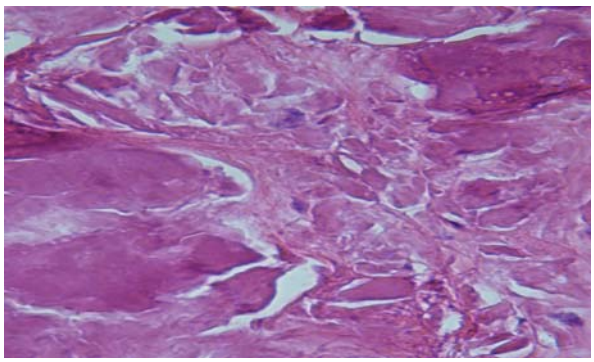

Fig. 2. Hyaline deposits with foci of calcification. H & E stain × 400.

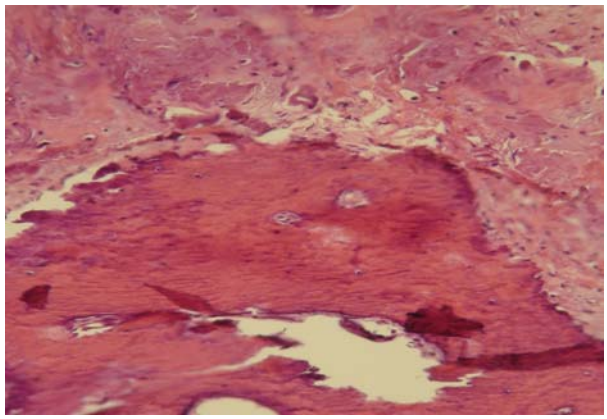

Fig. 3. Hyaline deposits with foci of ossification. H & E stain × 400.

DISCUSSION

Primary localized amyloidosis of eyelid manifest as unilateral or bilateral solitary or multiple firm, rubbery, waxy appearing painless fusiform or polypoid elevations (1, 5-7). It can be found in young or middle-aged, and occasionally, elderly persons. It develops slowly and in some cases the conjunctiva is diffusely thickened and slightly hyperemic (1, 11-14). Ptosis occurs when the deposits involve the upper tarsal conjunctiva or the levator muscles (1, 7-9).

Histopathological findings in our case were compatible with amyloidosis. It seems that ossification and calcification were significant due to chronicity. Systemic examination and all laboratory tests were normal which excludes systemic amyloidosis. Also, the only localized symptom or sign was ptosis due to upper lid mass attached to tarsus, making the diagnosis of secondary amyloidosis unlikely.

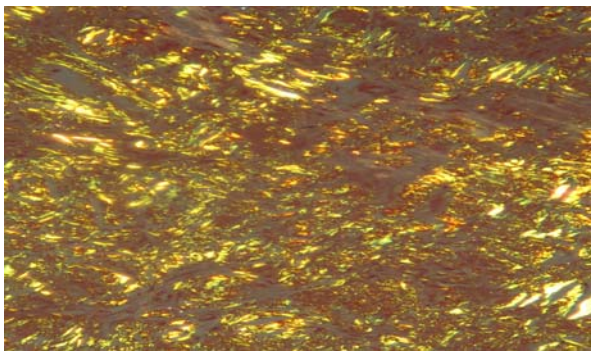

Fig. 4. Amyloid material with apple green birefringence in Congo-red staining.

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