

Amyloidosis of the Upper Eyelid

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Introduction

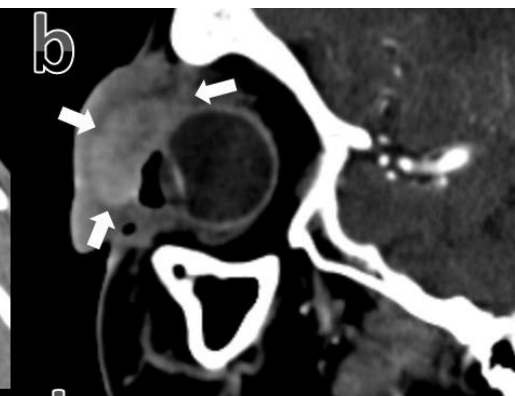
- Amyloidosis is a heterogeneous group of diseases characterized by extracellular amyloid deposits in different organs
 - Amyloid is a pathologic proteinaceous substance, deposited between cells in various tissues and organs of the body in a wide variety of clinical setting. The gold standard of amyloid detection is the demonstration of apple-green birefringence on congo red staining
 - The clinical presentation of amyloidosis can be classified by the clinicopathological features, the disease locations and magnitude of amyloid deposits¹
 - Amyloidosis may be systemic (generalized), involving several organ systems, or it may be localized, when deposits are limited to a single organ
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- 1. Rootman J. Degenerations and depositions. In: Rootman J, editor. Diseases of the Orbit: A Multidisciplinary Approach, 2nd ed. Philadelphia, PA: Lippincott Williams & Wilkins; 2003:555–564.

Introduction

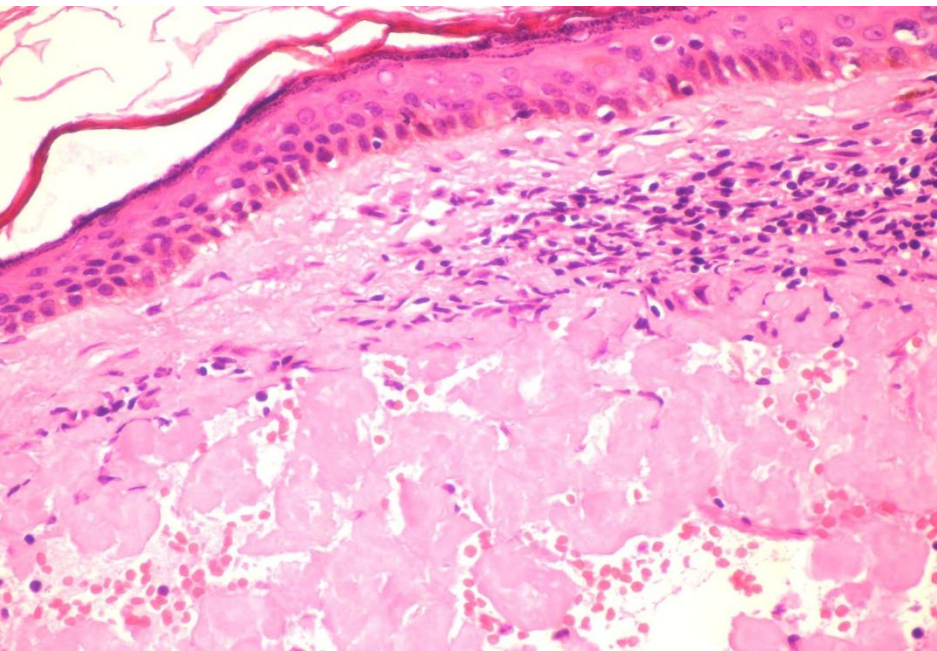
- On clinical grounds, the systemic - or generalized - pattern is sub classified into two groups, primary amyloidosis which is associated with immunocyte dyscrasia and secondary amyloidosis, a common complication of an underlying chronic inflammatory or tissue destructive process
- Sites of periocular and orbital amyloid deposit- are the lacrimal gland, eyelid, conjunctiva, and ocular adnexa, and these are generally associated with primary localized diseases; however, all patients should be investigated to rule out systemic involvement. Localized amyloidosis has no effect on survival²
- Because periocular and orbital amyloidosis is rare, the definitive diagnosis can be delayed, which could lead to disease progression
- 2. Naxer S, Behnes CL, Schittkowski MP. Amyloidosis – a rare differential diagnosis of an orbital tumour. Klin Monbl Augenheilkd. 2011;228(6): 555–564. German.

Case

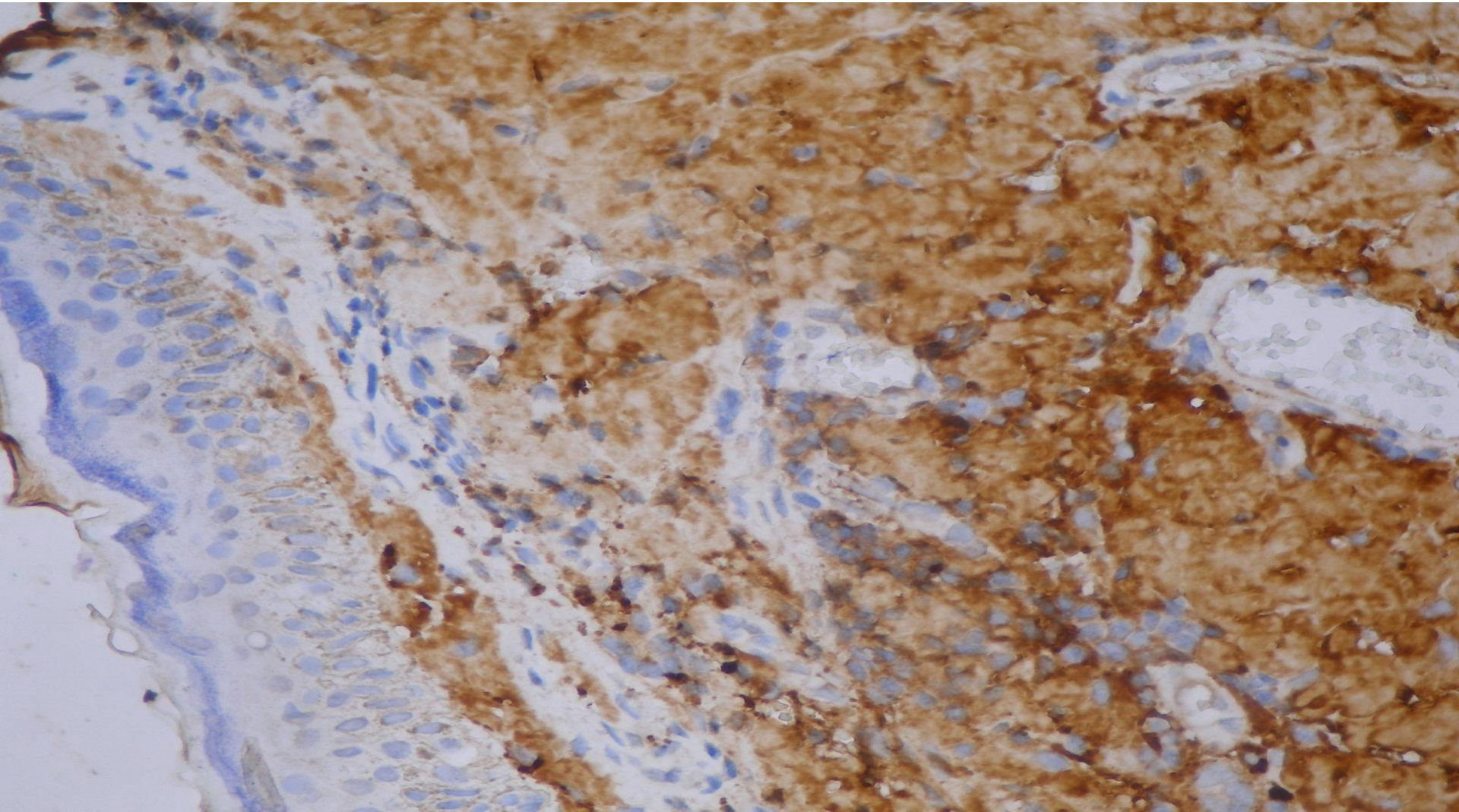
- A 67 year old woman was admitted for firm, painless, nodular, slowly growing unilateral lesion of right upper eyelid
- The lesion had been observed more than 8 years
- She hasn't been done any ocular examination before. Her medical history was normal
- On admission visual acuity was 0,05/0,6 (VOS = 0.05/0,6) and intraocular tension was within normal limits (TOS = 18/15 mmHg)
- Slit lamp biomicroscopy and fundus examination couldn't have done completely because of the lesions mass effect
- In the right eye we found limited ocular motility in both adduction and abduction but the eye showed orthotropia in the primary position
- The patient had routine laboratory tests, Computed tomography (CT) imaging demonstrated right eyelid swelling and an ill-defined isodense upper eyelid lesion
- The lesion showed diffusely enhancement after contrast media administration
- The patient underwent partial excision of the tumour-like lesions



- On MRI, there was prominent soft tissue haziness around the mass and diffuse prominent enhancement after contrast media administration
- The lacrimal gland and orbicularis oris muscle was infiltrated
- Evaluation by light microscopy revealed amorphous superepithelial amorphous eosinophilic deposits with Hematoxylin and eosin stained sections and amyloidosis was confirmed by a positive Congo red staining revealing a positive green birefringence when using with polarized light



- An immunohistochemical examination revealed that marked λ - chain-positivity was present in dermis



Discussion

- The clinical presentation depends on the location of the disease
- The diagnosis of eyelid primary amyloidosis is based on symptoms, patient's anamnesis, clinical examination and finally a biopsy specimen^{3,4}
- Radiological examinations, tomography or ultrasound are useful for the correct therapeutical approach and to exclude a possible systemic- involvement of the disease
- The demonstration of amyloid in and of itself is not particularly helpful to the clinician, and an accurate determination of the type of amyloid is just as important as recognizing the presence of amyloid
- The diagnosis of AL amyloidosis requires demonstration of amyloid in tissue and demonstration of a plasma cell dyscrasia. In approximately 10% of cases the disorder overlap with overt multiple myeloma

- 3. Ramsey MS, Fine BS and Cohen SW: Localized corneal amyloidosis: case report with electron microscopic observations. Am J Ophthalmol 73(4): 560-565, 1972.
- 4. Bozkurt B, Kiratli H, Soylemezoglu F and Irkec M: In vivo confocal microscopy in a patient with conjunctival amyloidosis. Clin Experiment Ophthalmol 36(2): 173-175, 2008.

Discussion

- The medical treatment of symptomatic primary eyelid amyloidosis is based on the use of ascorbic acid to reduce the bleeding tendency of these lesions and anti- inflammatory drugs, locally or general
- The surgical approach is based on the resection of the affected structures, with direct closure of the areas of loss in the initial localized forms, or with more complex and different reconstructive procedures in cases of greater extent of lesions
- Other therapeutic options are electro- cauterization, cryotherapy, and selective debridement

Conclusion

- Primary eyelid amyloidosis is a rare clinical entity that often leads to misdiagnosis
- Macroscopically differential diagnosis includes basal cell or squamous cell carcinoma, lachrymal gland carcinoma or lymphoma
- Ultrasonography and tomography are usually able to suggest the correct diagnosis but they are also mandatory to establish the relationships of the masses with the surrounding structures before surgery
- Moreover, general clinical examination and ophthalmologic tests are also indicated to complete the preoperative evaluation which should lead to a correct therapeutic strategy, avoiding partial or multiple treatments
- In fact, a surgical approach alone may often be associated with a high risk of incomplete excision and local recurrence, in the early or late postoperative period

Conclusion

- Radical surgical excision, even in highly specialized oculoplastic centers, may cause severe impairment of both structure and function of the eyelid
- For these reasons, other methods such as electro cauterization, cryotherapy, selective curettage and radiotherapy have already been described in addition to surgery as useful tools to remove amyloid vegetations