Original Article

Surgery treatment for the proliferative retinal detachment associated with macular hole in the morning glory syndrome

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Abstract

Purpose: To evaluate the efficacy of vitrectomy with peripapillary photocoagulation and silicone oil tamponade for the proliferative retinal detachment associated with macular hole in children with morning glory syndrome.

Methods: Eight children with morning glory syndrome (mean age 8.0±2.8 years; range 5–13 years) were included; all patients had unilateral eye disease and were misdiagnosed as having bilateral squint or amblyopia for years, with best corrected visual acuity less than 0.1. Five patients could not cooperate with the fundus examination and one patient had lens opacities. B ultrasound confirmed that all eight patients had retinal detachment and optic disc dysplasia. All patients underwent standard 3-port pars plana vitrectomy surgery (20G for three cases and 23G for five cases). At surgery, all patients were confirmed to have morning glory syndrome, macular hole, and proliferative retinal detachment; two cases had a funnel-shaped bulge. All the retinal detachment involved the macular area, and macular hole was detected in the abnormal expansion excavation of optic disk. The epiretinal membrane and subretinal membrane were completely peeled during the surgery. Combined photocoagulation in abnormal expansion excavation of optic disk and silicone oil tamponade were also performed.

Results: All eyes achieved anatomical resolution. After follow-ups ranging from eight months to four years, the visual function for all patients was improved by postoperative refractive correction associated with vision training. Best corrected visual acuity was 0.01 to 0.2 at the final follow-up, no retinal detachment recurred, and no silicone oil fluid flowed into the subretinal space. The silicone oil was successfully removed postoperatively after 1.5 years.

Conclusion: Vitrectomy with peripapillary photocoagulation and silicone oil tamponade is effective in treating the proliferative retinal detachment associated with macular hole in children with morning glory syndrome. (***Eye Science 2013***; ***28***: –)

Keywords: Morning glory syndrome; Peripapillary photocoagulation; Proliferative retinal detachment; Macular hole

Introduction

Morning glory syndrome (MGS) is a rare congenital optic disc dysplasia consisting of a conical excavation of the posterior pole including the optic disc. This excavation is filled with glial tissue, and a pigment ring slightly protrudes into the peripapillary area. MGS is often accompanied by persistent hyper plastic primary vitreous, congenital cataracts, microphthalmia, retinal defects, choroidal defects, lens defects, scleral defects, etc. Retinal detachment may develop in some morning glory syndrome patients[1–2], as the optic nerve is located centrally and surrounded by a deeply excavated scleral defect. Fluid sometimes accumulates within and beneath the retina and tractional forces may cause hiatus retinal detachment. The holes often are located in the abnormal optic disc area or at the optic disc edge and can be difficult to detect. The best treatment for these patients is also unclear. This study provides a retrospective review since 2003 of eight patients (eight eyes) with proliferative retinal detachment (PVD) and macular hole(MH); all patients were MGS children. All cases underwent vitreous surgery and the outcomes are reviewed.

Material and methods

Subjects

Eight cases of MGS were involved, three were

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males and five were females, and all were aged 5 to 13 years (6.5 ± 2.1 years). All patients had unilateral eye disease and had been misdiagnosed for bilateral squint or amblyopia for years. All eyes (5 eyes were esotropia and 3 eyes were exotropia) were associated with refractive errors and were able to make normal eye movements. All patients had normal intraocular pressure, and denied any family history; no other abnormalities were detected. Five patients could not cooperate with the fundus examination and one patient had lens opacities. B ultrasound confirmed that all eight patients had retinal detachment and optic disc dysplasia. During the surgery, all were confirmed to have MGS, MH, PVD, and subretinal proliferation. Two cases had a funnel-shaped bulge. Large excavated discs (about 1.5PD in diameter) were present, with retinal vessels radiating from the periphery. A tuft of translucent fibrous tissue covered the central part of the excavation and peripapillary chorioretinal pigmentary change was evident. The macular area with its xanthophyll pigment was displaced to the temporal margin of the excavation, while the papillary region was filled with acoustically dense material reminiscent of a (withering) flower and surrounded by a broad elevated, yellowish-white zone of chorioretinal pigmentary disruption. No central artery or vein was clearly distinguished; the blood vessels appeared to arise from the peripheral disc substance and course radially over the peripapillary annulus toward the retinal periphery. The vessels appeared to increase in number and to run an abnormally straight course, while tending to branch at acute angles. Serious macular detachment was involved and macular holes were detected in the abnormal expansion excavation of the optic disk.

Methods

All patients underwent standard 3-port pars plana vitrectomy surgery (20G for three cases and 23G for five cases), seven transparent lenses were well-preserved and one patient with cataract was treated using phacoemulsification. The epiretinal and subretinal membranes were completely peeled during the surgery; subretinal proliferation could be cut through the macular hole. The internal limiting membrane peeling and the gas/liquid exchange process were followed by flattening of the peripapillary retina with heavy liquids. Joint photocoagulation in the abnormal expansion excavation of the optic disk and silicone oil tamponade were also performed. The laser should be directed obliquely away from the posterior pole to avoid the macular area.

Results

Table 1 shows the clinical data for the patients. All eyes achieved anatomical resolution. After follow-ups ranging from eight months to four years, the visual function for all patients was improved by postoperative refractive correction associated with vision training. Best corrected visual acuity reached 0.01 to 0.2 at the final follow-up. No retinal detachment recurred and no silicone oil flowed into the subretinal space. The silicone oil was successfully removed postoperatively after 0.5 years in five cases and after 1-1.5 years in three cases. No recurrence of retinal detachment occurred during the follow-ups and the MHs closed in five cases (5/8, 62.5%).

Table 1 Basic data of patients in this study

case	age	sex	cataract (+/-)	PVR (+/-)	silicone oil filling time	macular hole (+/-)	Follow-up Time postoperative	preoperative visual acuity	postoperative visual acuity (last follow-up)
1	5	male	-	+	5 m	-	4y	FC/30 cm	0.01
2	11	female	-	+	17 m	+	8m	FC/30 cm	0.01
3	6	male	-	+	7 m	+	2y	FC/30 cm	0.05
4	7	male	-	+	6.5 m	+	3y	0.05	0.2
5	6	female	-	+	14 m	-	2.5y	0.01	0.05
6	9	female	+	+	3 m	+	10m	0.02	0.1
7	7	male	-	+	11 m	-	1y	FC/30cm	0.1
8	13	female	-	+	4 m	+	4y	0.01	0.2

cataract(+/-) - + with cataract; - without cataract

macular hole(+/-) - + closed macular hole; - unclosed macular hole

PVR -+ with proliferative vitreoretinopathy; - without proliferative vitreoretinopathy

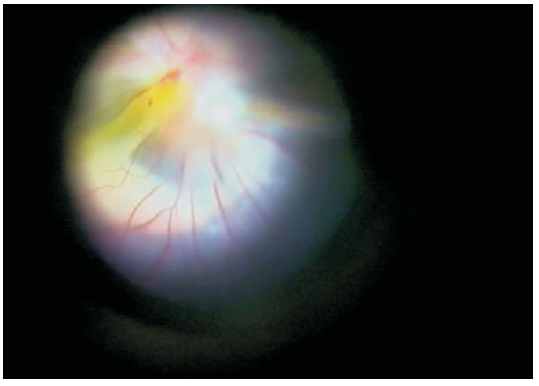

Figure 1 Proliferative retinal detachment in morning glory syndrome

Discussion

MGS is a rare, usually unilateral condition, where the disc is usually enlarged, with a funnel-shaped excavation surrounded by a usually raised pigmentary border.The macula is usually implicated and hides within the abnormal disc tissue. The retinal vessels emerge from the rim of the disc in a radial pattern, like the spokes of a bicycle wheel. Kindler, in 1970, was the first to clinically define and note the remarkable resemblance to the flower of the same name[3]. MGS may be a primary mesenchymal abnormality and also may be the result of dilation of the optic nerve head caused by dysgenesis of the terminal optic stalk, which fails to close, thereby leading to persistent excavation of the optic nerve head[1–2]. Retinal detachment is a frequent complication of the morning glory optic disc syndrome.

Some authors have reported that the liquid vitreous does not reach the subretinal space via a retinal tear[4]. Others have argued that glial hyperplasia on the disc, with progressive tangential traction on the adjacent retina and very small retinal holes lying in the optic disc may provide a fluid pathway, as these are likely to be covered by tissues before the disc. They suggested that vitreous surgery combined with peripapillary photocoagulation was an effective treatment[5–6].

In this study, the maculas of the eight patients were all located within the abnormally enlarged optic disk and were accompanied with significant MHs. The markedly enlarged scleral opening, with poorly differentiated retinal tissue and pigment epithelium herniating through the scleral opening, down alongside the optic nerve, may contribute to the development of the MHs. Patients are usually misdiagnosed with bilateral squint or amblyopia and this delays timely treatment because they are too young to cooperate, and fundus and B ultrasound examinations are lacking. All eight cases in this study had been misdiagnosed and were prescribed with long-term vision training. Early diagnosis and timing of treatment prior to the onset of proliferative retinopathy would be of great benefit to these patients a lot. Fundus examination, B ultrasound, and CT are important in children with unexplained amblyopia.

During the surgery, all patients were confirmed to have MGS, MH, PVD, and subretinal proliferation. The proliferation membrane was peeled completely and silicone oil was used to fill the vitreous cavity. Vitrectomy needs to be performed thoroughly, and all the traction efforts have to be relieved since most patients are children and are more likely to develop postoperative proliferative vitreoretinopathy. Managing vitrectomy in children remains challenging because the vitreous is dense, tough, and tightly adhering to the retina, and posterior vitreous detachment is less likely to occur. Vitrectomy must be handled carefully to avoid the formation of iatrogenic macular or retinal holes.

The differential diagnosis of MGS at surgery is particularly important because combined peripapillary photocoagulation is not required for several diseases with the optic disc abnormality such as:1) posterior sclera staphyloma, which often occurs in highly myopic eyes after the posterior scleral expansion; 2) papillary defects, where rare straight vascular vessels may occur at the optic disc edge as shown by the fundus fluorescein angiography;3) deep papillary depression in glaucoma or congenital papillary depression; this depression does not exceed the optic disc edge and fundus fluorescein angiography shows no abnormality;4)posterior choroidal defect, where the morphology of the papilla is normal, with no depression, and the choroidal defect is pale; 5) a variety of retrobulbar tumors, such as gliomas, meningiomas, or nerve sheath tumor, etc.

The current studies show that a typical MGS with associated retinal detachment may be attributed to

the tiny retinal holes in tissue lying in the optic disc. The report of dye flowing from the subarachnoid space into the subretinal space seems to provide a conclusive evidence for the connection between these two spaces. In addition, inserted gas bubbles may pass from the vitreous cavity through the perineural space, the sheath window, and into the orbit, suggesting that the vitreous, the subarachnoid, and even the orbital spaces may be interconnected.

Endophotocoagulation was applied around the margin of the optic disc to block the abnormal communication between the optic disc and the subretinal space, so that complete retinal reattachment would be probable. When dealing with macular holes in this study, the clinically significant holes were easily noted and closed, but a considerable possibility remained that small holes persisted in the margin of the optic disc. As a result, a combined peripapillary photocoagulation would be preferable.

Most physicians agree that excessive laser treatment would cause damage to the retinal nerve fiber layer in the region of the papillomacular bundle and increase the risk of severe visual loss. Sehepems et al. proposed that a low-energy argon laser beam would produce chorioretinal adhesion without causing damage to the optic nerve fibers. Since the macular area was located deep within the abnormal excavation of the optic disk in this study, peripheral 360° photocoagulation would not damage the papillomacular bundle region. The temporary silicone oil tamponade in MGS eyes appears to provide good long-term anatomic success of the repair of the MHs and the detached retina. However, children are too young to position their heads downward for enough days and their risk of complications is higher than in adults. Therefore, the silicone oil must be removed during the period when the upright effect of silicone oil is still effective (4–6 months postoperative), when the retina has achieved anatomical resolution and the holes have been completely closed. No complications of the silicone oil tamponade, such as cataract, glaucoma, or uveitis occurred in this study and the retina remained flat after the silicone oil removal.

In this research, vitrectomy with peripapillary photocoagulation and silicone oil tamponade was applied to treat PVD associated with MH in children with MGS. All eyes achieved anatomical resolution and the silicone oil was successfully removed 0.5 year postoperatively in five cases and 1–1.5 years postoperatively in three cases. The vision function achieved a satisfying improvement with postoperative refractive correction and vision training. The following points concluded from this research need to be addressed.

In summary, vitrectomy with peripapillary photocoagulation and silicone oil tamponade appears to be effective in treating the PVD associated with MH in children with MGS.

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