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Surgical approach to pediatric optic disc pit maculopathy: a case report

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Purpose: *To evaluate postoperative results of pars plana vitrectomy combined with an inverted internal limiting membrane-flap technique and intravitreal injection of viscoelastic material for optic disc pit maculopathy complicated by serous macular detachment in a child.*

Observations: *Ocular examination included best-corrected visual acuity (BCVA) testing, slit-lamp biomicroscopy, dilated fundus examination, intraocular pressure measurement, optical coherence tomography (OCT) and color fundus photography. BCVA in the left eye was 20/50 (0.4). Preoperative OCT findings showed distortion of retinal layers, serous macular detachment and a large schisis cavity in the left eye. Foveolar depression was not determined due to the height of intraretinal fluid and subretinal fluid extending towards the optic disc pit. Retinal thickness in the macular area was 507 μm.*

Pars plana vitrectomy was performed in combination with an inverted internal limiting membrane internal limiting membrane-flap technique and intravitreal injection of viscoelastic material, followed by 15% C3F8 gas endotamponade.

A follow-up OCT examination in 3 months showed decreased subretinal fluid, residual edema, and restored foveolar depression. Retinal thickness in the macular area was 328 μm. BCVA of the left eye improved to 20/32 (0.63).

Conclusions: *Pars plana vitrectomy with an inverted internal limiting membrane-flap technique for optic disc pit maculopathy allows to reduce an amount of intra- and subretinal fluid. A visco-associated flap fixation technique creates conditions for its stabilization, which ultimately contributes to improving anatomical and functional outcomes during surgery and in the postoperative period.*

Key words:

optic disc pit, pars plana vitrectomy, retina

Introduction. Optic disc pit (ODP) is a rare anomaly that is characterized by a round or oval-shaped depression of gray, yellow or black color with a typical inferotemporal location. ODP was first described by Wiethe in 1882 in a 62-year-old woman [1].

The prevalence of ODPs is 1 in 11,000 patients, affecting men and women with equal frequency [2]. The pathology is mainly unilateral, but may be bilateral in 10-15% of patients. The etiology of the optic disc pits remains unclear. There are congenital and acquired ODPs associated with trauma, glaucoma, and pathologic myopia. The disease can be asymptomatic for a long time; however, the optic disc pit can be complicated by a decrease in visual acuity as serous macular detachment develops within the third to fourth decades of life in 25-75% of cases [3].

Pathogenesis of the ODP formation remains open. ODP can result from incomplete closure of fetal fissures affecting the lamina cribrosa, mainly in the first trimester of embryogenesis [4]. The origin of congenital pits of the optic nerve head (ONH) is associated with autosomal dominant inheritance according to the theory of heredity [5]. Several different sources have been proposed to explain the mechanism of subretinal fluid accumulation: the vitreous cavity, cerebrospinal fluid arising from the subarachnoid space, leakage from blood vessels at the base of the pit, or leakage from the choroid [6]. Some authors consider increased intracranial pressure as a starting mechanism in the development of serous macular detachment.

The purpose of this study was to evaluate the postoperative results of pars plana vitrectomy combined with an inverted ILM-flap technique and intravitreal injection of viscoelastic material for optic disc pit maculopathy (ODP-M) complicated by serous macular detachment.

Case Report

An 11-year-old girl presented with a three-month history of decreased vision in her left eye. There was no previous history of eye diseases.

The patient underwent an eye examination, including best-corrected visual acuity (BCVA) testing, slit-lamp biomicroscopy, dilated fundus examination, intraocular pressure (IOP) measurement, spectral-domain optical coherence tomography (SD-OCT) and color fundus photography (Figure 1, 2). BCVA in the left eye was 20/50 (0.4). Preoperative OCT findings showed distortion of retinal layers, serous macular detachment and a large schisis cavity in the left eye. Foveolar depression was not determined due to the height of intraretinal fluid (IRF) and subretinal fluid (SRF) extending towards the ODP. Retinal thickness in the macular area was 507 μm.

In this case, pars plana vitrectomy (PPV) was performed in combination with an inverted internal limiting membrane (ILM)-flap technique and intravitreal injection of viscoelastic material, followed by 15% C_3F_8 gas endot-

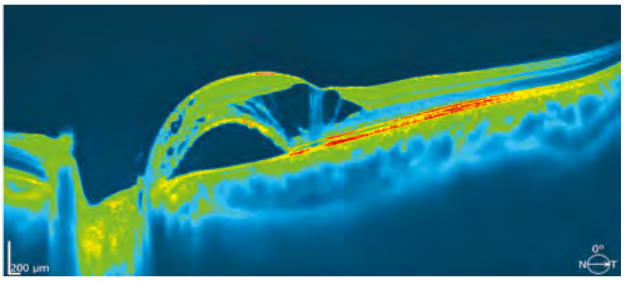

Figure 1. Preoperative OCT shows serous macular detachment and large schisis cavity in the left eye of the patient.

amponade in order to prevent subretinal fluid migration and subsequent detachment of macula (Figure 3). Written informed consent was obtained from the parents.

The surgery was performed under general anesthesia using the Constellation Vision System® (Alcon Inc., Geneva, Switzerland), under the control of Topcon OMS-800 OFFISS operation microscope with a BIOM for non-contact wide-angle viewing system (10 000 cuts/minute (cpm); aspiration, 200 mmHg; infusion pressure, 30 mmHg). Standard 3-port, 25-gauge PPV was performed, followed by an incomplete posterior vitreous detachment (PVD) by active aspiration with the vitreous cutter over the optic disc. A dye solution (TWIN, AL.CHI.MI.A S.r. L., Italy) was injected intravitreally with a stream directed toward the posterior pole of the eye. The dye was removed from the vitreous cavity, and an inverted ILM-flap technique was performed. The technique consists of partial peeling of the ILM at a distance of about 3 mm from the ONH, preserving its attachment to the temporal margin of the optic disc, and placing the flap over the optic disc pit. (Figure 3 B, C, D). Viscoelastic material (CELLUGELтм, Alcon Laboratories, Inc., Fort Worth, TX, USA) was injected in the vitreous cavity (Figure 3 E) prior to fluid-air exchange to prevent ILM-flap displacement. Fluid-air exchange was performed, followed by an injection of 15% perfluoropropane (C_3F_8). The patient was instructed to maintain a face-down position for two weeks after surgery.

Based on the anatomical (subretinal fluid resorption, macular reattachment) and functional (BCVA) data, as well as the presence of postoperative complications, treatment efficacy was evaluated. The follow-up period was three months.

There were no complications during the surgery and the early postoperative period. The ILM flap was formed according to the above procedure and fixed with a viscoelastic material, which prevented from its displacement

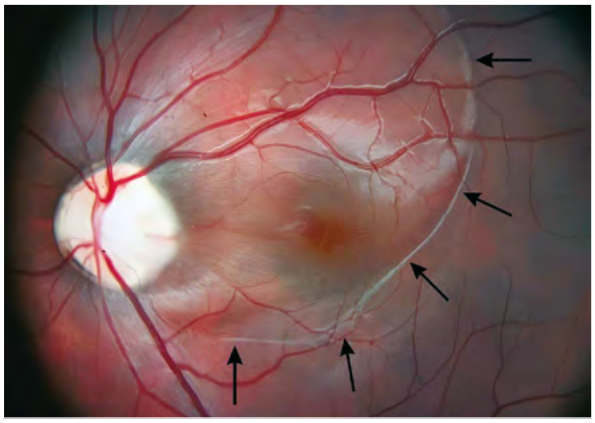

Figure 2. Preoperative color fundus photo of the left eye. Arrows show boundaries of serous macular detachment.

during the fluid-air exchange. Intravitreal gas injection of 15% perfluoropropane was performed to achieve macular reattachment. The volume of the intraocular gas bubble was 85% and 70% in the early postoperative period and at the time of discharge, respectively; IOP was 15-16 mmHg.

A follow-up OCT examination in 3 months showed decreased subretinal fluid, residual edema, and restored foveolar depression (Figure 4). Retinal thickness in the macular area was 328 µm. BCVA of the left eye improved to 20/32 (0.63).

In addition, OCT scans determined a significant reduction in central macular thickness in all sectors (Table 1).

Discussion

Our study describes a case of surgical treatment of ODP-M in an 11-year-old girl. ODP was congenital with localization along the temporal margin of the optic disc, which confirms the observation data that congenital optic disk pits involve the temporal part while the acquired ones develop mainly at the upper or lower pole of the optic disc [7]. There was no family ocular history.

Single cases of treatment of ODP-M are controversial because there is no consensus on surgical intervention or the methods of surgery, especially in children. The main treatment approaches are conservative methods and surgical interventions. PPV is one of the most common techniques of surgical treatment for ODP.

Conservative treatment, including dehydration and corticosteroid therapy, is not widespread because it has a temporary result. Laser therapy in combination with intravitreal injection of expansile gas in a non-vitrectomized

Table 1. Central macular thickness

Term	Central macular thickness, µm				
	CS	SS	IN	TS	NS
Before surgery	507	474	513	348	615
At 3 months postop	328	374	391	317	442

Note. CS – central sector, SS – superior sector, IN – inferior sector, TS – temporal sector, NS – nasal sector.

eye has a curative effect compared to intravitreal gas injection as monotherapy, 72% vs 50%, respectively [8, 9]. Xenon photocoagulation along the temporal margin of the optic disc was described by J.D. Gass in 1969 [10]. Laser therapy provides reliable chorioretinal adhesion that will act as a barrier between the ODP and subretinal space, preventing further serous macular detachment. However, the widespread use of juxtapapillary laser photocoagulation remains controversial due to the risk of laser-induced damage to the nerve fiber layer in the region of the papillomacular bundle. The most common approach to treatment is surgical, which involves vitrectomy combined with ILM-peeling, laser photocoagulation along the margin of the optic disc, and intravitreal gas injection. PPV is effective in reducing subretinal fluid and leads to a visual improvement in up to 95% [11].

Vitrectomy with induction of a posterior vitreous detachment (PVD) is an effective method, because it eliminates the traction of the vitreous body around the optic disc pit, which in turn leads to passive fluid migration into the intraretinal space.

In our case, PPV followed by an inverted ILM-flap technique contributed to the subretinal fluid resorption by closing a connection between the vitreous cavity and the ODP, macular reattachment, and, as a result, reduction of central macular thickness in all sectors, which was accompanied by an improvement in visual acuity. Intravitreal injection of viscoelastic material ensured ILM-flap fixation (Figure 3 E – see cover page 3). Laser photocoagulation was not carried out during the vitreous surgery in order to prevent postoperative complications. Intravitreal gas injection of 15% C3F8 was performed to create a temporary barrier blocking the passage of fluid through the ODP.

Conclusion

PPV with an inverted ILM-flap technique for ODP-M allowed to reduce an amount of intra- and subretinal fluid, the visco-associated flap fixation technique created conditions for its stabilization, which ultimately contributed to improving anatomical and functional outcomes during surgery and in the postoperative period.

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Disclosures

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Ethics approval for research involving humans*. This study was performed in line with the principles of the Declaration of Helsinki.*

Abbreviation. *BCVA – best-corrected visual acuity, ILM – internal limiting membrane, IOP – intraocular pressure, IRF – intraretinal fluid, OCT – optical coherence tomography, ODP – optic disc pit, ODP-M – optic disc pit maculopathy, ONH – optic nerve head, PPV – pars plana vitrectomy, PVD – posterior vitreous detachment, SRF – subretinal fluid.*

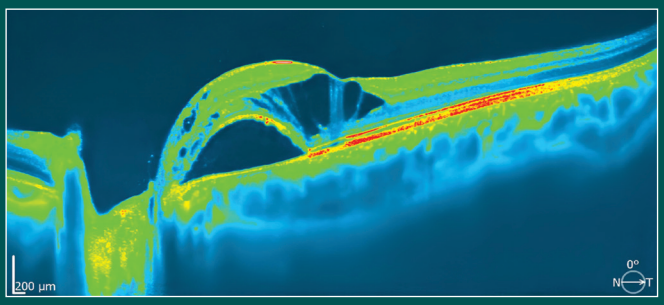

Figure 1. Preoperative OCT shows serous macular detachment and large schisis cavity in the left eye of the patient.

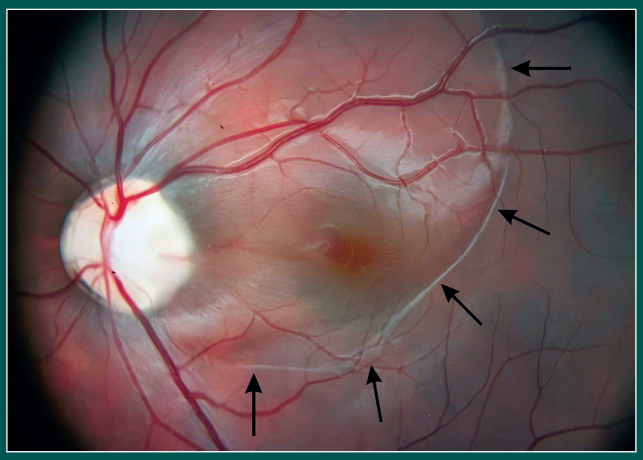

Figure 2. Preoperative color fundus photo of the left eye. Arrows show boundaries of serous macular detachment.

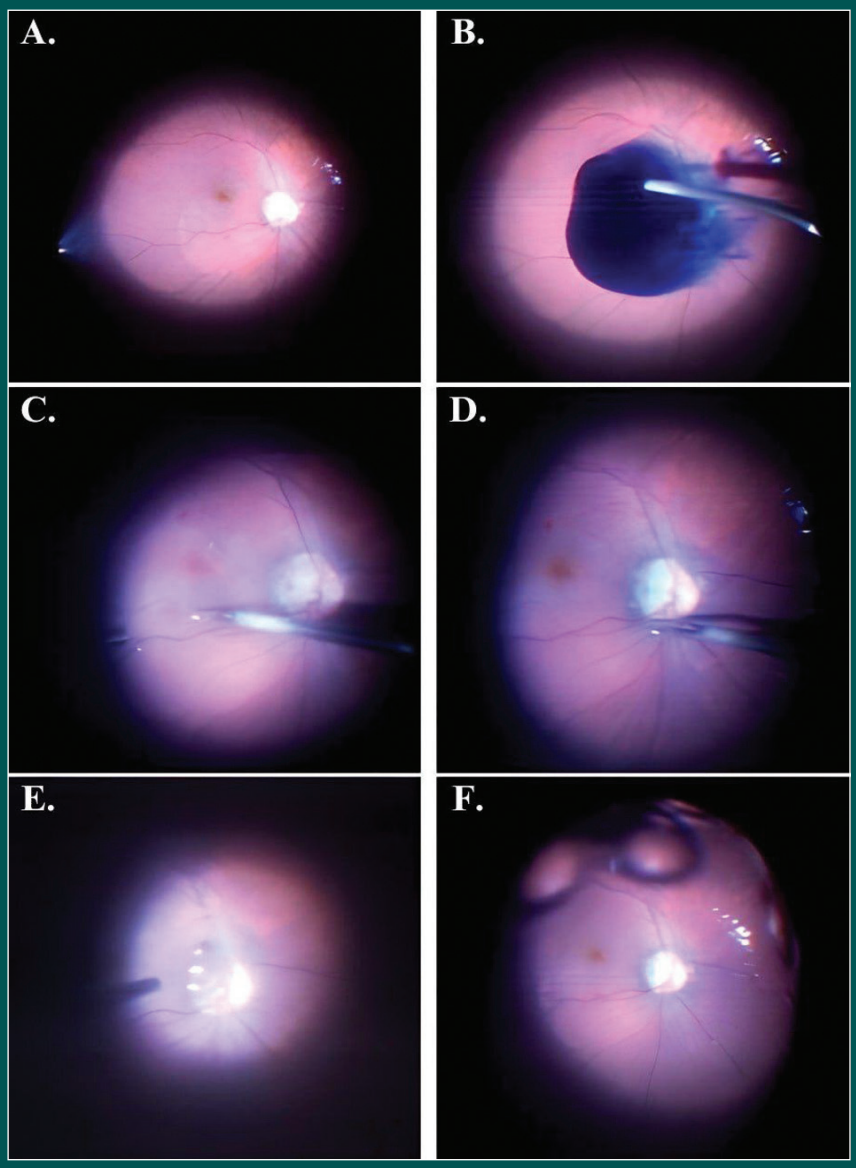

Figure. 3. Stages of the operation:

A. Boundaries of serous macular detachment.
B. Injection of the dye into the vitreous cavity.
C. ILM-peeling with an ILM-flap technique.
D. Placing the ILM-flap over the optic disc pit.
E. Injection of viscoelastic material.
F. Fluid-air exchange.

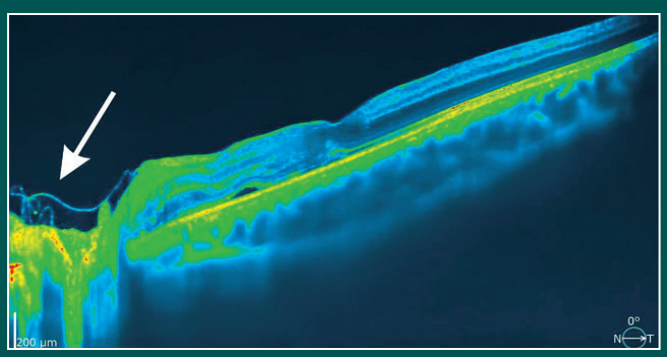

Figure 4. At 3 months postop, OCT examination of the left eye. Arrow shows the ILM-flap.