·Clinical Research·

Vitrectomy for vitreous amyloidosis

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Abstract

• AIM: To study the clinical features of vitreous amyloidosis and the effect of vitrectomy for it.

• METHODS: We treated 6 eyes (4 Patients) with vitreous amyloidosis from 2008 to 2009, and followed up for 18-30 months after vitrectomy.

• RESULTS: The visual acuity ranged from counting fingers to 3/50 before surgery and 15/50-40/50 after vitrectomy. No severe complication was observed.

• CONCLUSION: Vitrectomy is an effective and safe treatment for eyes with vitreous amyloidosis. A long period of follow-up after surgery should be performed.

• KEYWORDS: vitreous amyloidosis; surgical treatment

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INTRODUCTION

Vitreous amyloidosis is a rare condition that mainly occurs in Familial Amyloidotic Polyneuropathy (FAP) [1-5]. Vitreous opacities may be the earliest occurring or, in some cases, only symptom of this disorder [5-10]. In such cases a family history of amyloidosis is usually present.

We treated 6 eyes (4 Patients) with vitreous amyloidosis from 2008 to 2009, and followed up for 18-30 months after vitrectomy. Now we reported the outcome of surgery as follows.

PATIENTS AND METHODS

Case 1 A 38 years old man presented with a ten year complaint of decreased vision and floaters in both eyes. His family history was positive in three of ten family members. Visual acuities measured counting fingers at 1 foot in both eyes, with normal intraocular pressures. Slit lamp examination showed linear vitreous opacities attaching to the posterior lens capsule by "footplate". Fundus examination after pupillary dilatation revealed dense vitreous opacities with a glass-wool appearance, which block the view to the fundus in both eyes (Figure 1A). B-scan ultrasound examination showed that bilateral vitreous was full of middle-echo spot (Figure 2A). Pars plana vitrectomy was carried out in both eyes. Intraoperatively, the eyes showed severe vitreous amyloid deposition and had very strong vitreoretinal adhesions. The performation of surgical posterior vitreous detachment was rather difficult. The diseased vitreous was removed as thoroughly as possible. Local endolaser was applied to the area of degeneration in the inferior retina.

Case 2 A 35 years old man, the younger brother of patient 1, presented with a five year history of bilateral floaters with progressive blurring of vision. The visual acuities, intraocular pressures, slit lamp examination and fundus examination were similar to that of patient 1. B-scan ultrasound examination showed bilateral vitreous opacities and posterior vitreous detachment (Figure 2B). Pars plana vitrectomy was carried out in the left eye. Intraoperatively, the left eye showed numerous amyloid depositions in vitreous. The complete posterior vitreous detachment had existed and the diseased vitreous was removed completely.

Case 3 A 55 years old woman presented with a two year complaint of decreased vision and floaters in both eyes. She had a cousin who had the similar history to hers. Visual acuities measured counting fingers before eye in the right eye and 1/50 left eye, with normal intraocular pressures. Slit lamp examination showed linear vitreous opacities attaching to the posterior lens capsule by "footplate" (Figure 3A). Fundus examination after pupillary dilatation revealed vitreous opacities with a white streak appearance, which blurred the view to the posterior pole in both eyes. B-scan ultrasound examination showed that bilateral vitreous was full of hypoechoic spot. Pars plana vitrectomy was carried out in both eyes. Intraoperatively, the both eyes showed numerous amyloid depositions in vitreous and had very tight vitreoretinal adhesions. The surgical posterior vitreous detachment was performed difficultly and the diseased vitreous was removed as thoroughly as possible. One iatrogenic break in the nasal serrata was treated with cryopexy in the right eye. Fluid-gas exchange was performed after that, and then the intraocular gas was replaced with 16% perfluoropropane.

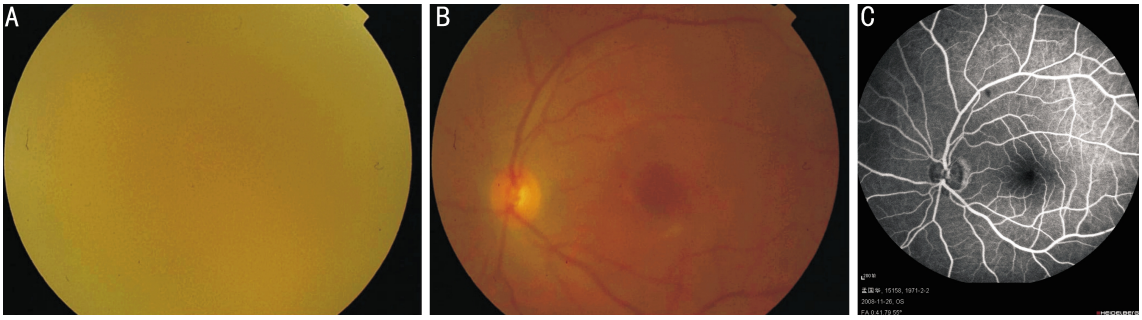

Figure 1 The left eye of patient 1 A:Fundus photograph before vitrectomy showing severe vitreous opacities; B:Postoperative fundus photograph of the same eye; C: FFA post vitrectomy of the same eye showing normality

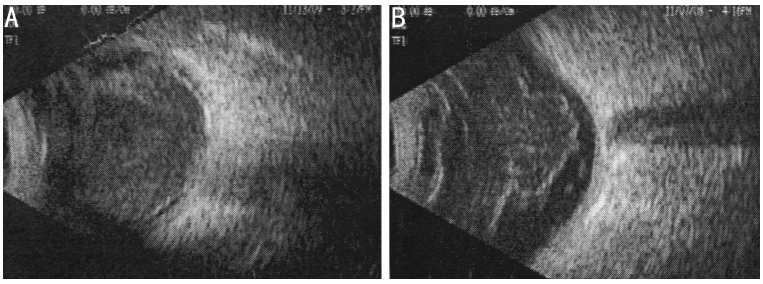

Figure 2 Ultrasonographic picture in the left eye of patient 1 A:Ultrasonographic picture showing vitreous was full of middle-echo spot; B: Ultrasonographic picture showing vitreous opaciyies and posterior vitreous detachment

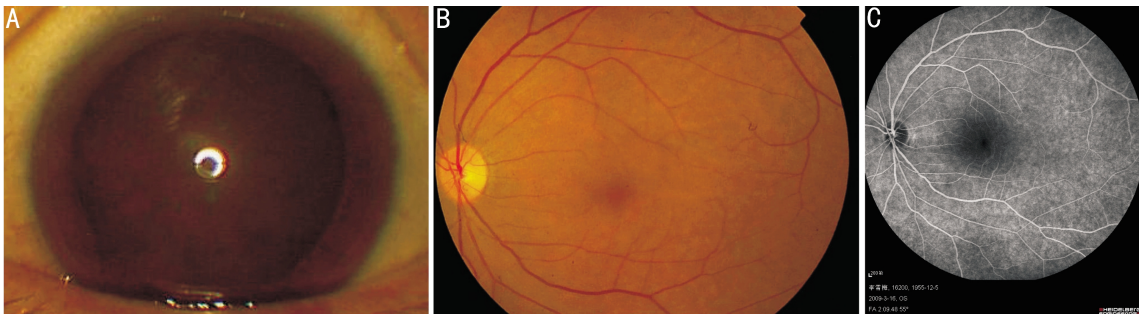

Figure 3 Photograph of the left eye of patient 3 A:Photograph of the left eye of patient 3 before vitrectomy, showing vitreous opacities attaching to the posterior lens capsule by "footplate"; B:Postoperative fundus photograph of the same eye; C:FFA post vitrectomy of the same eye showing normality

Case 4 A 66 years old woman presented with a two year complaint of decreased vision and floaters in the right eye. Her family history was negative. Visual acuities measured 3/50 in the right eye and 10/50 left eye, with normal intraocular pressures. Slit-lamp examination showed linear vitreous opacities attaching to the posterior lens capsule by "footplate" in the right eye. Fundus examination after pupillary dilatation revealed sheet-like veils vitreous opacities (Figure 4A). The retina seemed normal by indirect ophthalmoscope examination in both eyes. B-scan ultrasound examination showed that vitreous was full of hypoechoic spot in the right eye. Fundus fluorescein angiography (FFA) of the right eye showed opacities in the refractive media, fluorescent shelter before the optic disc, and dye staining in the walls of vessels in the inferior retina (Figure 4B, C). Pars plana vitrectomy was carried out in the right eye. The performation of surgical posterior vitreous detachment was very difficult and the diseased vitreous was removed as thoroughly as possible. Local endolaser was applied to the area of abnormal vessels in the inferior retina.

RESULTS

Pathological analysis of all vitreous specimens showed Congo red staining and confirmed the diagonosis of vitreous amyloidosis. All patients were followed up for 18-30 months after vitrectomy. Visual acuities, intraocular pressures, slit lamp examination, fundus examination, and FFA were observed (Figure 1B, C; Figure 3B, C).

The visual acuities ranged from counting fingers to 3/50 before surgery and 15/50-40/50 after vitrectomy (15/50 in 1 eye, 25/50-30/50 in 3eyes, 40/50 in 2 eyes).

Intraocular pressures were normal in all eyes throughout the follow-up period. No severe complication such as retinal detachment, recurrent vitreous opacities or glaucoma was observed throughout the follow-up period. FFA post vitrectomy showed dye leakage from peripheral capillary and dye staining in the walls of vessels in 1 eye respectively. The results of FFA in the other eyes were normal.

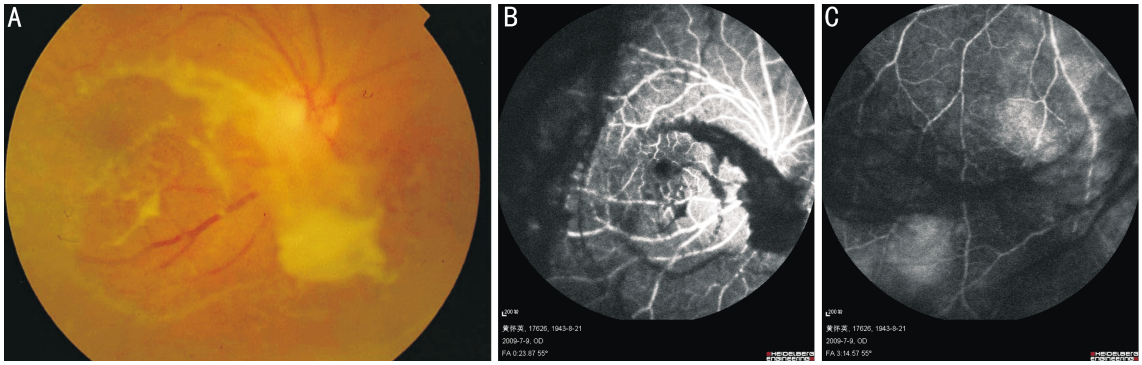

Figure 4 The right eye of patient 4 A:Fundus photograph before vitrectomy showing vitreous opacities; B:FFA of the same eye showing fluorescence shelter before the optic disc; C:FFA of the same eye showing dye staining in the walls of vessels

DISCUSSION

Familial amyloid polyneuropathy (FAP) caused by mutations in the transthyretin (TTR) gene is the commonest form of hereditary amyloidosis. Until 20 years ago, FAP was thought to be restricted to endemic occurrence in certain areas. However, owing to progress in biochemical and molecular genetic analyses, FAP is now believed to occur worldwide [11]. TTR is a tetrameric plasma protein (prealbumin), which mainly synthesised in the liver. TTR is also synthesised by the retinal pigment epithelium cells and in the choroid plexus of brain [12]. The chief function of TTR is to transfer thyroxine and retinol-binding protein. It polymerises into a β-pleated structure of amyloid fibril and deposits in the organ and tissue abnormaly causing disease.

FAP may implicate multiple tissues and organs, including peripheral nerves, kidneys, and cardiac muscle, as well as within the eye. In FAP patients, phenotypic differences in age of onset, location and severity of lesions occur.

Amyloid may deposit in the vitreous body, conjunctiva, iris, eyelid, trabecular meshwork, lacrimal gland, and ciliary nerves [13]. The main ocular manifestations are: abnormal conjunctival vessels, keratoconjunctivitis sicca and pupillary abnormalities. Less common features include vitreous opacities and secondary glaucoma [12]. The incidence of vitreous opacities in FAP varies from 5.4% to 35% and the density of the opacities determines the visual acuity and symptoms [14].

Many literatures have been reported that vitrectomy was a safe treatment for vitreous amyloidosis and improved patient's visual acuity significantly [15,16]. The difficulties of surgery lie in removing the vitreous completely and postoperative recurrence of vitreous opacities. The latter may be primarily caused by two mechanisms, dispersion of residual vitreous opacities and intraocular production of amyloid fibrils that occur even after surgery. There is a high risk of iatrogenic retinal breaks formation intraoperatively because of the tight vitreoretinal adhesions, especially in the vicinity of retinal vessels. Doft, Irvine et al had reported for developing of retinal detachment and postoperative recurrence of vitreous opacities [17,18]. So, the surgeon should not only remove the vitreous as thoroughly as possible, but also avoid the occurrence of iatrogenic retinal break intraoperatively. The surgical operation should be performed gently and carefully. In addition, the surgeon should check the whole retina before ending the procedures, and the iatrogenic break should be treated with photocoagulation or cryopexy. If not, severe complication such as retinal detachment may occur.

In this study, one iatrogenic break in the nasal serrata was found when checking intraoperatively. We treated it with cryopexy, and then, fluid-gas exchange, tamponading with 16% perfluoropropane was performed. After the above treatment，no postoperative complication occured. The posterior vitreous detachment is a relatively rare condition that occurs in vitreous amyloidosis. In this study, 1 case of eye had pre-existing posterior vitreous detachment before surgery. It reduced the difficulty of surgery significantly. Glaucoma in FAP may be due to amyloid protein released into the aqueous leading to trabecular meshwork obstruction, or due to raised episcleral venous pressure. So, the long-term monitoring and controlling of intraocular pressure in patients with vitreous amyloidosis is very important.

Qiao *et al* [3] reported for 1 case of vitreous amyloidosis complicated with glaucoma. They thought that vitrectomy is helpful for controlling of intraocular pressure in such kind of patient, and the urgent performation of anti-glaucoma surgery is not necessary.

With regard to the therapy for FAP, liver transplantation has been widely employed as the only potential cure for TTR-related FAP patients, since TTR is mainly produced in the liver [19]. Although more than 90% of the mutant TTR is produced in the liver, and after liver transplantation its levels in the serum can be reduced to less than 1% of pretransplant levels, it continues to be produced within the eye by the

retinal pigment epithelium, thus allowing continued ocular involvement. Hara et al [20] followed-up of a number of FAP patients after liver transplantation for a long period. Their observation showed that 36% patients developed vitreous opacities and 18% patients developed glaucoma. They thought that patients with FAP who undergo liver transplantation continue to have a long-term risk of severe ocular manifestations, especially vitreous opacities and glaucoma, which can restrict their daily lives.

In this study, FFA examination of eyes with vitreous amyloidosis showed dye leakage from peripheral capillary and staining in the walls of vessels in 1 eye respectively, and the majority of the eyes were normal. We didn't found characteristic change for vitreous amyloidosis in FFA, which was consistent with the previous literatures[21,22]. In this study, the visual acuities of patients with vitreous amyloidosis were improved significantly, and no severe complication was observed throughout the follow-up period. The results were satisfying.

Vitreous amyloidosis may complicate with not only severe ocular complications such as secondary glaucoma, retinal vein occlusion [23-25], but also systemic diseases. So, careful long-term follow-up of patients with vitreous amyloidosis is essential.

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