Intraoperative optical coherence tomography findings during surgery for optic disc pit-associated maculopathy

Hyun Goo Kang¹,², Sung Eun Park¹, Eun Young Choi¹,², Sung Chul Lee², Min Kim¹,²

¹Department of Ophthalmology, Institute of Vision Research, Gangnam Severance Hospital, Yonsei University College of Medicine, 211 Eonjuro, Gangnam-gu, Seoul 06273, Republic of Korea
²Department of Ophthalmology, Institute of Vision Research, Severance Hospital, Yonsei University College of Medicine, 134 Shinchon-dong, Seodaemun-gu, Seoul 06273, Republic of Korea

Correspondence to: Min Kim. Department of Ophthalmology, Institute of Vision Research, Gangnam Severance Hospital, Yonsei University College of Medicine, 211 Eonjuro, Gangnam-gu, Seoul 06273, Republic of Korea. minkim76@gmail.com

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Dear Editor,

We report our experience using a continuous intraoperative optical coherence tomography (OCT)-enabled microscope system for real-time anatomical evaluation of the optic pit during surgery in an 8-year-old girl who presented with symptomatic optic disc pit (ODP) maculopathy. ODP is an extremely rare congenital anomaly secondary to colobomatous malformation of the optic nerve head, with a reported incidence of 0.00009%¹. Although patients are frequently asymptomatic, approximately 40%-50% of patients experience visual symptoms because of subretinal fluid (SRF) accumulation beneath the macula, manifesting as serous macular detachment or retinoschisis¹. The pathogenesis, nature, and source of the fluid in ODP-associated maculopathy remain unclear: whether the fluid is liquefied vitreous through a sieve-like connection or cerebrospinal fluid (CSF) through a channel in the peripapillary subarachnoid space is unknown². Recent advances in imaging technologies, such as enhanced depth imaging (EDI) OCT and intraoperative OCT, have allowed for detailed analysis, with the latter allowing in vivo imaging during surgical maneuvers. Ehlers et al³ used such a device in 2011 and documented a fascinating case of successful aspiration of intraretinal fluid over ODP. Intraoperative collapse of the retinoschisis was demonstrated, and the findings strongly suggested a connection between the vitreous cavity and subretinal space. Endodrainage was further endorsed by Shukla⁴ and other authors in subsequent studies confirming this connection, although the success rates were variable⁵. However, our experience may contradict the previously mentioned reports of a connection between the vitreous and subretinal space³-⁴,⁶.

An 8-year-old girl was referred to our center for serous macular detachment associated with ODP in her left eye. She complained of vision blurring for 1mo. Her visual acuity was 20/67 OS. Fundus examination revealed a dull gray inferotemporal pit and a large serous macular elevation (Figure 1A). OCT showed an abruptly ending hyperreflective lamina cribrosa and an optically empty space corresponding to the pit (Figure 1B). A small hyporeflective tract was seen at the temporal margin of the disc with associated outer retinal cystic cavities. There was no definite channel connecting the vitreous to the subretinal space. The right eye exhibited a normal posterior pole. After frank discussions regarding the potential outcomes and possible need for additional surgery, the guardians agreed to surgical treatment, with consent obtained for intraoperative imaging under an institutional review board-approved protocol. This study adhered to the tenets of the Declaration of Helsinki.

Small-gauge sutureless vitrectomy was performed with continuous high-definition intraoperative OCT using the Zeiss Rescan 700 device with the Lumera 700 microscope (Carl Zeiss Meditec Inc., Dublin, USA). The posterior hyaloid was firmly attached to the optic disc and throughout the posterior pole, necessitating posterior detachment with the aid of preservative-free triamcinolone acetonide. An anomalous translucent vitreous remnant firmly attached to ODP was visualized (Figure 1C) and carefully removed using endoforceps. We noted no change in the height of SRF after these maneuvers (Figure 1D). Fluid-air exchange was performed, with aspiration via a soft-tipped cannula immediately over the site of ODP, with no change in the SRF height even after extensive aspiration (Figure 1E). The vitreous cavity was then refilled with fluid, the internal limiting...
membrane (ILM) was partially peeled, and the resulting ILM flap was stuffed into ODP. Then, juxtapapillary laser photocoagulation was performed with fluid-air exchange. Autologous platelet concentrate was sprayed over ODP. Finally, pneumatic tamponade with C3F8 was performed.

At 3 mo after surgery, her visual acuity was 20/50. OCT revealed a decrease in SRF, although the retinal architecture was not completely restored. In addition, a seemingly enclosed optic pit area with heterogeneously hyperreflective material over the previously visualized hyporeflective tract was observed (Figure 1F, 1G).

Our images were similar to those published by Gowdar et al.[7] who also observed a hyporeflective tract connecting the retinal schisis cavity to ODP on preoperative EDI-OCT. Considering the age of our patient and the consequent lack of vitreous liquefaction, it is difficult to conclude that the extensive SRF with minimal retinoschisis was due to a channel connecting the vitreous cavity to the subretinal space.

Instead, in cases where endodrainage was possible, the surgical induction of posterior vitreous detachment with aggressive aspiration may have iatrogenically created such channels through the attenuated retina. In the present case, we observed a strongly adherent membrane over ODP, also noted by Gowder et al.[7] on EDI-OCT. Akiba et al.[8] described this membrane as a pulsating translucent membrane that was likely to be a persistent anomalous Cloquet's canal and condensed vitreous strands. Aggressive aspiration and traction on this membrane may induce the formation of connections between the vitreous cavity and the subretinal space, which may explain the variable success rates reported in the literature[9-10].

Reviews of surgical treatments for optimal outcomes have converged on a maximal approach[11-13]. In the present case, we additionally considered the patient’s youth and the difficulties of a second surgery. We performed laser photocoagulation for demarcation and compaction of the retinal layers to prevent recurrent fluid influx. ILM flap stuffing and gas tamponade provided mechanical pressure over the pit and aided in physiological sealing of the lamina cribrosa through the formation of a permanent barrier. Finally, autologous platelet concentrate was sprayed over ODP to facilitate physiological sealing.

There are some noticeable differences between our case and that described by Ehlers et al.[3], such as in the patient age and retinal architecture. The disparity in surgical outcomes may suggest heterogeneity in the etiology of ODP-maculopathy, which will require future multi-center studies to uncover.

In conclusion, as opposed to previous findings, we did not observe a connection between the vitreous cavity and the subretinal space; instead, our findings appear to indicate a connection between the subretinal space and a gap in the

Figure 1 Multimodal imaging at various stages of treatment for an 8-year-old girl with ODP-associated maculopathy A: A fundus photograph shows an inferotemporal ODP, a large serous macular elevation, and yellow subretinal deposits; B: OCT reveals an abruptly ending hyperreflective lamina cribrosa, a heterogeneously reflective lesion filling the gap, and a small hyporeflective tract at the temporal margin of the disc; C: After the induction of posterior vitreous detachment, an anomalous translucent membrane firmly attached to the pit can be observed; D: After membrane removal, aggressive aspiration just over the pit did not result in reduction of the macular elevation; E: Extensive aspiration after fluid-air exchange results in no changes in the height of the serous macular detachment; F: At 3 mo after surgery, a gray translucent membrane-like structure is observed over the pit; G: OCT shows a seemingly enclosed optic pit, with compaction of the retinal layers at the temporal margin of the optic disc and reduced serous macular detachment, although there is some residual fluid and the retinal architecture has not fully recovered. Heterogeneously hyperreflective material can be noted over the pit.
lamina cribrosa in the optic pit. Both pre- and intraoperative imaging appears to suggest that a fluid source other than liquefied vitreous may be involved, such as CSF. With this understanding in mind, surgeons may be able to optimize outcomes by designing a maximal approach that targets the closure of this tract.

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REFERENCES