Primary Conjunctival Amyloidosis Mimicking Conjunctival Neoplasm: A Case Report

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Abstract

Amyloidosis is a group of disorders characterized by the extra cellular deposition of a substance called amyloid in various tissues. When present as a conjunctival mass with recurrent haemorrhage the condition mimicks a neoplastic lesion. We report such a patient who had complaints of a conjunctival mass with recurrent subconjunctival haemorrhage. The diagnosis of amyloidosis was made by excisional biopsy and confirmed by Congo red staining. Further work up did not reveal any evidence of systemic amyloidosis.

Key words : Conjunctival amyloidosis; Congo red; Neoplastic leison.

Introduction

Primary conjunctival amyloidosis is extremely rare entity with a characteristic feature of tendency to bleed. It is a chronic disease commonly involving tarsal and forniceal conjunctiva. When presenting as a conjunctival mass it mimicks a neoplastic lesion like lymphoma or papilloma [1]. The disease causes significant ocular discomfort and may also be a rare cause of ptosis. We report a case of conjunctival amyloidosis clinically mimicking a neoplastic lesion. The diagnosis was confirmed by biopsy of the lesion and histopathological examination (HPE).

Case Report

A 60 years old female came to the Department of Ophthalmology with recurrent bleeding and chronic discomfort in the left eye for the last one year. She had noticed a slowly growing conjunctival mass in the left eye for

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last 6 months. The patient had no preceding history of any ophthalmic complaints or systemic symptoms. There was no history of eye surgery or ocular trauma. Ophthalmic examination showed a circumscribed reddish mass in the left lower palpebral conjunctiva. Rest of the ocular and systemic examination was normal. The mass was excised and sent for HPE to rule out any neoplastic pathology. Grossly the excised mass measured 0.8 x0.5x0.2cm with a reddish pink colour. HPE revealed a polypoidal lesion lined by conjunctival epithelium with sub epithelial tissue showing deposits of amorphous, eosinophilic, pale hyaline material, with interspersed ectatic and congested blood vessels (figure 1). Areas of hemorrhage were also seen. Congo red staining revealed the characteristic salmon pink colour of the deposits (figure 2) which on polarized microscopy exhibited a characteristic applegreen birefringence suggestive of amyloid deposition. A diagnosis of amyloidosis was established. A further detailed work up revealed no evidence of systemic amyloidosis. Serum electrophoresis was normal with absence of monoclonal band.

Discussion

Primary conjunctival amyloidosis is a rare disease which may mimic an allergic or neoplastic etiology and is apparently the most

Figure 1: Biopsy section. Microphotograph showing subconjunctival deposits of pale, eosinophilic acellular material distinctive of amyloid. (H& E stain,X 200)



Figure 2: Congo red staining of biopsy section showing typical red-orange color of amyloid deposits. (Congo-red stain, X100)



common non familial ophthalmological manifestation of amyloidosis. It results from extra cellular deposition of insoluble fibrous amyloid proteins in the organs and tissues [2]. It may occur as an uncommon complication of trachoma[3] or may be unusual cause of ptosis. The term amyloidosis was coined by Virchow in 1854. The first case of localized ocular amyloidosis was reported in 1871. The disease is usually unilateral as seen in our patient though bilateral involvement is not uncommon [2]. The characteristic feature of the disease is recurrent tendency to bleed[4]. The early diagnosis is difficult as yellowish deposits indicative of amyloidosis are not obvious at this stage[5]. The patients may also present clinically with lid swelling, ptosis, chronic ocular discomfort, irritation, foreign body sensation, papillary hyperplasia and recurrent conjunctival haemorrhage[1]. The classic "salmon-pink" conjunctival infiltrate has been associated with lymphoproliferative disorders; however amyloid should also be considered as it may be clinically indistinguishable from such disorders [6]. When presents as a conjunctival mass as in our case the differential diagnosis include lymphoma, papilloma and allergic conditions [1]. HPE supplemented by special stains is necessary to determine the final diagnosis. The possibility of amyloidosis in extra ocular sites must also be ruled out. Our case did not have any evidence of amyloidosis in a follow up period of two years.

Conclusion

Most of the patients with conjunctival amyloidosis present with yellowish pink haemorrhagic mass in the conjunctiva and such patients usually do not show any evidence of systemic amyloidosis. Hence conjunctival amyloidosis should be kept in the differential diagnosis of any patient presenting with a conjunctival mass and recurrent subconjunctival haemorrhage. Biopsy and HPE are infallible tools for diagnosing such a condition.

References

- Vemuganti GK. Inflammatory and non neoplastic lesions of eyelids, eye ball and Orbit. J Cytol. 2007; 24: 26-30.
- Rodrigues G, Sanghvi V, Lala M. A rare cause of unilateral upper and lower eyelid swelling: Isolated conjunctival amyloidosis. *Korean J Ophthalmol.* 2001; 5: 38-40.

- Pecora JL, Sambursky JS, Vargha Z. Radiation therapy in amyloidosis of the eyelid and conjunctiva: A case report. *Ann Ophthalmol.* 1982; 14: 194-196.
- 4. Lee HM, Naor J, DeAngelis D, Rootman DS. Primary localized conjunctival amyloidosis presenting with recurrence of subconjunctival hemorrhage. *Am J Ophthalmol.* 2000; 129(2): 245-7.
 - 5. Mesa-Gutierrez JC, Huguet TM, Garcia NB, Ginebreda JA. Primary localized conjunctival amyloidosis: a case report with a ten-year followup period. *Clinical Ophthalmol.* 2008; 2(3): 685-687.
- Spitellie PH, Jordan DR, Gooi P, Brownstein S, Burns BF. Primary localized conjunctival amyloidosis simulating a lymphoproliferative disorder. *Ophthal Plast Reconstr Surg.* 2008; 24(5): 417-9.
- Rosenblatt M, Spitz GF, Friedman AH, Kazam ES. Localized Conjunctival Amyloidosis: Case Reports and Review of Literature. *Ophthalmologica*. 1986; 192: 238-245. (DOI: 10.1159/000309653).