Bilateral Uveal Effusion Associated With Scleral Thickening Due to Amyloidosis

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A 45-year-old man with primary systemic amyloidosis was found to have bilateral uveal effusions secondary to thickened sclera according to magnetic resonance imaging of the orbits. The patient was treated with bilateral sclerectomies and vortex vein decompression, and had an excellent outcome. Light microscopy of excised sclera revealed severe infiltration of the tissue by amyloid. To our knowledge, this is the first report of amyloid infiltration of the sclera leading to uveal effusion.


Amyloid deposition has been reported both in ocular adnexae such as the eyelids, conjunctiva, and lacrimal gland, and components of the globe such as the cornea and vitreous body.1,2 Deposits in the cornea may result in recurrent erosion and visual impairment while vitreous involvement may be misdiagnosed as uveitis or old hemorrhage.2 Amyloid infiltration of the sclera has also been described, although there is no mention of any deleterious effect on the eye or vision.1,2 This clinicopathologic report describes a patient with primary systemic amyloidosis and scleral infiltration by amyloid, resulting in massive uveal effusion that responded to scleral surgery.

REPORT OF A CASE

A 45-year-old Filipino man with known primary systemic amyloidosis had a 4-month history of gradual bilateral painless loss of vision associated with the recent onset of metamorphopsia. Best-corrected visual acuity was 20/80 OU. There was 1+ flare and cells in the anterior chamber of each eye. Exudative retinal detachments with shifting subretinal fluid were present in each eye. Bilateral optic disc swelling consistent with optic disc drusen was also noted (Figure 1). Except for hypertension, his medical history was unremarkable. Results of other investigations, including a screening panel of tests for systemic vasculitis were normal.

The patient had first visited with infertility, and amyloidosis was diagnosed following testicular biopsy. There was known infiltration of the liver and kidneys by amyloid. There was no family history of amyloidosis.

Computed tomographic (CT) scanning of the orbits demonstrated thickening of the posterior aspect of each globe that was consistent with thickened coats of the eye, but scleral thickening could not be distinguished from choroidal thickening or retinal detachment in either eye. Magnetic resonance imaging (MRI) of the orbits revealed grossly thickened posterior sclera and choroidal engorgement (Figures 2 and 3). A tapering course of high-dose oral prednisone (1.5 mg/kg) was given without clinical improvement. Best-corrected visual acuity decreased to 20/200 OD and counting fingers OS, and the exudative retinal detachments increased in both eyes.

Scleral resection with vortex vein decompression was performed in the right eye by excising 8×8-mm pieces of partial-thickness sclera between the rectus muscles in each quadrant. The sclera was dissected posteriorly until the intrascleral portion of each vortex vein was exposed and decompressed (Figure 4). Two sclerostomies were performed to allow drainage...
of suprachoroidal fluid. Fifty per-
cent sulphahexafluoride gas was in-
jected into the vitreous to repressur-
ize the eye.

One day after surgery the reti-
nal detachment in the right eye had
almost completely resolved, and by
3 days after surgery there was no
evidence of retinal detachment or
choroidal effusion. Four months
after surgery, visual acuity had im-
proved to 20/60 OD, and the retina
remained completely flat. Subse-
quently, scleral resection surgery was
performed in the left eye with a simi-
lar outcome (Figure 1).

Microscopy confirmed the pres-
ence of Congo red–positive amy-
loid material in all the scleral speci-
mens (Figure 5). In addition, the
material was resistant to potassium

**Figure 1.** Left, Fundus photographs showing inferior exudative retinal detachment of the left eye in the sitting position 1 day before surgery. Right, Complete reattachment of retina and choroid 3 months after scleral surgery. There are small areas of chorioretinal atrophy in the macula. Note the mildly elevated optic disc and blurred optic disc margins consistent with optic disc drusen in both eyes.

**Figure 2.** Axial T2-weighted magnetic resonance imaging scan of the right orbit obtained with a surface coil showing abnormally thickened coats of the globe (arrowheads), retinal detachment (asterisk), and subchoroidal effusion within engorged choroid (arrows).

**Figure 3.** Axial contrast-enhanced T1-weighted magnetic resonance imaging scan with fat saturation of the right orbit shows differentiation between enhancing engorged choroid (white arrows) and surrounding scleral thickening (black arrows) comprising thickened coats of globe as well as retinal detachment (asterisk).

**Figure 4.** Photograph of a partial-thickness sclerectomy in the inferonasal quadrant. The incision begins along the line of the rectus muscles insertion and is approximately 8 × 8 mm long. Note the piece of excised sclera (asterisk) and decompressed vortex vein (arrow) as it exits from the sclera.
permanganate oxidation, confirming that the amyloid protein was of the primary type.³ Technical difficulties prevented typing the amyloid protein using immunohistochemistry.

**COMMENT**

Amyloid deposition in the sclera is uncommon, with only a small number of reports of patients with secondary amyloidosis.¹² Other ocular structures that may be affected by amyloid deposition include the cornea, choriocapillaris, the optic nerve dural sheath, and vessels of the optic nerve.¹ Vitreous involvement is found in hereditary transthyretin-related amyloidosis.² Globe involvement is usually asymptomatic, though pain and visual disturbance may complicate corneal and vitreous amyloidosis.² To our knowledge, this case is the first report of uveal effusion secondary to scleral infiltration by amyloidosis.

Medical treatment and conventional vitreoretinal surgery are ineffective in treating the uveal effusions seen in idiopathic uveal effusion syndrome and nanophthalmos.⁴⁻⁶ Partial-thickness sclerectomy and vortex vein decompression is a well-documented procedure for uveal effusion associated with abnormally thickened sclera.⁶ The presence of uveal effusion in this patient with ocular amyloidosis and the success of scleral surgery in relieving the effusions supports the theory that an abnormally thickened sclera interferes with the normal transcleral flow of fluid.⁴ This disturbance of bulk fluid flow across the choroid and sclera leads to accumulation of proteinaceous fluid in the choroid and subretinal space and the clinical appearance of shifting subretinal fluid characteristic of nonrhegmatogenous retinal detachment.⁷

This case report also highlights the importance of MRI scans in the investigation of patients with uveal effusion. In contrast to CT scanning, it is possible with MRI to differentiate a retinal detachment, choroidal and scleral thickening, and to show subtle ciliochoroidal effusions (Figures 2 and 3).

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**REFERENCES**


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Figure 5. Section of the sclera showing severe involvement by amyloid (left, Congo red, original magnification ×100; right, Congo red under polarized light, original magnification ×100).