

Case Report

Case Report: Fibroglial Retinal Tissue in Contractile Morning Glory Disc Anomaly

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Keywords

Contractile morning glory syndrome · Retinal detachment · Tractional retinal detachment · Congenital optic disc anomaly · Contractile · Disc anomaly

Abstract

The purpose of the present case is to describe a patient with tractional retinal detachment (RD) associated with contractile morning glory: a 17-year-old female, with a history of failed surgery for RD when she was 2 years old in her right eye (OD), nystagmus, and a limited visual acuity in the left eye (OS). The slit lamp examination showed phthisis bulbi in OD and the anterior segment was unremarkable in OS. Dilated fundus examination revealed a tractional RD in the posterior pole and peripapillary and preretinal fibrosis without evidence of intravitreal dispersion of retinal pigment epithelial cells. After surgery treatment, the RD resolved and the posterior segment showed a staphylomatous excavation around the optic disc anomaly with irregular contractions that folded the macular area. This were unrelated to light, breathing, or eye movements. Although morning glory disc anomaly is associated with RD, the early diagnosis can reverse structural changes. In this case, the rare association with contractile movements was found posterior to the pars plana vitrectomy after all the fibroglial epiretinal tissue was removed.

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Introduction

The morning glory syndrome (MGS) or morning glory disc anomaly (MGDA) was described by Kindler [1]. The syndrome is characterized by peripapillary staphyloma with axial retrodisplacement of optic nerve, absence of lamina cribosa, an aberrant radial configuration of

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retinal vessel at the edge of the optic cup, and chorioretinal pigmentary changes that reminds of the morning glory flower [2]. It is a rare disease and no gender predisposition has been found [2].

The development of retinal detachment (RD) in patients with MGDA is well recognized [3]. The different pathogenesis theories support all types of RD in MGDA: exudative, tractional, and rhegmatogenous [4].

Different mechanisms have been described in the pathogenesis of RD in MGDA, for example, the communication of the subretinal space with the vitreous cavity, subarachnoid space, or orbital tissues. Based on spectral domain-optical coherence tomography, it has been reported that tractional force of the vitreous is the major cause of the RD in these particular cases of MGDA. Primary tractional forces of the vitreous can induce tractional RD and further formation of the tiny retinal break at the weakest area of the retinal tissue within excavated optic disc [4]. In this report, we describe the follow-up of a patient with tractional RD with important fibrous tissue in a patient with contractile MGDA.

Case Report

A 17-year-old female presented with a history of failed surgery for RD when she was 2 years old in her right eye (OD), nystagmus, and a limited visual acuity in the left eye, and two weeks prior, a sudden decrease in central vision in the left eye (OS). At her initial visit, the best-corrected visual acuity was 20/1,200 in her OS and no light perception in her OD. The slit lamp examination showed phthisis bulbi in OD, and anterior segment was unremarkable in OS. Dilated fundus examination revealed a tractional RD in the posterior pole and peripapillary and a preretinal fibrosis without evidence of intravitreal dispersion of retinal pigment epithelial cells. Surgical treatment with pars plana vitrectomy was proposed.

Surgery was under local anesthetic. A standard pars plana vitrectomy equipment (Constellation, Alcon Surgical, Fort Worth, TX, USA), M822 microscope (Leica Microsystems Schweiz, AG) and 3D visualization system (TrueVision System). A 3-port, 25-gauge pars plana vitrectomy and core vitrectomy were performed. Creation of posterior vitreous detachment, vitreous base shave, and meticulous peeling maneuvers with standard 25-gauge forceps in preretinal membranes were performed (Fig. 1).

A fluid-air exchange was completed and subretinal fluid was not extracted because we did not find regmatogenous lesion. Silicone oil (5,000 centistokes) was used as the tamponade to finish the surgery.

One day after the surgery, the RD resolved (Fig. 2). On the posterior segment of the OS, it showed staphylomatous excavation around the optic disc anomaly with irregular contractions that involved and folded the macular area.

One month after the surgery, best-corrected visual acuity improved to 20/200 in the OS and irregular contractions have been kept unrelated to light, breathing, or eye movements (online suppl. Video 1 available at www.karger.com/doi/10.1159/000510958).

Discussion

The MGDA is generally a unilateral sporadic congenital abnormality of the optic disk. The embryologic basis is unclear. It may be the result of a defect in the formation of the posterior sclera with secondary herniation of neural tissue and interference with resorption of primary hyaloidal elements [3, 5].

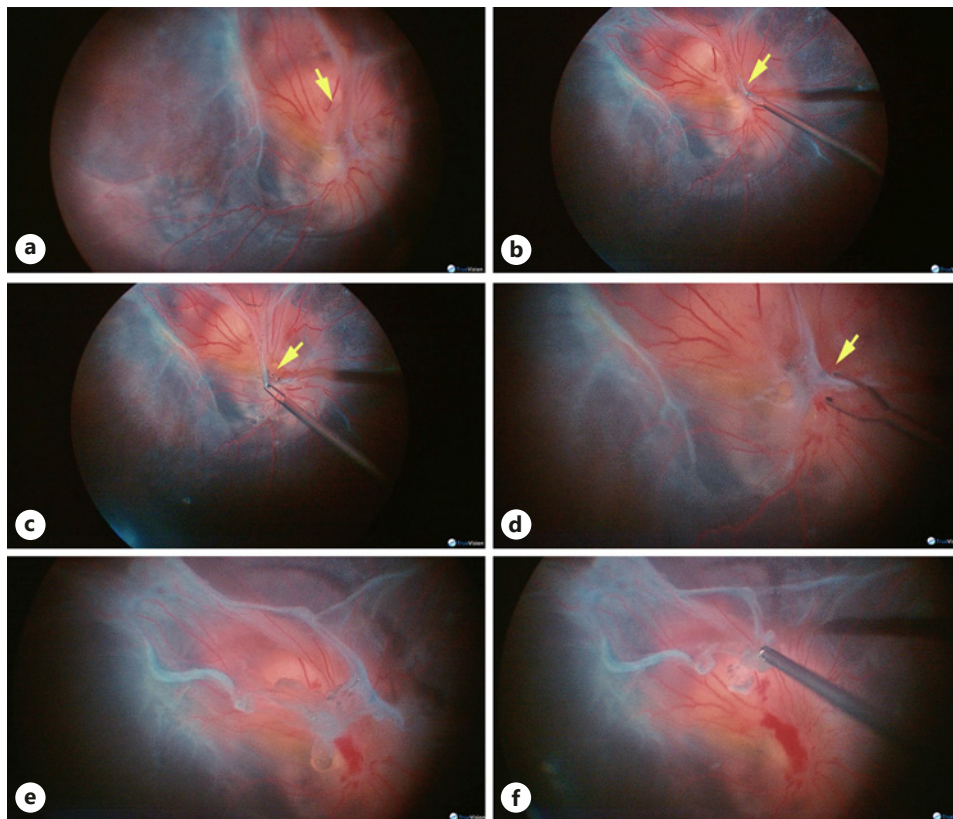


Fig. 1. Intraoperative photography fundus of the OS. **a** Peripapillary subretinal fluid is observed (yellow arrow). **b–d** The fibroglial tissue over the posterior pole is removed with forceps from the papilla. **e, f** Once released, the tissue is removed from the posterior pole and withdrawn with the vitrector.

Identifying MGDA is important because of its association with RD involving the posterior pole or peripapillary retina [6, 7]. It is suggested that the formation of subretinal fluid in the MGS may be related with a defect in the optic disc, similar to optic nerve pits [8], with the presence of an abnormal communication between the vitreous cavity, subretinal space, and subarachnoid space [9]. It has been demonstrated a communication between the vitreous cavity and the perineural space [10], vitreous cavity and subretinal space through the retinal opening in the optic cup [8], and between subarachnoid space and subretinal space [11]. Another theory is where retinal tears in the fovea and in the peripapillary retina have been responsible for the RDs. Possibly, the peripapillary retinal tears are the result of traction from peripapillary fibroglial tissue [8, 12]. Lytvynchuk et al. [4] described a case with MGS and RD. They evaluated the tissues with an intraoperative optical coherence tomography, and the analysis revealed a strong adhesion between posterior condensed vitreous and peripapillary area and macula. Primary tractional forces of the vitreous can induce tractional RD and further formation of the retinal break at the weakest area of the overstretched retinal tissue within excavated optic disc [4]. Zhang and colleagues [12] reported 8 cases of proliferative RD associated with macular hole in young patients with MGS. It has been reported that the most common location for the retinal breaks is in the retinal tissue within anomalous and retracted optic disc. In our case, the fibroglial tissue that was observed during the surgery may have caused a very small retinal break in the peripapillary retina, but the clinical examination, pre- and intraoperative, failed to reveal the presence of a retinal break. After the removal of the fibroglial tissue, the retina reattached without an internal drainage. It is recom-

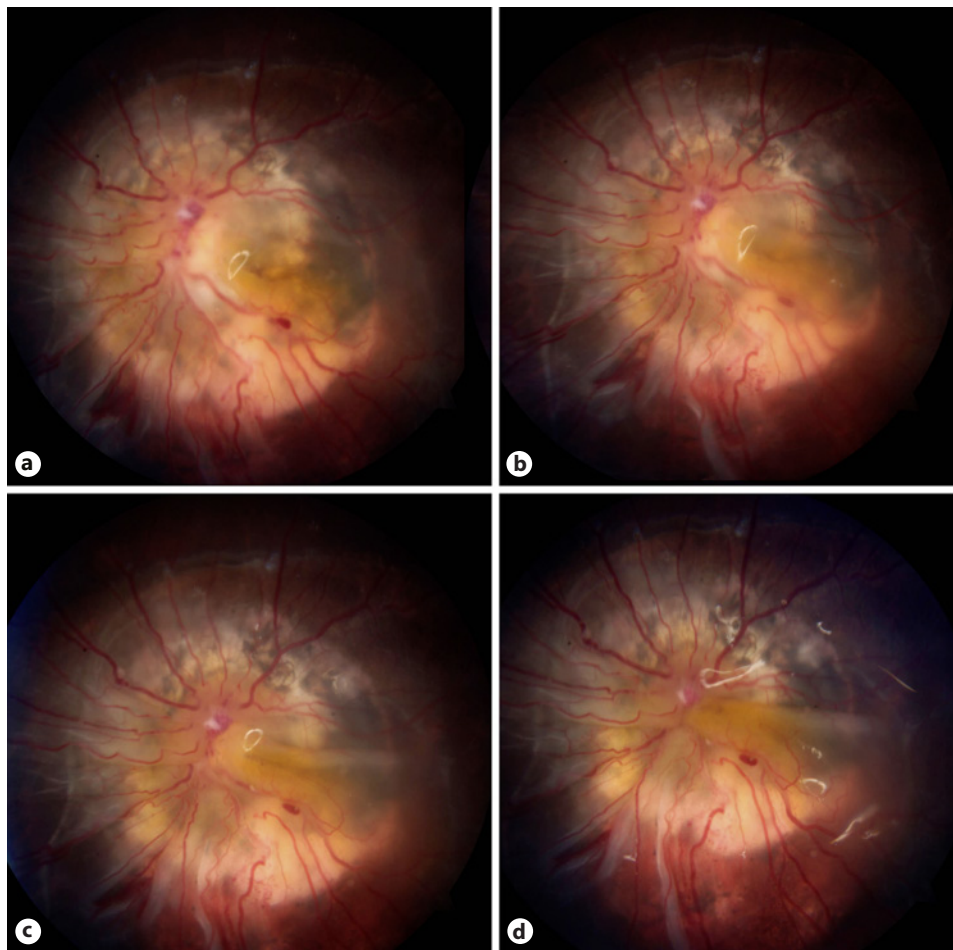


Fig. 2. Posterior pole photographs. **a** MGDA is observed; vascular tortuosity and retina attached in the posterior pole. **b, c** Contractile movements at the level of the papilla that cause folding of the retina. **d** The configuration of the lesion changed remarkably with the contraction, into prominent radial folds.

mended an early PPV since the subretinal fluid of these patients has been related with a poor visual prognosis.

It has been reported in histopathology studies a mass of connective tissue occupying the physiological cup possibly derived from the primitive hyaloid system [13] and the association with congenital ocular anomalies like persistent hyperplastic primary vitreous, suggesting that the regression of the hyaloidal vasculature is easily compromised in the presence of optic disc defects, but in this case the morphology and the tissue response during surgical management was not characteristic of persistent hyperplastic primary vitreous. Another theory of the origin of this tissue is that the RD originated a proliferative vitreoretinopathy, but without the clinical evidence of regmatogenous lesion or cells of the pigment epithelium in the vitreous cavity, it cannot be supported. In the literature review that we have done, we did not find the characteristics of this tissue in MGDA; therefore, the origin of this particular tissue remains unclear.

The contractile movements from the optical disc are a rare association that have been documented in cases of MGDA and peripapillary staphyloma [14]. The mechanism of the contractions remains unclear [9, 14–16]. The main theories of the contractile movements are classified into 2 groups: pressure balance and muscular contraction [9]. An alternative pressure balance mechanism hypothesis proposes that there is an anomalous communication between the subarachnoid space and the juxtapapillary subretinal space. Changes in

transient pressure gradients occur between the 2 compartments, causing the flow of fluid back and forth along the optic nerve causing the contraction and expansion [9, 15]. The pressure balance mechanism includes peripapillary subretinal fluid and the contraction related to the respiratory cycle, Valsalva maneuver, forced eye closure, or intraocular pressure changes [17].

The muscle contraction mechanism has been proposed by several authors [15, 17–19]. The presence of nonvascular contractile smooth muscle cells in the posterior choroid and sclera of normal eyes and heterotopic smooth muscle in optic disc colobomas has been reported [18, 20]. Sawada and colleagues [15] reported, as a triggering factor, the strong light stimulus to the fellow eye, which is similar to the reaction of the ciliary muscle and sphincter iridis muscle; nevertheless, it did not happen in the affected eye. The contraction mechanism provoked by consensual reflex is supported by other authors [17, 19, 21]. Wise reported a case of contractile peripapillary staphyloma and proposed an atavistic retractor bulbi muscle since the case presented the contractile movements induced by forced lid closure [19]. Lee and colleagues [22] demonstrated contractile movement mainly around the optic nerve margin in a horizontal direction in computerized analysis and support smooth muscle contraction for the pathogenesis of the contractile movement. A remarkable feature in this case are the contractile movements that were evident after the surgery when the fibroglial tissue was removed. However, the muscular contraction theory seems to be more suitable in the present case. The configuration of the lesion changed markedly; the detached portion of the retina was thrown into prominent radial folds and none of the conditions under which fundus contractile movements were registered produced an alternating pressure gradient between the vitreous cavity and the subretinal space. It was not related with the respiratory cycle, Valsalva maneuver, forced eye closure, light exposure, intraocular pressure changes, and also during the surgery, the contractile movements were not present. Therefore, we conclude that the presence of smooth muscle is the origin of the contractile movements in this case.

Conclusion

The highlight of this case is the tractional RD associated with the MGDA and the contraction of the epiretinal tissue causing the RD. Before the surgical procedure and with the tissue's contraction, subretinal fluid was visible in the peripapillary area. However, after the PPV a contractile movement of the MGDA was observed. We can assume that the contractile movements were not related with respiratory cycle, intraocular pressure changes, ocular movements, or light exposure.

Statement of Ethics

The case report has been approved by the IRB and has received ethical approval. The case report and all conducted studies adhere to the Declaration of Helsinki. Informed written consent was obtained from the patient for publication of this case report.

Conflict of Interest Statement

The authors declare that they have no conflicts of interest regarding the publication of this case report.

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Author Contributions

Ramirez-Estudillo, Abel, MD: conception, design, drafting, analysis, and interpretation of the work. Torres-Navarro, Karla, MD: analysis, drafting, and interpretation of the work. Rojas-Juárez, Sergio, MD: revising it critically for important intellectual content. Ramirez-Galicia Ximena, MD: drafting and analysis of the work. Palafox-Cornejo Berenice, OD: acquisition and analysis of data for the work. Galicia-Castillo Adriana, MD: final approval of the version to be published. All authors approve the final version of the manuscript for publication.

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