European Journal of Ophthalmology / Vol. 18 no. 3, 2008 / pp. 473-475

SHORT COMMUNICATION

Needle-shaped deposits on retinal surface in a case of ocular amyloidosis

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PURPOSE. *To report a case in which optical coherence tomography (OCT) showed needle-shaped deposits on the retinal surface oriented toward the vitreous cavity and immunohistochemical findings suggested light chain-related amyloidosis.*
METHODS. *A 59-year-old man with no systemic complications had bilateral neovascular glaucoma and vitreous opacities in the right eye. Vitrectomy was conducted on the right eye and the excised vitreous was examined histopathologically.*
RESULTS. *Glass wool-like opacities were observed during vitrectomy. Postoperative fundus examination of the right eye showed retinal hemorrhage and white deposits around blood vessels and on retinal surface. Fluorescein angiography revealed hyperfluorescence of the optic disc, non-perfusion areas, and vascular focal staining. OCT depicted needle-shaped deposits perpendicular to the retinal surface oriented toward the vitreous. Histologic examination of deposits revealed positive reaction for Congo red stain, and immunohistochemical examination demonstrated positive reactivities for anti-lambda and anti-kappa light chains (precursors of amyloid protein), suggesting a diagnosis of light chain-related amyloidosis.*
CONCLUSIONS. *In this case, OCT showed needle-shaped deposits perpendicular to the retinal surface. Special staining with Congo red revealed the deposit to be amyloid deposition. Immunohistochemical staining suggested light chain-related amyloidosis. Vascular obstructive lesions and neovascular glaucoma secondary to retinal vascular damage in amyloidosis warrant particular attention. (Eur J Ophthalmol 2008; 18: 473-5)*

KEY WORDS. *Glass wool-like opacities, Light chain-related amyloidosis, Neovascular glaucoma, Optical coherence tomography, 25-Gauge transconjunctival vitrectomy*

Accepted: December 19, 2007

INTRODUCTION

Amyloidosis is a disease involving amyloid protein deposition at one or more sites in the body. Detection of precursor proteins in serum and genetic diagnostics are important for establishing a definitive diagnosis. Reported cases of amyloidosis with vitreous opacities mainly had familial amyloidotic polyneuropathy, while isolated vitreous amyloidosis is rare (1). Immunohistochemical demonstration of amyloid protein in a resected vitreous sample has not previously been reported.

We encountered a patient with vitreous opacities in whom light chain-related amyloidosis was diagnosed by immunohistochemical staining. From the reflection of amyloid deposition on the retinal surface depicted on optical coherence tomography (OCT), needle-shaped depositions oriented from the retinal surface toward the vitreous cavity were observed for the first time.

Case report

A 59-year-old man presented at our hospital in March 2006 because of deteriorating vision in both eyes. The patient had undergone vitrectomy and cataract surgery for vitreous

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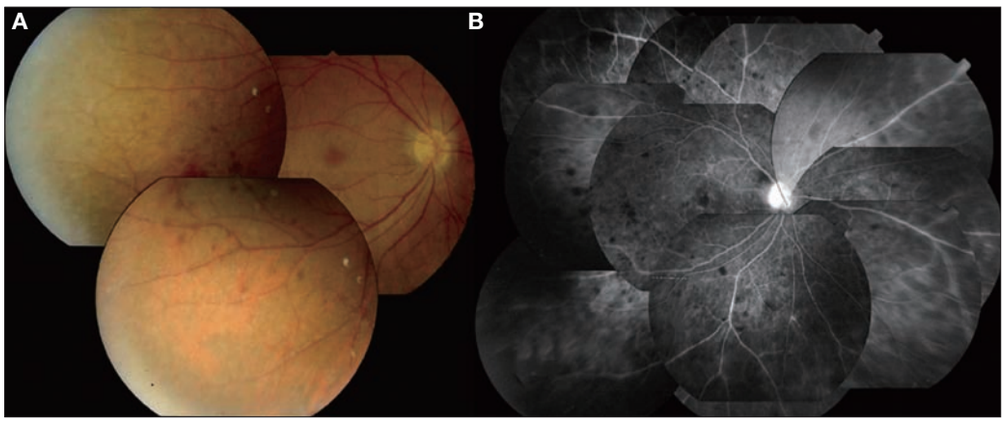

Fig. 1 - *Postoperative fundus findings of right eye.* **(A)** *Fundus photography showing retinal hemorrhage and white deposits around blood vessels and on retinal surface.* **(B)** *Fluorescein angiography showing hyperfluorescence of optic disc and blocked fluorescence due to retinal hemorrhage, and focal staining of retinal vascular wall.*

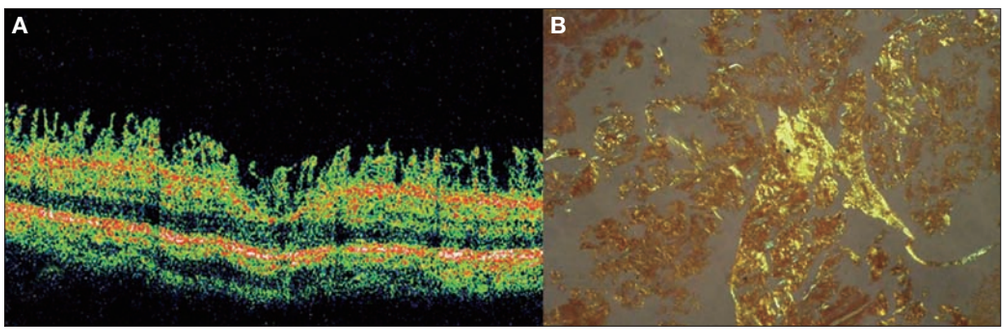

Fig. 2 - *Postoperative optical coherence tomography (OCT) and pathologic findings of right eye.* **(A)** *OCT depicting needle-shaped deposits oriented from the retinal surface toward the vitreous cavity in the macular region.* **(B)** *Yellowish green birefringence under a polarized light microscope (original magnification, ×200).*

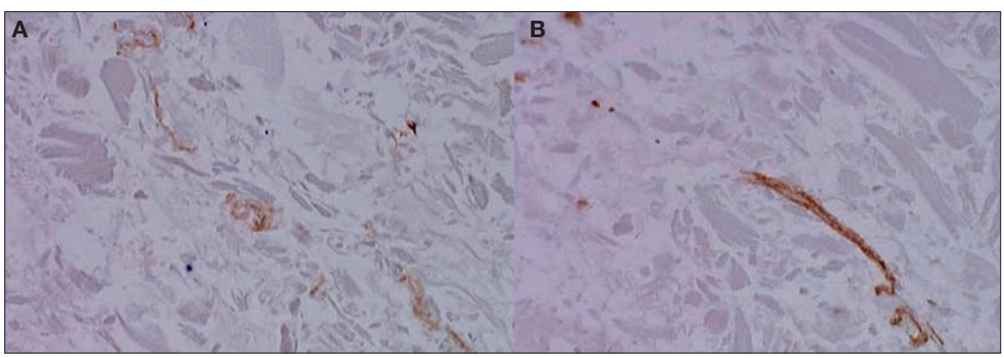

Fig. 3 - *Immunohistochemical findings.* **(A)** *Immunohistochemical study showing positive reactivity for lambda chain, a precursor of amyloid protein (original magnification, ×200).* **(B)** *Immunohistochemical study showing positive reactivity for kappa chain, a precursor of amyloid protein (original magnification, ×200).*

opacities in both eyes in 1997, and also Seton surgery for neovascular glaucoma of the left eye in 2004. He had no systemic complications and his family history was unremarkable. The cause of vitreous opacity was unknown because histopathologic examination had not been conducted after the vitrectomy.

At presentation, corrected visual acuity tested with the standard Japanese decimal visual acuity chart was 1.2 in the right eye and 0.08 in the left eye. Intraocular pressure was 50 mmHg in the right and 38 mmHg in the left. Intraocular lenses were implanted in both eyes. Examination revealed neovascular glaucoma in both eyes and opacities in the right eye. White deposits on the papillary border, neovascularization, and peripheral anterior synechia in the angle were observed in both eyes. In the left eye, glaucomatous disk cupping was observed but no retinal abnormality was found, and OCT also showed no abnormality. In the right eye, fundus visibility was poor due to poor mydriasis, therefore OCT was not possible. From the above findings, vitrectomy was scheduled only for the right eye showing recurrent vitreous opacities.

In March 2006, 25-gauge transconjunctival vitrectomy was conducted using the Accurus 800CS^{TM} (Alcon Surgical, Fort Worth, TX) with a 25-gauge system (Medical Instrument De-

velopment Laboratories, San Leandro, CA). When right vitrectomy was conducted, glass wool-like opacities were observed extending from the periphery to the ciliary body. Intraoperative fundus examination revealed retinal hemorrhage and white deposits on the retinal surface. Since ocular amyloidosis was suspected clinically, the excised vitreous was investigated histopathologically.

Post-vitrectomy examinations of the right eye are shown in Figure 1. Fundus examination showed retinal hemorrhage as well as white deposits around blood vessels and on the retinal surface (Fig. 1A). Fluorescein angiography (FA) revealed hyperfluorescence in the optic disc, blocked fluorescence due to retinal hemorrhage, focal staining of the retinal vascular wall, and nonperfusion areas (Fig. 1B). OCT depicted needle-shaped deposits perpendicular to the retinal surface oriented toward the vitreous (Fig. 2A). Staining of the deposits with hematoxylin and eosin (HE) showed a homogenous eosinophilic amorphous substance, which stained positively with Congo red. The Congo red–stained area showed yellowish green birefringence when observed by polarizing microscope (Fig. 2B) confirming a diagnosis of amyloidosis. Immunohistochemical staining revealed positive reactivities for anti-lambda light chain (Fig. 3A) and anti-kappa light chain (Fig. 3B), both of which are precursors of light chain amyloid protein. Amyloid AA protein was negative. Although deposition of amyloid protein was found in the conjunctiva, none was observed in the trabecular meshwork.

DISCUSSION

In patients with amyloidosis, vitreous opacity reportedly recurs at a high frequency despite vitrectomy (2). The present case also had recurrent disease after the vitrectomy conducted in 1997. A previous study suggested that vitreous opacity probably occurs via deposition of amyloid protein on the retinal vascular wall followed by spread from the vascular wall to the vitreous (3). In the present case, OCT showed needle-shaped depositions perpendicular to the retinal surface oriented toward the vitreous, and this image presumably depicts the in vivo adhesion of amyloid protein to the retinal surface and/or spread of amyloid into the vitreous.

An increase in outflow resistance due to amyloid deposition in the trabecular meshwork has been reported to be the mechanism of glaucoma development secondary to amyloidosis (4). In the present case, there was no amyloid protein deposition in the trabecular meshwork. Rather, secondary angle-closure glaucoma with angle neovascularization was observed, suggesting that intraocular ischemia may have caused glaucoma.

In patients with amyloidosis, fundus findings of venous dilatation, linear and dot hemorrhages, soft exudates, multifocal sheathing, and pigmentary mottling have been reported. Similar findings were found in this case. The fundus findings showed no improvement more than 1 year after vitrectomy, therefore recurrence is likely. A case of amyloidosis presenting rubeotic glaucoma with FA findings of retinal neovascularization and extensive retinal occlusion was reported, and panretinal photocoagulation was conducted (5). Stenosis of the retinal intravascular lumen was observed histopathologically in an autopsied eye (3). Therefore, we speculate that in the present case, neovascular glaucoma probably developed due to expansion of the nonperfusion areas resulting from vascular obstructive lesions secondary to amyloidosis.

This case indicates that ocular amyloidosis may be associated with retinal vascular damage, and attention should be paid to the development of secondary vascular obstructive lesions leading to neovascular glaucoma.

This study was supported by regular departmental research funds. There are no conflicts of interest regarding this work or its submission. None of the authors has proprietary interest.

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