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Case report

Nd:YAG capsulotomy for the management of posterior capsular amyloidosis

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ABSTRACT

Purpose: To describe the accumulation of amyloid in Berger's space.
Observations: A 35-year-old man with autosomal-dominant, familial transthyretin-associated amyloidosis and bilateral vitreous opacities had a recurrence of amyloidosis following vitrectomy. The recurrent amyloid was attached to the posterior capsule of the lens. Phacoemulsification followed by neodymium:yttrium-aluminum-garnet (Nd:YAG) capsulotomy was helpful in restoring vision.
Conclusions and importance: Amyloid can reaccumulate in Berger's space, which is difficult to reach in phakic eyes during vitrectomy and can then cause decreased vision. Recognition of this interesting anatomic problem will allow for faster visual rehabilitation of the patient.

1. Introduction

Familial transthyretin (TTR)-associated amyloidosis is a rare autosomal-dominant disorder with variable phenotypic presentation due to extracellular deposition of amyloid fibrils composed of TTR.[1] Ocular involvement is common and can be in the form of abnormal conjunctival vessels, keratoconjunctivitis sicca, pupillary abnormalities, vitreous opacities and glaucoma.[2,3] We report the case of a 35-year-old man with autosomal-dominant, familial TTR-associated amyloidosis and bilateral vitreous opacities. After undergoing vitrectomy, he had a recurrence of ocular amyloid attached to the posterior capsule of the lens with complaints of glare and blurred vision. Vision was restored after phacoemulsification and neodymium:yttrium-aluminum-garnet (Nd:YAG) capsulotomy.

2. Case report

A 35-year-old man was referred to our clinic for the evaluation of bilateral vitreous floaters. He complained of a four-month history of decreased vision in the left eye. Family history was notable for amyloidosis in the father and the paternal grandmother. On examination, his visual acuity was 20/20 right (OD) and 20/150 left (OS). The anterior segment examination was normal. Fundus examination showed bilateral central, dense, strand-like opacities in the vitreous, worse in the left eye. Optical coherence tomography showed bilateral vitreous opacities with normal foveal contour and retinal architecture. A transthoracic echocardiogram was performed, which showed a thickened ventricular septum, suggestive of an infiltrative cardiomyopathy. An abdominal fat aspirate was positive for amyloidosis. Deoxyribonucleic acid (DNA) sequence analysis carried out on peripheral blood detected a mutation in exon 2 of the transthyretin (TTR) gene with a DNA change of c.157 T > A.

The patient underwent 25-gauge pars plana vitrectomy in the left eye, and the vision improved to 20/20 one month after surgery. Mass spectrometry of the vitreous sample confirmed the presence of TTR amyloid. One year later, the patient required 27-gauge pars plana vitrectomy in the fellow eye for progression of vitreous opacities with an accompanying decline in the vision to 20/40. Subsequently, the vision improved to 20/20 in the right eye following the vitrectomy.

One year after vitrectomy in the right eye and two years after vitrectomy in the left eye, the patient complained of disabling blur and glare in both eyes. On examination, the visual acuity was 20/25 OD and 20/20 OS. However, measurement of vision with the brightness acuity tester on the medium setting demonstrated a decline in vision under conditions of glare to 20/200 in the right eye and 20/70 in the left eye. Early nuclear sclerosis was present in both eyes, and dilated examination revealed bilateral dense opacities in the retrolental space with attachments to the posterior capsule of the lens (Fig. 1). Fundus examination showed vitreous opacities along the superior vascular arcade and in Cloquet's canal of the right eye. Optical coherence tomography demonstrated bilateral hyperreflective spots in the superficial retinal layers (Fig. 2).

Given the location of the amyloid deposits on the posterior lens capsule and the presence of cataract, a decision was made to perform cataract extraction followed by neodymium:yttrium-aluminum-garnet (Nd:YAG) posterior capsulotomy in the right eye. The visual acuity was

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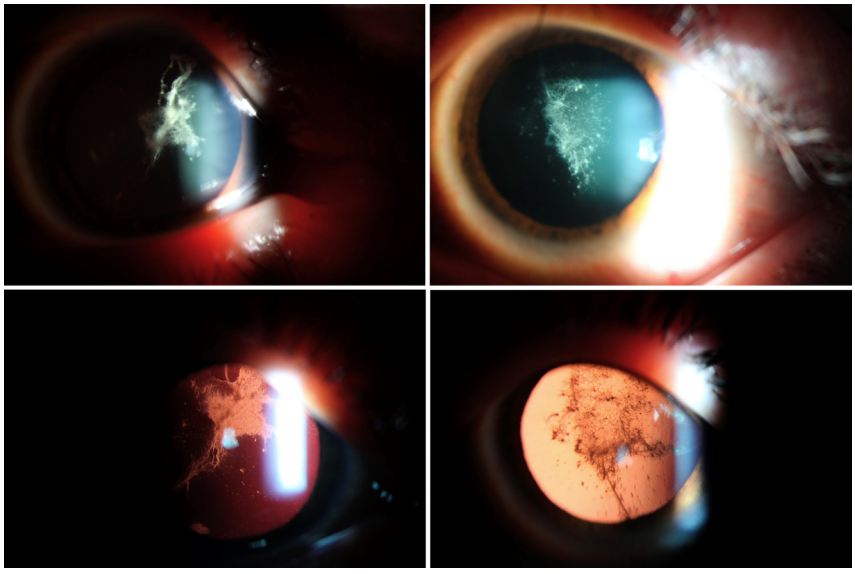

Fig. 1. Slit lamp photographs before lens extraction, showing amyloid deposits attached to the posterior lens capsule OD (left) and OS (right).

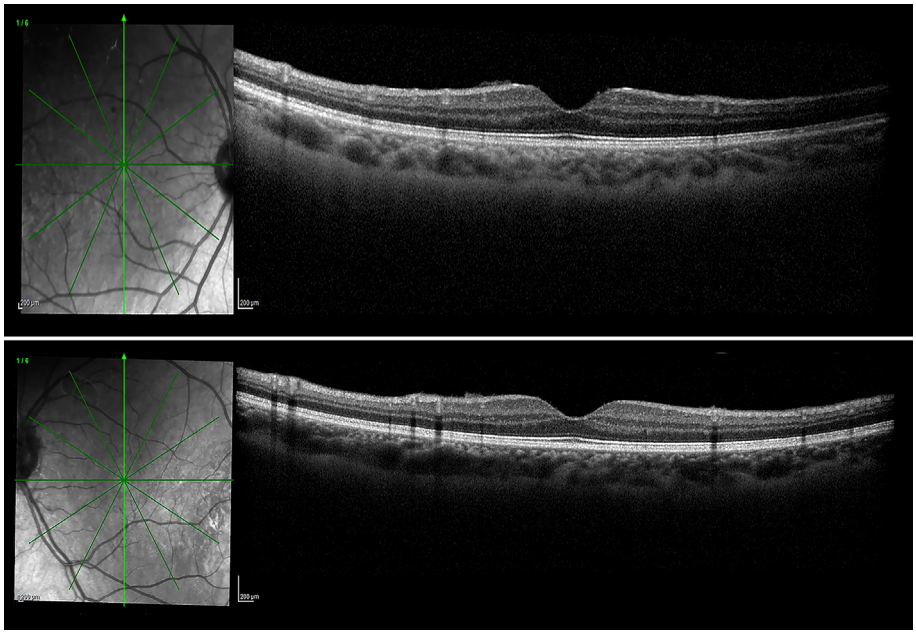

Fig. 2. Optical coherence tomography showing hyperreflective retinal deposits OD (top) and OS (bottom).

20/25 OD one month after phacoemulsification, at which time posterior capsulotomy was performed with energy settings of 1.5 mJ/pulse and 1 pulse per burst. Due to dispersion of the amyloid material partially obscuring the view during the laser procedure, two sessions were required with the first creating an opening in the posterior capsule and the second treating a remnant plaque of amyloid just posterior to the intraocular lens. Following the procedure, the patient had 20/20 vision OD with resolution of symptoms of glare. He received similar management in the left eye in a single session with clear 20/20 vision at one month following treatment (Fig. 3). At six months following the capsulotomies, he noted no decrease in vision.

3. Discussion

Familial transthyretin (TTR)-associated amyloidosis is a rare autosomal-dominant disorder caused by mutations in the TTR gene. TTR is a transport protein for thyroxine and vitamin A. Mutations can lead to the production of an abnormal protein, which deposits extracellularly in the form of amyloid fibrils.[1] Ocular involvement can occur and vitreous deposits are considered pathognomonic of TTR-associated amyloidosis.[2] Pars plana vitrectomy for vitreous opacities results in restoration of visual acuity.[3,4] However, recurrence of amyloid has been reported, most commonly in the retrolental space.[5–7] Some report that this recurrence is most likely due to a re-opacification of remnant vitreous in an incompletely vitrectomized eye.[5–7] Others report that recurrence can occur even with a complete vitrectomy and lens removal due to intraocular amyloid synthesis.[4] Although the liver is the source of circulating TTR, there is experimental proof of TTR synthesis in the rat eye retinal pigment epithelium.[8] De novo synthesis of mutated TTR in the retinal pigment epithelium may explain the continued ocular amyloid deposition even after liver transplant.[9]

The presentation in our patient with bilateral retrolental opacities could be explained by the accumulation of amyloid in Berger's space and production of amyloid in the eye. Supportive evidence of retinal

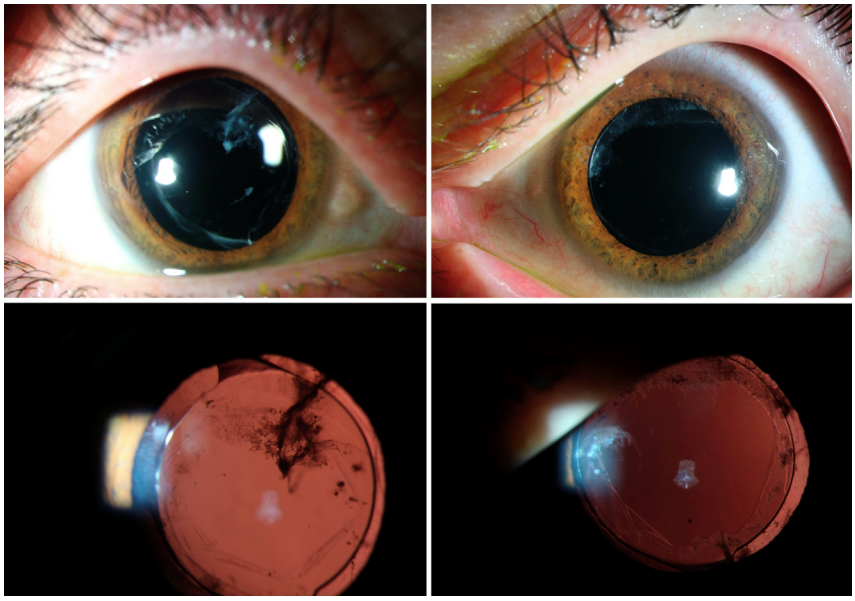

Fig. 3. Slit lamp photographs showing the opening in the posterior lens capsule after phacoemulsification and Nd:Yag capsulotomy OD (left) and OS (right).

origin of amyloid in our patient is shown by the hyperreflective deposits seen on the retinal surface on optical coherence tomography, similar to previous reports.[10,11] The vitrectomy performed likely allowed access of amyloid to Berger's space. The lens epithelium then likely provided a scaffold, supporting the accumulation of amyloid material. Basement membrane has previously been reported to be important for ongoing in-vivo amyloid fibrillogenesis and deposition. The hypothesis is that the basement membrane engulfs the TTR molecules, following which there is upregulation of basement membrane components, such as type IV collagen which, in turn, causes further deposition of TTR.[12] In previously reported cases of recurrence of ocular amyloid after vitrectomy, authors have described management by repeat vitrectomy.[5–7] In our patient, further vitrectomy could not have been safely performed, given the direct attachment of the amyloid deposits to the posterior lens capsule, and cataract surgery followed by capsulotomy led to an improvement in vision. Furthermore, the disruption of the posterior capsule by capsulotomy served to remove the membrane that appears to be central to the pathology of amyloid deposition. Similar to amyloid that accumulated in this space in our patient, we speculate that this space may allow for accumulation of cells in other conditions, such as vitreoretinal lymphoma.

4. Conclusions

In conclusion, recurrence of amyloid following vitrectomy can occur in the retrolental space with attachment of deposits to the posterior lens capsule. Glare testing may be important in recurrence of amyloidosis presenting with visual complaints and good visual acuity, as it can provide an objective measurement to guide further management. Cataract extraction followed by laser posterior capsulotomy can be beneficial in such patients.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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Conflict of interest: Dr. Pulido has stock in LAgen Laboratories, which supplies induced pluripotent stem cell-derived RPE cells for in vitro studies, and has no relevant financial interests. The following authors have no financial disclosures: SM, SK.

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