CASE REPORT

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Amyloid in Conjunctiva

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ABSTRACT The protein structure known as amyloid exhibits apple-green birefringence when stained with congo-red. The condition known as amyloidosis is caused by the deposition of amyloid fibrils in extracellular tissue. Amyloidosis may be systemic or localized. Localized amyloidosis is limited to an organ such as heart, skin, and eye. Amyloid in conjunctiva is a rare case presentation. The use of radiotherapy is one of the many available treatment options. In our clinic, a resistant conjunctival amyloidosis case was determined. The patient's right eye had a yellow, waxy lesion. Radiotherapy was applied on the resistant case and it was followed for 6 months in our clinic. The yellow waxy lesion on the right eye had regressed.

Keywords: Amyloidosis; conjunctival disease; radiotherapy

Amyloid is distinguished by its apple-green birefringence with congo-red. The extracellular tissue deposition of these fibrils named amyloidosis.[1] According to, the type and the quantity of the amyloid deposition and clinical presentation of the disease varied.[2] Due to the progressive nature of this disease, the affected tissue may stop functioning.[2-4] According to the literature, amyloidosis could be divided into systemic and localized. In systemic amyloidosis, the fibrils in the circulation system could be deposited in multi organ such as the heart, kidney, and more. By contrast, localized amyloidosis is limited to an organ for instance heart, skin, and eye due to the local amyloid production of monoclonal B cells or plasma cells.[1,4-6] On the other hand, localized ocular amyloidosis is separated into 2 groups primary and secondary. Primary localized ocular amyloidosis has no predisposing factors. However, secondary localized ocular amyloidosis is the conclusion of underlying circumstances for instance infectious or inflammatory situations, trauma.[2,3,7] Localized amyloidosis is rarely observed, and 4% of cases occur in the orbit.[8]

We present a case report with conjunctival amyloidosis that is treated in our clinic.

CASE REPORT

An untraumatized 43-year-old female patient had a history of right eye redness and itching. After examination by the ophthalmologist, bilateral eye redness was detected. Both color vision and visual acuity were within normal ranges. Performed incisional biopsy proved that the lesion is conjunctival nodular amyloidosis. Histopathological analysis revealed the following: CD68(+) histiocyte infiltration, polytypic staining with kappa and lambda with plasmocytes, accumulation of Congophilic amorphous substance in the subepithelial region, and green birefringence with congo-red staining under polarized light. An

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magnetic resonance imaging (MRI) scan was performed to check for disease invasion. The MRI scan was unremarkable.

The patient was also examined for systemic amyloidosis, rheumatological disorders, and lymphoproliferative conditions like multiple myeloma. There was no indication that these illnesses existed. There was no evidence of amiloid deposition in the aspiration of abdominal fat. No abnormalities were detected in the kappa and lambda light chains, or in the protein electrophoresis. The 24-hour urine protein, β2-microglobulin and immunoglobulin levels, biochemistry, coagulation function, routine blood work, and urinalysis were all within normal ranges. The patient who was discussed in the multidisciplinary council was accepted as having bilateral primary localized amyloidosis because of that amyloidosis was not observed in the abdominal adipose tissue biopsy, urinary and blood parameters were within normal limits, the MRI showed involvement limited to the orbit, and no uptake was seen in the positron emission tomography scans. Amyloidosis was limited to the conjunctiva. The bilateral nodular red-yellow waxy lesions caused the cosmetically concerned patient to be excised locally. The right eye showed signs of progression about 2 months following the procedure. The lesion extending to the fornices caused thickening of the lower eyelid. Blepharoptosis and subconjunctival involvement were not observed.

The ophthalmologist did not advise the patient to have another excision after the first. Radiotherapy was administered at the patient's request due to cosmetic concerns despite being under hematology follow-up, the patient did not receive a new systemic evaluation. Radiation therapy (RT) was administered to the lesion on the right eye using the volumetric modulated arc therapy technique with a total dose of 20 Gray (Gy) in 10 fractions with 6 MV (Figure 1). The organ at risk was within normal limits (Figure 2). Computed tomography cone beam was used for field verification. During the 2nd week of treatment, the right eye experienced grade 1 watery eyes as an acute side effect, and beginning in the 3rd week, grade 1 dry eyes were observed. There have been reports of grade 1 dry eye as a persistent side effect. The 6-month follow-up showed no progression or recurrence (Figure 3). Color vision, optic coherence tomography, and visual acuity were all within normal ranges.

Patient information consent was obtained from the patient for case report of the photographic, medical and radiological data of the patient.

DISCUSSION

One of the limited types of amyloidosis is isolated ocular amyloidosis. According to a retrospective study, ophthalmic presentation is found in 11.8% of patients with systemic amyloidosis.[4] Additionally, Kang et al. pointed out that 25 % of orbital and ocular adnexal amyloidosis cases had associated systemic diseases or malignancies.[9] Although ocular amyloidosis is primarily localized, systemic diseases such as lymphoma and myeloma could be considered as a differential diagnosis in the presence of ocular amyloidosis.[1,2,4,10] In our case, both systemic amyloidosis and malignancies have been excluded.

Ocular amyloidosis is known to involve the eyelids, conjunctiva, ocular adnexa, and lacrimal

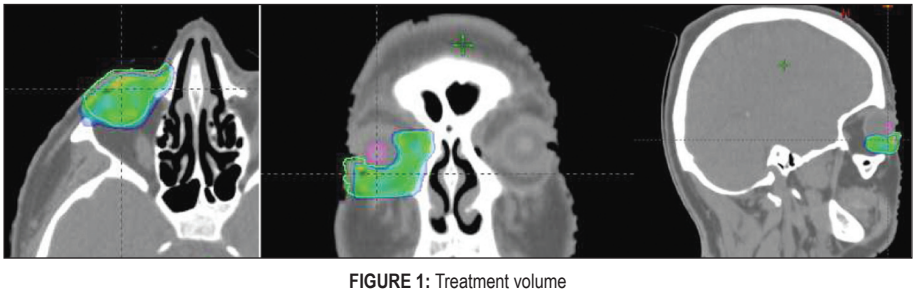

FIGURE 1: Treatment volume

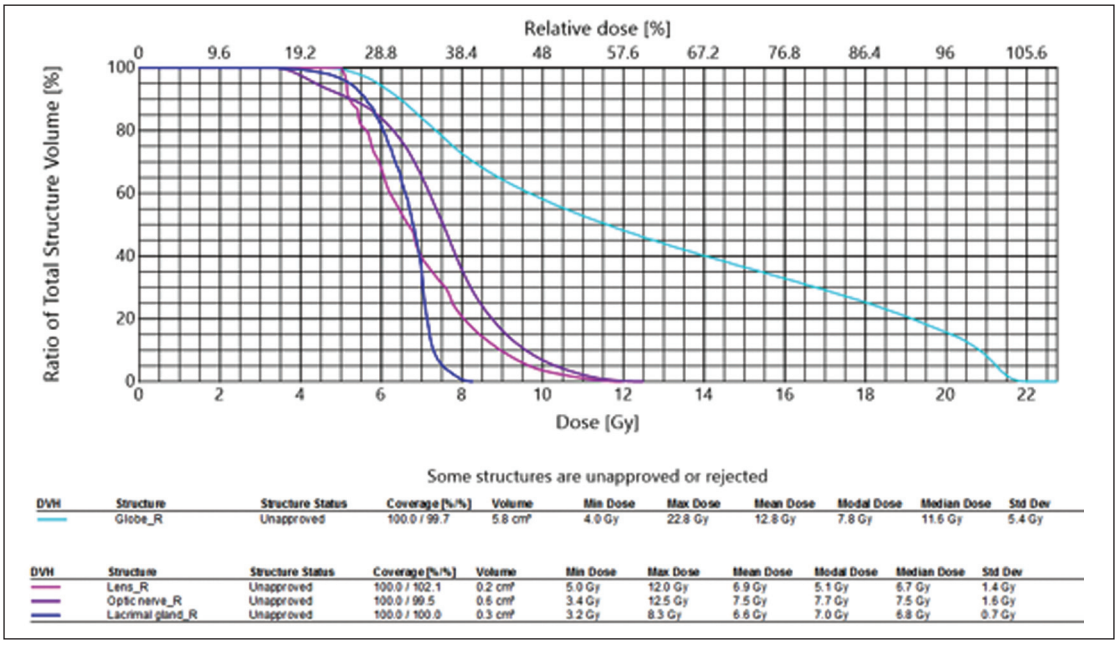

FIGURE 2: Organ at risk

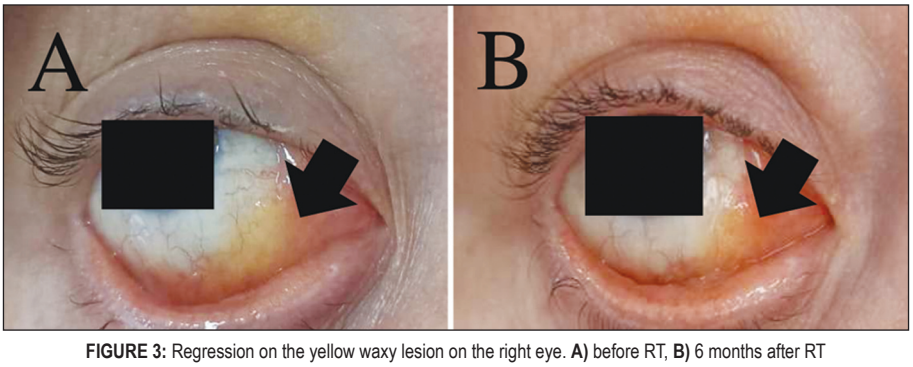

FIGURE 3: Regression on the yellow waxy lesion on the right eye. **A)** before RT, **B)** 6 months after RT

glands. Nevertheless, the most common presentation of orbital amyloidosis is conjunctival.[1,2] Conjunctival amyloidosis is difficult to diagnose because of the lack of suspicion and subtle symptoms.[3,10] The majority of these originate from fornices and pass through the *bulbus*.[2,10] Although the lesion in early stages could be an asymptomatic nodule that varies in colors such as salmon, red, or pale, the diagnostic demonstration of early-stage conjunctival amyloidosis is waxy, yellow, fragile painless deposits.[3,10] In our case, only yellow wax-like deposits were present. Ptosis, and global displacement may be presenting symptoms caused by involvement of the eyelid and other adnexal tissues.[2] A biopsy from the afflicted tissue could be used to diagnose the illness.[3,4]

Cryotherapy and CO_2 laser vaporization are considered treatment modalities. As conservative measures, steroids, and lubricants could be used. Especially, in cases with unremovable masses or possible recurrence, radiotherapy should be taken into consideration.[11,12] As a treatment for our case in which local excision was performed, we administered radiotherapy.

Localized eyelid and conjunctival amyloidosis can be treated surgically, according to a study involving 11 patients.[13] However, Kang et al. noted that observation of cases without systemic involvement and radiotherapy at unresectable phenomena could be alternative treatment options.[9] Additionally, radiotherapy was recommended either alone or prior to surgery in cases that could not be surgically removed, as well as postoperatively in cases with frequent and residual recurrences.[5,6,14]

Because benign focal clones of monoclonal B cells or plasma cells lead to primary local amyloidosis, low-grade lymphoma dose regiments are utilized for dose determination of the radiation therapy.[5,6,14] Additionally, the dose of the primary localized orbital amyloidosis was based on successful radiotherapy experiences of tracheobronchial and laryngeal amyloidosis.[6] The utilized radiation dose ranged from 20 to 39 Gy and there are orbital amyloidosis case reports indicating local control at 20-34 Gy at a follow-up ranging between 2-6 years.[5,6,14] Additionally, the literature documents radiation therapy with doses ranging from 18 to 30 Gy that show remarkable progress and no recurrence.[12] Furthermore, due to the studies using low-dose RT in lymphoproliferative diseases, it is predicted that the same effect can be observed even with doses below 20 Gy.[14] We provided local control with 20 Gy RT in our patient and their follow-up continues in our clinic. In conjunctival amyloidosis, a hematological disorder, options such as observation without treatment and cryotherapy are available. However, RT was administered at the patient's request due to cosmetic concerns.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Ömer Erol Uzel;* ***Design:*** *Meltem Dağdelen;* ***Control/Supervision:*** *Ömer Erol Uzel, Ahmet Murat Sarıcı;* ***Data Collection and/or Processing:*** *Hatice Kübra Topal;* ***Analysis and/or Interpretation:*** *Ömer Erol Uzel, Meltem Dağdelen;* ***Literature Review:*** *Hatice Kübra Topal;* ***Writing the Article:*** *Hatice Kübra Topal;* ***Critical Review:*** *Meltem Dağdelen, Ömer Erol Uzel.*

REFERENCES

1. Gorevi PD, Schur P, Romain P. Overview of amyloidosis. Up to date.2011;10(3). [Link]
2. Sultana S, Robi NH, Rashid R, Kadir, SMU. Amyloidosis of the orbit and adnexia. International Journal of Ocular Oncology and Oculoplasty. 2018;4(2):105-9. [Link]
3. Suesskind D, Ziemssen F, Rohrbach JM. Conjunctival amyloidosis — clinical and histopathologic features. Graefes Arch Clin Exp Ophthalmol. 2015;253(8):1377-83. [PubMed]
4. Lee PH, Liao IC, Lee WA. Rare presentations of primary amyloidosis as ptosis: a case report. BMC Ophthalmol. 2022;22(1):43. [Crossref] [PubMed] [PMC]
5. Khaira M, Mutamba A, Meligonis G, Rose GE, Plowman PN, O'Donnell H. The use of radiotherapy for the treatment of localized orbital amyloidosis. Orbit. 2008;27(6):432-7. [Crossref] [PubMed]
6. 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. [Crossref] [PubMed]
7. Ali Z, Fernando B. A rare case of amyloidosis of the eyelid and conjunctiva. Case Rep Ophthalmol Med. 2016;2016:2915196. [Crossref] [PubMed] [PMC]
8. Li J, Liu R, Ren T, Wang N, Guo Q, Xu L, et al. Clinical analysis of 37 Chinese patients with ocular amyloidosis: a single center study. BMC Ophthalmol. 2024;24(1):294. [Crossref] [PubMed] [PMC]
9. 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. [Crossref] [PubMed]
10. Moorman CM, McDonald B. Primary (localised non-familial) conjunctival amyloidosis: three case reports. Eye (Lond). 1997;11(Pt 5):603-6. [Crossref] [PubMed]
11. Mora-Horna ER, Rojas-Padilla R, López VG, Guzmán MJ, Ceriotto A, Salcedo G. Ocular adnexal and orbital amyloidosis: a case series and literature review. Int Ophthalmol. 2016;36(2):281-98. [Crossref] [PubMed]
12. Al-Nuaimi D, Bhatt PR, Steeples L, Irion L, Bonshek R, Leatherbarrow B. Amyloidosis of the orbit and adnexae. Orbit. 2012;31(5):287-98. [Crossref] [PubMed]
13. Cui Y, Li B, Chen T, Zhao Y, Li DM. [Clinical and pathological features and surgical treatment of eyelid/conjunctival amyloidosis]. Zhonghua Yan Ke Za Zhi. 2016;52(10):775-779. Chinese. [PubMed]
14. Truong MT, Kachnic LA, Grillone GA, Bohrs HK, Lee R, Sakai O, et al. Long-term results of conformal radiotherapy for progressive airway amyloidosis. Int J Radiat Oncol Biol Phys. 2012;83(2):734-9. [Crossref] [PubMed]