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Case Report

Lower Eyelid Conjunctival Amyloidosis: A Case Report

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Abstract

Amyloidosis is characterized by extracellular deposition of protein amyloid fibrils in tissues, including conjunctiva, which are insoluble in proteolytic proteins. Conjunctival amyloidosis specifically in the palpebral conjunctiva is a rare finding in clinical practice [1,4,5,7,10]. The authors described the case of a 37-year-old Bangladeshi origin female patient, with lesion located in the lower eyelid conjunctiva, elevated, well-defined, yellowish. Incisional biopsy of the lesion was performed, and the histological examination confirmed conjunctival amyloidosis. Conjunctival amyloidosis is a rare finding in the literature. The management of conjunctival amyloidosis depends on the extent of local involvement and patient's general health status, varying from symptomatic, with use of ocular lubricants, to surgical, by removing the lesions.

Keywords: Conjunctival Amyloidosis; Eyelid; Amyloid Fibrils

Introduction

Conjunctival amyloidosis is an uncommon condition that occasionally is associated with systemic involvement. Is classified by extent, it may be localised to one organ or may be systemic. It can also be classified according by amyloid protein type, with AL and AA subtypes being the most common [4,5,7,10].

Amyloidosis is caused by the extracellular tissue deposition of amyloid fibrillar proteins. Amyloid fibrils are insoluble polymers comprised of low molecular weight subunit proteins, which adopt a beta-pleated sheet configuration. The deposition of these insoluble proteins can lead to tissue toxicity and disease. At least 27 different human protein precursors of amyloid fibrils are now known. Some types of amyloidosis are hereditary and have been linked to genetic mutations. For others there is no hereditary component [4].

The clinical presentations of conjunctival amyloidosis are diverse. Usually appears as confluent fusiform lesions or polypoidal papules that have a salmon-coloured or yellow-pink colour. Any conjunctival surface may be involved, but the superior fornix and tarsal conjuctiva are the most frequently affected. This deposition may result in ptosis [2]. Patients may also present with recurrent subconjunctival haemorrhage [11,12] from the accumulation of amyloid in the walls of the blood vessels. The eyelid, extraocular muscles and lacrimal gland may also be affected by amyloidosis.

Secondary amyloidosis is associated with a variety of chronic inflammatory diseases such as rheumatoid arthritis, ankylosing spondylitis, familial Mediterranean fever, osteomyelitis, inflammatory bowel disease and infective or neoplastic conditions. Few cases of secondary amyloidosis complicating psoriasis have been reported.

We present one patient with conjunctival amyloidosis who was referred to us with the suspicion of another conjunctival lesion.

Case Report

A 37-year-old female of Bangladeshi origin, referred to the oculoplastic clinic from local optician with the suspicion of conjunctival lesion. The right lower lid superior fornix and tarsal conjuctiva were affected, without any sign of orbital involvement. The lesion's colour was yellow, pink, the associated features were intrinsic vascularization. (Figures 1,2,3) Systemic evaluation revealed no related systemic abnormalities. Incisional biopsy performed was suggestive of AL (lambda sub-type) amyloid. Further investigation regarding systemic amyloidosis and neoplastic plasma cells disease have been performed which have shown negative results. Blood test results show increased levels of Serum IgA 4.85, Ser Kappa light ch23.4. The patient has been referred to the National Amyloidosis Centre (UCL Hospital, London) and diagnosed with Amyloidosis Type A1 localised. The patient remains stable over the last 9 months under conservative treatment.

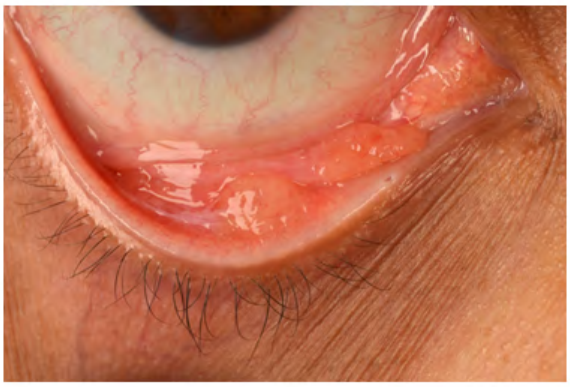

Figure 1

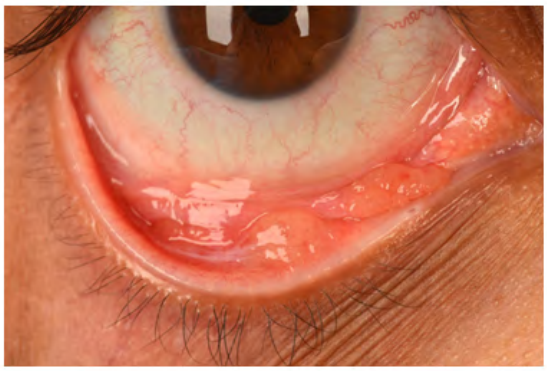

Figure 2

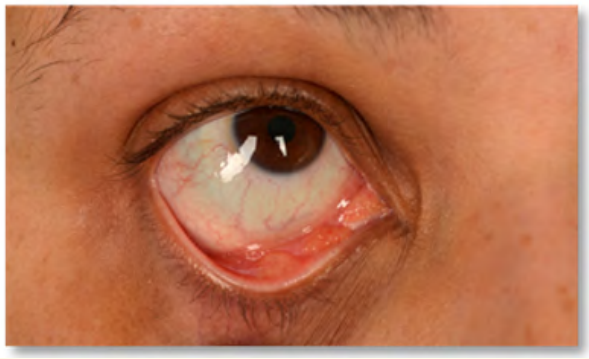

Figure 3

The family history was no contributory although the patient has suffered from chronic plaque psoriasis for the last 10 years, received and completed phototherapy on October 2020 with good response and is on treatment with Dovobet, Exorex and Emollients.

Secondary amyloidosis is associated with a variety of chronic inflammatory diseases such as rheumatoid arthritis, ankylosing spondylitis, familial Mediterranean fever, osteomyelitis, inflammatory bowel disease and infective or neoplastic conditions. Few cases of secondary amyloidosis complicating psoriasis have been reported.

Recommended treatment

This case has been managed so far conservatively using artificial tears.

Management modalities include observation, judicious use of artificial tears, excision, liquid nitrogen cryotherapy or low dose external beam therapy for localized conjunctival amyloidosis [6,8,9]. For patients with systemic manifestations, stem cell transplant, chemotherapy and steroids may be considered depending on the extend of the disease. Surgical debulking remains the standard treatment [3], but because complete excision isn't always possible, the disease may occur. Demirci and Leibovitch report a recurrence rate of 21-27% after surgical debulking of localized orbital amyloidosis. Newer therapies such as cryotherapy may further decrease recurrence rate by decreasing blood supply to the surrounding tissue, but more research is needed. Radiotherapy for localized amyloidosis has also been reported but is not considered the standard of care. For patients with recurrent amyloidosis, treatment varies depending on the disease manifestations [13].

Conclusion

Although conjunctival amyloidosis is a rare clinical entity to come across in the field of ophthalmology, it holds a clinical significance due to the list of differential diagnosis. The presenting symptoms may confuse the clinician. The presence of additional symptoms and the decision to a diagnostic biopsy may confound the clinical diagnosis. However, thorough ophthalmology and dermatology evaluation and the knowledge of such a rare condition can help to narrow down the differentials. The clinical differential diagnosis of conjunctival amyloidosis includes lymphoma, leukaemia, squamous cell carcinoma, sebaceous cell carcinoma, sarcoidosis and melanoma. The smooth salmon coloured appearance of amyloidosis is similar to lymphoma. This case emphasizes the atypical presentation of conjunctival amyloidosis as unilateral ocular mass progressing gradually, which can mislead the actual diagnosis. It also signifies the importance of history taking and clinical examination as the diagnosis of conjunctival amyloidosis is mostly a clinical diagnosis of exclusion of other possible differentials taking under consideration the patient's medical history. The diagnosis of conjunctival amyloidosis can only made with tissue biopsy and confirmed histopathology.

Systemic involvement should be ruled out in any patient who presents with an isolated conjunctival lesion. Amyloidosis can frequently affect the kidney, heart and liver. Test of cardiac, liver and kidney function including echocardiogram and electrocardiogram as well as serum and urine protein electrophoresis should be obtained. More specific immunofixation and free light chain electrophoresis of both the serum and urine should be obtained as well. Other tests such as bone marrow biopsy, abdominal fat biopsy and rectal biopsy should be considered. Imaging tests as CT or MRI may be helpful in localizing and detecting the extent of orbital disease.

In conclusion, conjunctival amyloidosis generally manifests as a yellowish-pink, haemorrhagic mass deep to the epithelium. Most patients show no evidence of systemic amyloidosis. Is managed well with conservative treatment. Finally, amyloidosis is a disease derived from the accumulation o various insoluble amorphous fibrils in different tissues that may be idiopathic (primary amyloidosis) or associated with other inflammatory conditions (secondary amyloidosis). Among the others, dermatologic diseases have been described with this condition, including psoriasis. Secondary amyloidosis occurs in patients with chronic infectious or inflammatory processes. Limited cases of amyloidosis and coexisting psoriasis have been reported.

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