Communication between the subretinal space and the vitreous cavity in the morning glory syndrome

Abstract • Background: The aim was to describe a pathogenic mechanism for a rhegmatogenous retinal detachment in a 69-year-old man with the morning glory syndrome. • Methods: During vitreous surgery for a retinal detachment, a membrane was removed that covered the optic disc anomaly and produced traction on the peripapillary retina. A retinal hole was found in tissue lying within the optic cup, and the hole was sealed using a autologous plasma-thrombin mixture. Silicone oil was used for retinal tamponade. • Results: A retinal hole in tissue lying within the optic cup provided a fluid pathway between the vitreous cavity and the subretinal space. Following vitrectomy surgery, bubbles of silicone oil passed through the retinal hole into the subretinal space of the macula. • Conclusion: This case demonstrates that a retinal hole in tissue lying within the optic disc anomaly of the morning glory syndrome provides a communication for fluid between the subretinal space and the vitreous cavity, resulting in a rhegmatogenous retinal detachment. Vitreous replacement with silicone oil resulted in the migration of silicone bubbles into the subretinal space.

Introduction

The morning glory syndrome is a rare unilateral condition characterized by an enlarged and excavated optic disc. The central area of the optic disc is covered by a fibroglial tissue, and the adjacent peripapillary region contains a ring of elevated subretinal tissue with pigmentary changes in the retinal pigment epithelium [1, 2]. The distinctive appearance of the optic disc anomaly is named for the morning glory flower [2].

The retinal vessels are visible only at the margin of the optic disc because the central area of the optic disc is covered with a fibroglial tissue. Anomalous branching of the retinal vessels may be observed near the vicinity of the optic disc margin, and some of the retinal vessels in the peripapillary area may appear to be abnormally straight, attenuated, and sheathed [3]. Occasionally the macula is displaced towards the optic disc [4].

Retinal detachments have been reported to occur in 30–38% of patients with the morning glory syndrome. In most retinal detachments a retinal break has not been found. The retinal detachment is usually limited to the peripapillary retina or the posterior pole, but occasionally extensive bullous detachments may develop [1, 5, 6].

We describe the case of a 69-year-old man with the morning glory syndrome who developed recurrent rhegmatogenous retinal detachments. During vitreous surgery a communication for fluid was noted between the vitreous cavity and the subretinal space via a small retinal hole in tissue lying within the central area of the morning glory disc anomaly.

Case report

A 69-year-old man had a history of a congenitally deformed left eye with no light perception. The right eye had the morning glory syndrome and a history of glaucoma. The right eye had under-
On 11 May 1993, the visual acuity in the right eye was hand motion. There was a retinal detachment limited to the posterior pole by scatter photocoagulation in the right eye. No retinal breaks were identified. A membrane covering the optic disc produced traction on the peripapillary retina. The next day, during vitrectomy surgery, the retinal vessels surrounding the morning glory disc anomaly appeared to be pulled and retracted posteriorly into the central area of the optic disc by a pigmented membrane. The macula was displaced toward the edge of the optic disc by a membrane from the surface of the optic disc, and a slit-like retinal hole was observed in tissue lying within the optic cup. The retinal hole provided a pathway for fluid between the vitreous cavity and subretinal space. The retina flattened following an air-fluid exchange with internal drainage of the subretinal fluid over the optic disc. A drop of thrombin (100 U/ml) was combined with two drops of the patient's autologous plasma to form a clot over the retinal opening in the optic disc. A fluid-air exchange was performed, and the retina was flattened following internal drainage of subretinal fluid over the optic disc. The retinal detachment usually involves the peripapillary retina and the posterior pole. In some cases spontaneous resolution of the retinal detachment has been observed [5]. In others, pigmentary changes in the retinal pigment epithelium suggested spontaneous reabsorption of subretinal fluid. This clinical observation suggests that the formation of subretinal fluid in the morning glory syndrome may be related to a defect in the optic disc. Furthermore, the histologic features of the morning glory syndrome are similar to that of optic nerve pits which are associated with retinal detachments.

Several pathogenic mechanisms have been proposed to explain how retinal detachments are commonly associated with the morning glory syndrome. Rhegmatogenous retinal detachments have been reported in a limited number of cases. In these cases retinal tears in the fovea and in the peripapillary retina have been responsible for the retinal detachments [8–10]. Possibly the peripapillary retinal tears are the result of traction from peripapillary fibroglial tissue [3]. Chang et al. demonstrated an apparent communication between the subarachnoid and subretinal spaces when metrizamide dye injected into the subarachnoid space appeared radiographically in the subretinal space [11]. Reattachment of the retina was observed after optic nerve sheath fenestration.

Discussion

Cogan described the histopathologic features in one patient with the morning glory disc anomaly. Within the optic cup there was a sheet of vascularized connective tissue, distortion of the peripapillary retina as a result of traction from an epipapillary connective tissue membrane, an enlarged scleral canal with an absent lamina cribrosa, and proliferation of glial and retinal pigment epithelial cells surrounding the optic disc [7]. The histopathologic features of the morning glory disc anomaly are similar to that of optic nerve pits, except that eyes with the morning glory syndrome have more extensive scleral defect with staphylomatous herniation, and retinal tissue that extends posteriorly surrounding the entire margin of the optic disc.

In most of the retinal detachments associated with the morning glory disc syndrome retinal breaks have not been found. The retinal detachment usually involves the peripapillary retina and the posterior pole. In some cases spontaneous resolution of the retinal detachment has been observed [5]. In others, pigmentary changes in the retinal pigment epithelium suggested spontaneous reabsorption of subretinal fluid. This clinical observation suggests that the formation of subretinal fluid in the morning glory syndrome may be related to a defect in the optic disc. Furthermore, the histologic features of the morning glory syndrome are similar to optic nerve pits which are associated with retinal detachments.

Seventeen intraocular operations for uncontrolled glaucoma and cataract surgery with implantation of an intraocular lens.

On 18 May 1993 bubbles of silicone oil migrated through the retinal hole into the subretinal space of the macula (arrowhead). The pigmented membrane covering the optic disc was removed.

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