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"MORNING GLORY" SİNDROMUNDA REQMATOGEN TOR QIŞA QOPMASININ VİTREORETİNAL CƏRRAHİYYƏSİ (KLİNİK HAL)

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XÜLASƏ

"Morning glory" sindromu (MGS) qıfvari görmə siniri diski, mərkəzi qlial toxuma və peripapilyar piqmentasiya ilə xarakterizə olunan nadir, anadangəlmə görmə siniri diski anomaliyasıdır. Çox vaxt birtərəfli olan bu sindrom yüksək miopiya, ambliopiya və inkişaf qüsurları ilə əlaqələndirilir, rastgəlmə tezliyi təxminən 100000 nəfərə 3,6 hal təşkil edir. Tor qişanın qopması bu sindromun əsas ağırlaşmalarından biridir və halların 26 - 40%-də rast gəlinir, daha çox seroz xarakter daşısa da, traksion və ya reqmatogen formada da baş verə bilər.

Məqsəd – tor və damarlı qişaların qopması ilə ağırlaşmış MGS olan xəstədə vitreoretinal cərrahiyyənin nəticələrini təqdim etmək.
Uşaqlıqdan yüksək miopiya (OD: (–)15 D; OS: (–)10 D) və ambliopiya anamnezi olan 35 yaşlı kişi son 2 ay ərzində sağ gözdə kəskin görmə itkisi (maksimal korreksiya olunmuş görmə itiliyi (MKGİ): 20/2000) şikayəti ilə müraciət etmişdir. Anamnezdə hər iki gözdə skleroplastika əməliyyatı, sol gözdə isə fakik linza implantasiyası və lazer fotokoaqulyasiyası icra olunduğu qeyd edilmişdir. B-skan ultrasəs müayinəsi zamanı reqmatogen tor qişa və damarlı qişanın qopması aşkar olunmuşdur. Xəstəyə saat 11 meridianında optik disk kənarı yırtığı ətrafında endolazer fotokoaqulyasiya, subretinal mayenin aspirasiyası və silikon yağı (5700 cSt) tamponadası ilə 25 ölçülü vitrektomiya əməliyyatı icra edilmişdir. İki ay sonra silikon yağının çıxarılması, İOL implantasiyası ilə kataraktanın fakoemulsifikasiyası və qaz-hava tamponadası əməliyyatları həyata keçirilmişdir. Silikon yağı çıxarıldıqdan 6 ay sonra tor qişanın tam bitişməsi müşahidə olunmuş, sağ gözün MKGİ göstəricisi 20/100-ə qədər artmışdır.

Yekun
Tor qişanın qopması ilə müşayiət olunan MGS arxa şüşəvari cismin qopmaması, sıx fibroz toxuma və uzun aksial oxunun olması kimi cərrahi çətinliklər yaradır. Erkən oftalmoloji diaqnostika və vaxtında aparılan cərrahi müalicə əməliyyatdan sonrakı dövrdə uğurlu anatomik nəticənin əldə olunmasına imkan verir.

Açar sözlər: *tor qişanın qopması, "morning glory" sindromu, vitreoretinal cərrahiyyə*

Müəlliflər münaqişələrin (maliyyə, şəxsi, peşəkar və digər maraqları) olmamasını təsdiqləyirlər

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VITREORETINAL SURGERY FOR RHEGMATOGENOUS RETINAL DETACHMENT IN A PATIENT WITH MORNING GLORY SYNDROME: A CASE REPORT (CLINICAL CASES)

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SUMMARY

Morning glory syndrome (MGS) is a rare congenital optic disc anomaly characterized by a funnel-shaped disc, central glial tissue, and peripapillary pigmentation. It is often unilateral, associated with high myopia, amblyopia, and developmental malformations, with a prevalence of approximately 3.6 per 100,000. Retinal detachment, occurring in 26-40% of cases, is a major complication, predominantly serous but also tractional or rhegmatogenous.

Purpose – to present the results of vitreoretinal surgery in a patient with MGS complicated by retinal detachment and choroidal detachment.
A 35-year-old male patient with a history of high myopia since childhood (OD: (–)15 D; OS: (–)10 D) and amblyopia presented with acute vision loss in the right eye Best-Corrected Visual Acuity (BCVA): 20/2000) over 2 months. History included bilateral scleroplasty and left-eye phakic lens implantation with laser photocoagulation. B-scan ultrasonography revealed rhegmatogenous retinal and choroidal detachment. The patient underwent 25-gauge vitrectomy with endolaser photocoagulation around an 11 o'clock disc-margin break, subretinal fluid aspiration, and silicone oil (5700 cSt) tamponade. Two months later, silicone oil removal, cataract phacoemulsification with intraocular lens (IOL) implantation, and gas–air tamponade was performed. At 6 months after silicone oil removal, the retina was reattached, with OD BCVA improving to 20/100.

Conclusion
Morning glory syndrome with retinal detachment poses surgical challenges, including absent posterior vitreous detachment (PVD), dense fibrotic tissue, and long axial length. Early ophthalmological diagnosis and timely surgical treatment allow a good anatomical outcome in the postoperative period.

Key words: *retinal detachment, morning glory syndrome, vitreoretinal surgery*

Morning Glory Syndrome, also known as morning glory disc anomaly, is a rare congenital anomaly of the optic disc with unclear etiology, first described by P. Kindler in 1970. Ophthalmoscopically, it is characterized by an enlarged, funnel-shaped optic disc surrounded by a ring of pigmentation and central glial tissue. The condition is typically unilateral and frequently associated with high myopia, amblyopia, and combined developmental anomalies (occasionally with intracranial malformations). The estimated prevalence is approximately 3.6 per 100,000 individuals. It affects females approximately twice as often as males, is usually unilateral, and involves the right eye in about 60% of cases [1 – 4]. Bilateral forms are rare and may have a hereditary component. The pathogenesis is thought to involve defective closure of the fetal fissure, mesenchymal abnormalities, persistence of the hyaloid system, or mutations in PAX6/PAX2 genes. MGS is often associated with basal encephalocele, cerebral vascular anomalies, renal malformations, facial skeletal abnormalities, as well as PHACE, CHARGE, and Aicardi syndromes [3]. Visual acuity is frequently poor from early childhood (e.g., due to high myopia and amblyopia), as observed in the present case as well. Retinal detachment represents the major complication in patients with MGS.

Retinal detachment develops in 26 - 40% of patients (various sources report 30–38%). Different types of retinal detachment may occur:

- Serous (exudative) — the most common type. Fluid enters the subretinal space from the subarachnoid space through defects in the optic nerve sheaths [5]. In the study by G.Cennamo et al. [6], retinal detachment was detected in 5 out of 9 eyes with MGS despite the absence of visible retinal breaks.
- Tractional — due to glial tissue traction, disc contraction, and vitreoretinal traction.
- Rhegmatogenous — less frequent, occurring in the presence of retinal breaks at the optic disc margin. The mechanism involves abnormal communication between the vitreous cavity, subarachnoid space, and subretinal space, combined with traction and possible choroidal exudation.

Only a limited number of publications in the literature address the management and surgical strategies for patients with this condition.

Purpose – to present the results of vitreoretinal surgery in a patient with MGS complicated by retinal detachment and choroidal detachment.

Case Presentation

pA 35-year-old patient with no chronic systemic diseases presented with a sudden, severe decrease in vision in the right eye over the preceding 2 months. He had a history of high myopia in both eyes since childhood (right eye: sphere (–)15 D; left eye: sphere (–)10 D), high-degree amblyopia since childhood, and BCVA was 20/100.

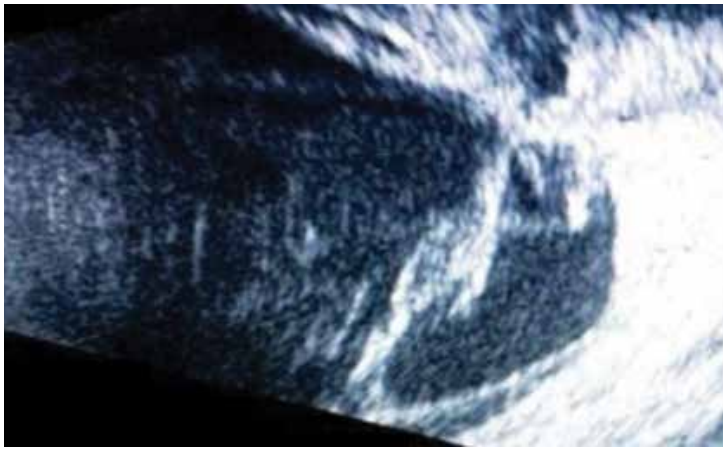

Figure 1. *Preoperative ultrasound scan of the patient's right eye: retinal and choroidal detachment.*

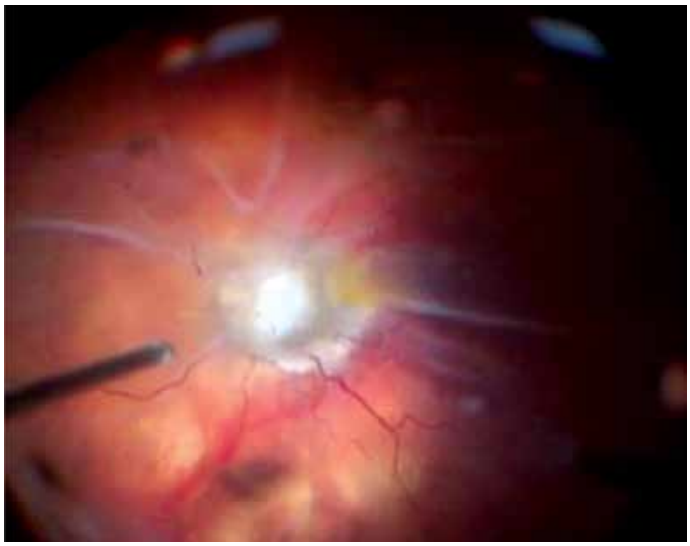

Figure 2. *Intraoperative fundus photography: dense, poorly mobile vitreous, absence of PVD, and dense fibrotic tissue at the optic disc.*

Scleroplasty had been performed on both eyes in childhood. No ocular trauma was reported. No regular topical medications were used. In 2015, phakic intraocular lens implantation and prophylactic barrier laser photocoagulation of the retina was performed in the left eye (OS). On admission, BCVA in right eye was 20/2000 and 20/20 in left eye. Intraocular pressure (IOP) was 5 mm Hg OD preoperatively.

Preoperative examination revealed initial cataract, dense and poorly mobile vitreous body without PVD, and fundus changes consistent with MGS, including total rhegmatogenous retinal detachment with macular involvement (macula-off) and choroidal detachment.

In the evaluation of congenital optic disc anomalies, including MGS, B-scan ultrasonography and optical coherence tomography (OCT) are widely used. Gray-scale B-scan ultrasonography in this patient revealed retinal and choroidal detachment. Notably, according to G.Cennamo et al., B-scan ultrasonography proved more informative than other methods for assessing optic nerve integrity (**Figure 1**). The patient did not undergo OCT before surgery, as the retinal detachment was total bullous and the image slices were not informative.

The surgical interventions were performed at the Pirogov National Medical and Surgical Center, Ophthalmology Center. The patient underwent standard 25-gauge three-port pars plana vitrectomy. Intraoperative examination of the vitreous cavity revealed dense, poorly mobile vitreous, absence of PVD, and dense fibrotic tissue at the optic disc (**Figure 2**). A retinal break was identified at the 11 o'clock position relative to the disc. Triamcinolone solution was used for vitreous visualization. PVD was induced by aspiration. To stabilize the retina, temporary perfluorodecalin (PFCL) tamponade was used; PFCL was introduced up to the level of the break to prevent its migration under the retina, followed by exchange to air and subsequent aspiration of subretinal fluid. Aspiration of thick subretinal fluid was performed through the break margins, followed by endo-laser photocoagulation around the break (spot size 200-300 μm, duration 0.1-0.2 s, power 150-400 mW titrated to achieve mild white burns, 2-3 rows of confluent or overlapping spots), removal of the perfluorocarbon liquid, exchange of BSS for sterile air, and final tamponade of the vitreous cavity with 5700 cSt silicone oil. The second stage of surgery (performed 2 months later) consisted of silicone oil removal, phacoemulsification of

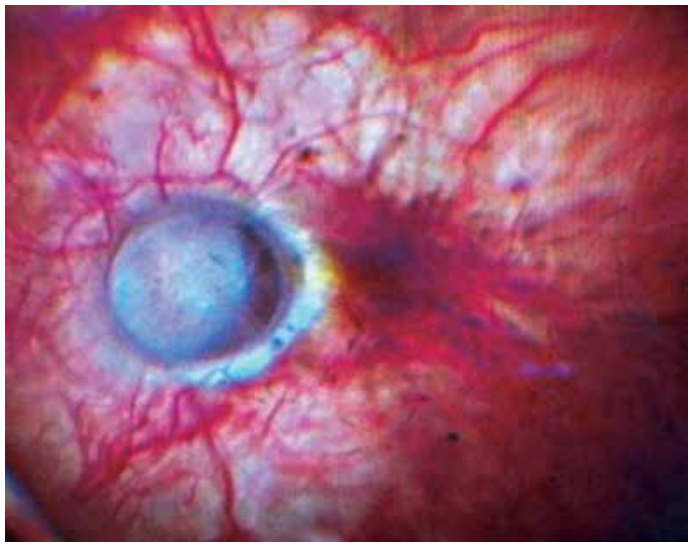

Figure 3. *Postoperative photography of posterior pole of the eye.*

cataract with intraocular lens implantation, and gas–air tamponade.

In the postoperative period, the patient was instructed to maintain a face-down position. Such a two-stage approach represents one of the recommended strategies: vitrectomy with creation of chorioretinal adhesions via laser photocoagulation around the affected area, combined with prolonged tamponade (using silicone oil or gas–air mixture), has enabled retinal reattachment in many reported cases. In the presented case, following tamponade removal, the retina remained attached (**Figures 3 and 4**), with the macula reattached postoperatively. BCVA in the right eye improved to 20/100 and IOP was 15 mm Hg OD. The follow-up period was 6 months after silicone oil removal. It should be emphasized that final visual acuity is often limited by the underlying amblyopia and high myopia, despite successful surgery and achievement of favorable anatomical outcomes. Additional preoperative OCT images demonstrating the retinal break are not available; only one postoperative OCT is presented, taken 6 months after tamponade removal (**Figure 4**). For rare pathologies like this, longer follow-up (up to 12-24 months) is ideal to assess risks

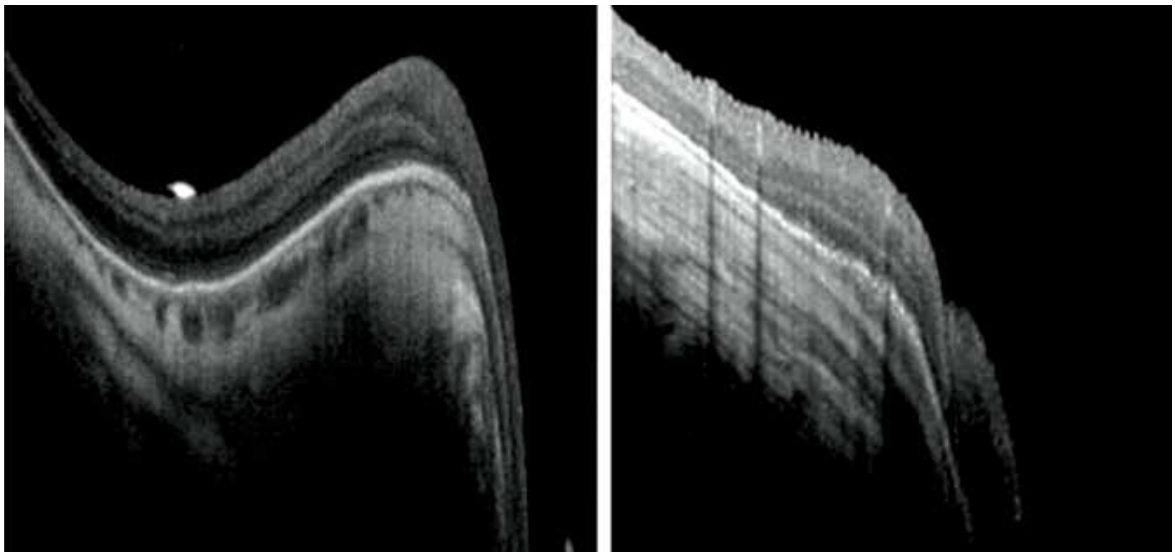

Figure 4. *Postoperative optical coherence tomography six months after surgery.*

such as proliferative vitreoretinopathy (PVR) in 20–30% of cases, but current observation confirms stable reattachment.

Results

Morning glory syndrome itself is extremely rare, and its combination with retinal and choroidal detachment is even more exceptional. This accounts for the limited number of clinical series and the absence of standardized treatment protocols. The literature describes various surgical techniques, including vitrectomy with retinal laser photocoagulation, drainage of subretinal fluid, transpupillary laser photocoagulation, and even defect closure using placental grafts [2, 7 – 9].

Surgical challenges encountered during vitreoretinal surgery in patients with MGS include:

1. Young patient age, which is frequently associated with the absence of PVD;
2. The need for vitreous staining to facilitate complete removal of vitreous structures;
3. Dense adherences of fibrotic tissue in the region of the optic disc;
4. In the presence of a retinal break at the optic disc margin, there is a risk of perfluorocarbon liquid migration into the subretinal space;
5. Increased axial length of the eye, which in some cases necessitates the use of additional instruments designed for highly myopic patients.

Virtually all authors underscore the necessity of an individualized approach and the simultaneous application of multiple surgical techniques. Our experience confirms the efficacy of vitrectomy combined with endolaser photocoagulation and tamponade using silicone oil or a gas–air mixture. This technique resulted in a favorable anatomical and functional outcome.

Conclusion

In conclusion, MGS represents a serious ophthalmological challenge: patients present with inherently low visual acuity, a high risk of retinal and choroidal detachment, and technically demanding treatment. Modern vitreoretinal surgical methods can achieve positive results; however, the visual prognosis remains limited by the underlying optic disc dysfunction. Further accumulation of clinical experience and publication of similar cases are essential for developing optimal treatment strategies. Additional studies will help standardize approaches and improve outcomes for these patients.

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