E-ISSN: 3050-9459
Journal of Ophthalmology and Advance Research

ATHENAEUM SCIENTIFIC PUBLISHERS

Case Report

Recalcitrant Primary Conjunctival Amyloidosis

Aashka Damani[1], George A Villatoro[1], Debra J Shetlar[1], Michael T Yen[1*]

[1]Cullen Eye Institute, Department of Ophthalmology, Baylor College of Medicine, Houston, TX, United States

***Correspondence author:** Michael T Yen, MD, Cullen Eye Institute, Department of Ophthalmology, Baylor College of Medicine, Houston, TX, United States; Email: myen@bcm.edu

Citation: Damani A, et al. Recalcitrant Primary Conjunctival Amyloidosis. J Ophthalmol Adv Res. 2026;7(3):1-5.

https://doi.org/10.46889/JOAR.2026.7302

Received Date: 28-07-2026

Accepted Date: 26-08-2026

Published Date: 03-09-2026

Abstract

Primary conjunctival amyloidosis is a localized deposition of amyloid in the conjunctiva with variable clinical appearance that can cause pain, proptosis or limited field of view. Classically, this condition is not associated with systemic amyloidosis. However, patients must undergo incisional or excisional biopsy to confirm the histopathologic diagnosis and a systemic work up to rule out systemic amyloidosis or alternative etiologies amyloidosis. Patients can present with a variety of clinical phenotypes including eyelid thickening, blepharoptosis, subconjunctival papules, subconjunctival hemorrhages and eyelid ecchymoses which can mimic a number of other diagnoses including sarcoidosis, lymphoma, pyogenic granulomas, papilloma, nevus and sebaceous carcinoma. Depending on the clinical severity of presentation treatments may range from supportive therapy with ocular lubrication to gold standard treatment, surgical excision to debulk However, this treatment is not definitive as this can lead to recurrence in 21-50% of cases and thus poses a clinical challenge for ophthalmologists. Here we present a case of localized, primary conjunctival amyloidosis with rapid recurrence, underscoring the recalcitrant nature of this condition.

Keywords: Conjunctival Amyloidosis; Sarcoidosis; Lymphoma; Subconjunctival Papules; Pyogenic Granulomas

Introduction

Conjunctival amyloidosis is an uncommon condition that can mimic other diagnosis such as subconjunctival hemorrhages, sarcoidosis, pyogenic granulomas and lymphoid lesions [1-11]. It is a localized deposition of beta-folded proteins that cause tissue toxicity and inflammation, leading to lesions with mass effect that can cause pain, proptosis and limited visual fields of view [12]. A tissue biopsy usually confirms the diagnosis, though mass spectrometry protein sequencing can provide additional therapeutic guidance by confirming amyloid subtype [13].

It is typically localized but can rarely be associated with systemic involvement [10]. Conjunctival amyloidosis can present with eyelid thickening, blepharoptosis, subconjunctival orange-yellow papules, subconjunctival hemorrhages and spontaneous eyelid ecchymosis [5,7]. Amyloid lesions can involve the bulbar and palpebral conjunctiva separately or conjointly, with variable reporting in the literature on which is more prevalent [9,14]. While cryotherapy or low-dose radiation have emerged as possible treatment options, surgical excision remains the gold standard [6,7,15,16]. However, complete excision is not always possible and recurrence occurs in about 21-50% of patients [5,8,9].

Case Presentation

A 35-year-old female presented for evaluation after noticing several growths on the undersurface of the upper eyelids over the

last year. On exam, she had protruding palpebral conjunctival masses that appeared fleshy and almost salmon patch in appearance. They were present in the bilateral lower eyelid fornices as well but were not as protrusive. A biopsy of the conjunctival masses was performed which revealed acellular material staining strongly with congo red and "apple-green" birefringence under polarized light (Fig. 1), consistent with amyloid deposits. Comprehensive systemic evaluation, including serum and urine protein electrophoresis, revealed no abnormalities, suggesting localized conjunctival amyloidosis. The decision was made to observe the lesions.

Over the course of two years, the lesions on the right upper eyelid began to prolapse from the superior conjunctival fornix and palpebral conjunctiva onto the ocular surface and visual axis (Fig. 1). The weight of the lesions caused a mechanical ptosis of the upper eyelid. The patient elected to have the lesions excised. An exam under anesthesia revealed large confluent reddish papules emanating from the palpebral conjunctival surface and extending into the superior conjunctival fornix. The lesions were excised flush with the surrounding palpebral and fornix conjunctiva. The conjunctival defect of the upper eyelid tarsus and superior fornix was then reconstructed with amniotic membrane grafts.

Follow Up

She was doing well one month post-operatively without evidence of significant recurrence (Fig. 1). At her three month follow up, the conjunctival amyloid lesions recurred (Fig. 1).

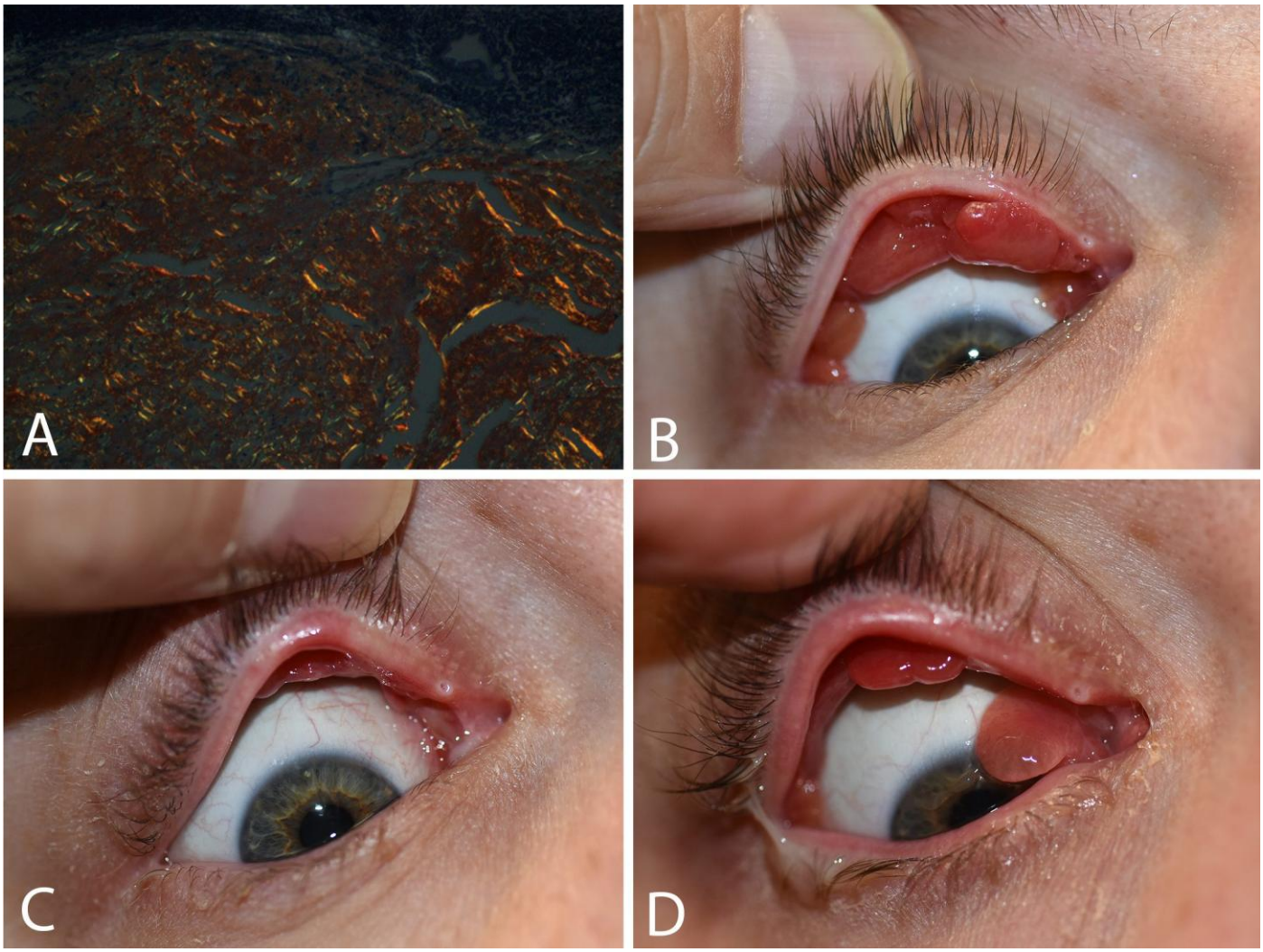

Figure 1: Clinical and histopathological features of conjunctival amyloidosis. (A) Histopathological examination demonstrating amyloid deposits with characteristic birefringence under polarized light. (B-D) Clinical photographs showing localized, nodular, salmon-pink conjunctival lesions involving the upper palpebral and bulbar conjunctiva.

Discussion

Primary conjunctival amyloidosis is an extracellular, localized deposition of amyloid fibrillar proteins that typically involves the palpebral conjunctiva, especially the superior fornix and tarsal conjunctiva, though it can occur anywhere in the conjunctiva. Amyloidosis is a disease of protein folding in which normally soluble proteins deposit as abnormal, insoluble fibrils that disrupt tissue structures and cause disease [17].

Though primary conjunctival amyloidosis is not classically associated with systemic amyloidosis, patients presenting with biopsy-confirmed conjunctival amyloidosis should undergo systemic work up [1-3,5]. Amyloid fibrils can deposit anywhere in the body and systemic amyloidosis classically causes complications in the heart, lung, liver and kidney. Thus, patients must undergo a thorough workup including echocardiogram, electrocardiogram and serum and urine protein electrophoresis and immunofixation [18]. In patients who need further workup, bone marrow biopsy almost always demonstrates a population of clonal cells. Even skin biopsies will demonstrate amyloid deposition and now rectal biopsies have fallen out in favor for noninvasive abdominal fat aspiration as they are less invasive and have equivalent success at identifying amyloid [12,19,20]. CT, MRI or radionuclide imaging may play a role in identifying amyloid-associated inflammation in other parts of the body as well [12].

Historically, amyloidosis was categorized anatomically with three categories primary (AL), secondary (AA) and hereditary. Now amyloidosis is characterized based on the composition of the amyloid subunit protein [12,21-23]. Thus far, more than 20 subtypes of amyloid have been identified [24]. Subtyping amyloid is critical to determining the therapeutic options that are most effective for a patient's disease.

Patients with systemic amyloidosis are treated with stem cell transplant, chemotherapy regimens and steroids or transplantation [25-28]. Localized conjunctival amyloidosis can be treated with artificial tears, excision, observation or radiotherapy [6,7,15,16]. Radiotherapy has been reported, but success has been variable and it is not currently the standard of care [16,29,30]. Gold standard therapy is surgical excision with recurrence in 21-50% of cases [5,8,9]. Cryotherapy is emerging as a newer option, decreasing blood supply to the vascular lesions [15].

This case underscores the recalcitrant nature of this condition with surgical debulking as the most common repeat procedure these patients undergo [14]. It's unclear why and which patients have recurrence, but chronic inflammation, trauma, surgery or amyloid subtype may play a role [13,31-33]. Follow-up in the literature ranges from 6 mos to 36 years, with this case being the most rapid to recur in 3 months [9,14]. There is one other reported case of tumor recurrence at 1 month but initial histopathology demonstrated only granulation tissue with subsequent biopsy 1.5 years later demonstrating amyloidosis, thus it is unclear whether the patient initially had primary conjunctival amyloidosis [33].

This patient is prototypical for conjunctival amyloidosis - female in her 4th decade of life with 2 years to diagnosis given her early nonspecific symptoms [9,14]. Contradictorily, systemic amyloidosis occurs in predominantly older males [34]. The nonspecific nature of conjunctival amyloid symptoms causes a significant delay in diagnosis, with many patients receiving correctional ptosis surgeries, sometimes repeatedly before receiving a confirmatory pathologic diagnosis [14,33]. Thus, it's critical to have a high index of suspicion and complete a thorough workup for ptosis etiologies before proceeding with interventional surgery. In fact, post-surgical inflammation may serve to exacerbate the underlying condition possibly contributing to recurrence [33].

Conclusion

In conclusion, conjunctival amyloidosis is a recalcitrant and sometimes rapidly recurrent disease that is treated with surgical excision and often requires repeat debulking over long follow up periods to maintain patient comfort and vision. This case demonstrates the most rapid recurrence of conjunctival amyloidosis in the literature to date.

Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Funding Statement
This research did not receive any specific grant from funding agencies in the public, commercial or non-profit sectors.

Acknowledgement
The authors have no acknowledgments to declare.

Data Availability Statement
The data supporting the findings of this study are available from the corresponding author upon reasonable request.

Ethical Statement
The project did not meet the definition of human subject research under the preview of the IRB according to federal regulations and therefore was exempt.

Informed Consent Statement
Informed consent was obtained from all participants included in the study.

Authors' Contributions
All authors contributed equally to this paper.

References

1. Purcell JJ, Birkenkamp R, Tsai CC, Riner RN. Conjunctival involvement in primary systemic nonfamilial amyloidosis. Am J Ophthalmol. 1983;95(6):845-7.
2. Iijima S. Primary systemic amyloidosis: an unique case complaining of diffuse eyelid swelling and conjunctival involvement. J Dermatol. 1992;19(2):113-8.
3. Shields JA, Eagle RC, Shields CL, Green M, Singh AD. Systemic amyloidosis presenting as a mass of the conjunctival semilunar fold. Am J Ophthalmol. 2000;130(4):523-5.
4. Shields JA, Shields CL. Atlas of Eyelid and Conjunctival Tumors. Lippincott Williams & Wilkins. 324-25.
5. Demirci H, Shields CL, Eagle RC, Shields JA. Conjunctival amyloidosis: report of six cases and review of the literature. Surv Ophthalmol. 2006;51(4):419-33.
6. O'Donnell B, Wuebbolt G, Collin R. Amyloidosis of the conjunctiva. Aust N Z J Ophthalmol. 1995;23(3):207-12.
7. Patrinely JR, Koch DD. Surgical management of advanced ocular adnexal amyloidosis. Arch Ophthalmol. 1992;110(6):882-5.
8. Leibovitch I, Selva D, Goldberg RA, Sullivan TJ, Saeed P, Davis G, et al. Periocular and orbital amyloidosis: clinical characteristics, management and outcome. Ophthalmology. 2006;113(9):1657-64.
9. Banerjee P, Alam MS, Subramanian N, Kundu D, Koka K, Poonam NS, et al. Orbital and adnexal amyloidosis: thirty years experience at a tertiary eye care center. Indian J Ophthalmol. 2021;69(5):1161-6.
10. Abdallah AO, Westfall C, Brown H, Muzaffar J, Atrash S, Nair B. Unilateral conjunctival AL kappa amyloidosis with trace evidence of systemic amyloidosis. Am J Case Rep. 2012;13:102-5.
11. Grossniklaus HE, Green WR, Luckenbach M, Chan CC. Conjunctival lesions in adults: a clinical and histopathologic review. Cornea. 1987;6(2):78-116.
12. Gertz MA, Lacy MQ, Dispenzieri A. Amyloidosis. Hematol Oncol Clin North Am. 1999;13(6):121133.
13. Hamill EB, Thyparampil PJ, Yen MT. Localized immunoglobulin light chain amyloid of the conjunctiva confirmed by mass spectrometry without evidence of systemic disease. Ophthalmic Plast Reconstr Surg. 2017;33(5):e108-10.
14. Kang S, Dehabadi MH, Rose GE, Verity DH, Amin S, Das-Bhaumik R. Ocular amyloid: adnexal and systemic involvement. Orbit. 2020;39(1):13-7.
15. Fraunfelder FW. Liquid nitrogen cryotherapy for conjunctival amyloidosis. Arch Ophthalmol. 2009;127(5):645-8.
16. Copperman TS, Truong MT, Berk JL, Sobel RK. External beam radiation for localized periocular amyloidosis: a case series. Orbit. 2019;38(3):210-6.
17. Pepys MB. Pathogenesis, diagnosis and treatment of systemic amyloidosis. Philos Trans R Soc Lond B Biol Sci. 2001;356(1406):203-10.
18. Pascali E. Diagnosis and treatment of primary amyloidosis. Crit Rev Oncol Hematol. 1995;19(3):149-81.

19. Kyle RA, Spencer RJ, Dahlin DC. Value of rectal biopsy in the diagnosis of primary systemic amyloidosis. Am J Med Sci. 1966;251(5):501-6.
20. Huang CY, Wang WJ, Wong CK. Skin biopsy gives the potential benefit in the diagnosis of systemic amyloidosis associated with cardiovascular involvement. Arch Dermatol. 1998;134(5):643-5.
21. Gertz MA. Secondary amyloidosis (AA). J Intern Med. 1992;232(6):517-8.
22. Mahloudji M, Teasdall RD, Adamkiewicz JJ, Hartmann WH, Lambird PA, McKusick VA. The genetic amyloidoses with particular reference to hereditary neuropathic amyloidosis, type II (Indiana or Rukavina type). Medicine (Baltimore). 1969;48(1):1-37.
23. Block WD, Carey JG, Curtis AC, Falls HF, Jackson CE, Rukavina JG. Primary systemic amyloidosis: a review and an experimental, genetic and clinical study of 29 cases with particular emphasis on the familial form. Medicine (Baltimore). 1956;35(3):239-334.
24. Misumi Y, Ando Y. Classification of amyloidosis. Brain Nerve. 2014;66(7):731-7.
25. van Buren M, Hené RJ, Verdonck LF, Verzijlbergen FJ, Lokhorst HM. Clinical remission after syngeneic bone marrow transplantation in a patient with AL amyloidosis. Ann Intern Med. 1995;122(7):508-10.
26. Dhodapkar MV, Jagannath S, Vesole D, Munshi N, Naucke S, Tricot G, et al. Treatment of AL-amyloidosis with dexamethasone plus alpha interferon. Leuk Lymphoma. 1997;27(3-4):351-6.
27. Dubrey S, Simms RW, Skinner M, Falk RH. Recurrence of primary (AL) amyloidosis in a transplanted heart with four-year survival. Am J Cardiol. 1995;76(10):739-41.
28. Kyle RA, Gertz MA, Greipp PR, Witzig TE, Lust JA, Lacy MQ, et al. A trial of three regimens for primary amyloidosis: colchicine alone, melphalan and prednisone and melphalan, prednisone and colchicine. N Engl J Med. 1997;336(17):1202-7.
29. Bennassi A, Kinj R, Chaabouni S, Khanfir K. Orbital amyloidosis and radiotherapy: a case report and review of literature. Cancer Radiother. 2022;26(8):1070-4.
30. Pecora JL, Sambursky JS, Vargha Z. Radiation therapy in amyloidosis of the eyelid and conjunctiva: a case report. Ann Ophthalmol. 1982;14(2):194-6.
31. Knowles DM, Jakobiec FA, Rosen M, Howard G. Amyloidosis of the orbit and adnexae. Surv Ophthalmol. 1975;19(6):367-84.
32. Khalifa YM, Bloomer MM, Margolis TP. Secondary localized sectoral keratoconjunctival amyloidosis from ocular trauma. Cornea. 2010;29(11):1328-9.
33. Lee PAL, Vaidya A, Kakizaki H, Takahashi Y. Amyloidosis in the bulbar conjunctiva following transconjunctival ptosis surgery. J Craniofac Surg. 2021;32(3):e280-81.
34. Kyle RA, Greipp PR. Amyloidosis (AL): clinical and laboratory features in 229 cases. Mayo Clin Proc. 1983;58(10):665-83.

About the journal

ATHENAEUM SCIENTIFIC PUBLISHERS

Journal of Ophthalmology and Advance Research is a peer-reviewed, open-access scholarly journal published by Athenaeum Scientific Publishers. The journal publishes original research articles, case reports, reviews, editorials and commentaries within its defined scope, with the aim of supporting scientific research and clinical knowledge in ophthalmology.

All manuscripts are evaluated through an independent peer-review process conducted in accordance with the journal's editorial policies and established publication ethics. Editorial decisions are made solely on the basis of academic merit.

Manuscript submission: https://athenaeumpub.com/submit-manuscript/