

Pars Plana Vitrectomy with Air Tamponade for Optic Disc Pit Maculopathy: Swept-source Optical Coherence Tomography Imaging Findings and Surgical Approach: A Clinical Case

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Abstract

Purpose: To describe how Swept Source Optical Coherence Tomography (SS-OCT) provides enhanced diagnostic information enabling optimisation of surgical strategy in a pediatric patient suffering from ODP maculopathy. We report the first case of successful management of ODP maculopathy via pars plana vitrectomy (PPV) and air tamponade.

Methods: Interventional case report.

Results: A five-year-old girl presented with ODP and serous macular detachment in her previously amblyopic left eye. SS-OCT showed optically opaque vitreous fibres perpendicularly attached to the base of the optic nerve head excavation and what appeared to be fibrous tissue at the base of the excavation of the optic nerve head and attached to the pit. Detachment of the posterior hyaloid was induced during PPV, the pre-optic nerve head fibrosis was removed with forceps and fluid/air exchange was carried out to induce air tamponade. The serous macular detachment had resolved 11 months after the surgery and the optic pit was reduced in size. No maculopathy recurrences or complications such as cataract were observed. Best corrected visual acuity did not improve possibly because of pre-existing amblyopia.

Conclusion: SS-OCT demonstrated anatomical features not previously described in ODP maculopathy and could assist in the choice of surgical technique. In patients with ODP maculopathy and documented optic nerve head vitreous attachment with peri-pit fibrosis, vitrectomy supplemented by tamponade with air alone may be a viable alternative to tamponade with gas with or without laser photocoagulation. This modified technique may reduce the risk of complications such as cataract formation.

Keywords: Swept Source Optical Coherence Tomography; SS-OCT; Optic Disc Pit maculopathy; PPV with air tamponade

Introduction

Optic Disc Pit (ODP) is a rare condition occurring in approximately 1 in 10,000 of the population. Serous macular detachment occurs in up to 50% of all cases. Currently the most widely accepted treatment for this condition is pars plana vitrectomy surgery (PPV) with or without internal layer membrane (ILM) peeling, gas tamponade and retinal laser photocoagulation. Improvements in Optical Coherence Tomography (OCT) imaging are helping clinicians to better understand ODP maculopathy.

We report a case of ODP maculopathy treated with PPV and air tamponade without ILM peel or laser photocoagulation. Swept-Source Optical Coherence Tomography (SS-OCT) allowed the visualisation of vitreous fibres and fibrous tissue attached to the optic nerve head and optic pit and led us to adopt a less complicated surgical approach to the management of this case.

Case History

A five-year-old girl presented with a history of 5 months of diminished vision in the left eye. A complete ophthalmic examination was performed, including OCT scans with both Fourier-domain OCT (FD-OCT) (Topcon 3D OCT-2000, Topcon Medical Systems, Oakland, New Jersey, USA) and SS-OCT (Topcon DRI OCT-1 Atlantis, Topcon Medical Systems, Oakland, New Jersey, USA). Best Corrected Visual Acuity (BCVA) at diagnosis was 0.0 logMAR in the right eye and 0.8 logMAR in the left eye. There was no past medical or ocular history of note and there was no reported history of ocular trauma. Anterior segment and intraocular pressure were normal in both eyes. A serous macular detachment was observed on dilated fundus examination of the left eye. FD-OCT confirmed this, as well as the presence of macular schisis. An average central retinal thickness (ACRT) of 474 μm was recorded (Figure 1). Total thickness measured included the neuroretina and the subretinal space. SS-OCT revealed vitreous fibres perpendicularly attached to the base of the optic nerve head excavation with absence of the Space of Martegiani and what appeared to be fibrous tissue at the base of the excavation of the optic

nerve and attached to the pit. The size of the optic pit on the SS-OCT scans was 550 μm by 460 μm . (Figure 3). Neither of aforementioned

vitreous fibres or fibrous tissue was visible in the FD-OCT scans (Figure 1).

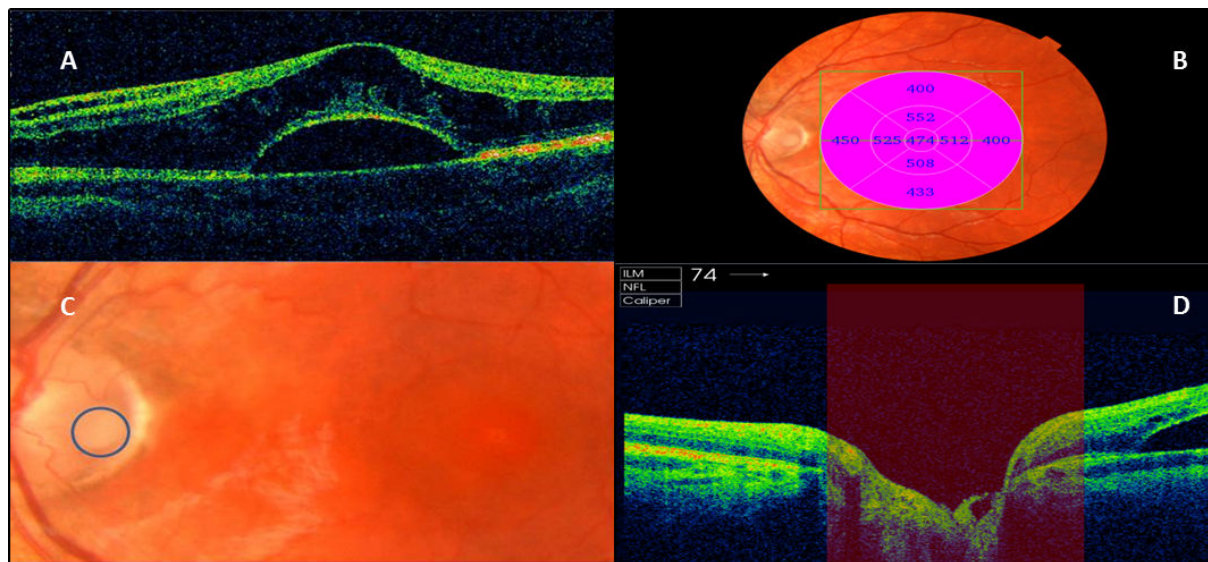


Figure 1: Pre-operative FD-OCT scan showing: macular schisis with serous detachment (A). Average central retinal thickness in the macular Grid (B). Pre-operative appearance of Optic Disc Pit . Highlighted with a blue circle (C) and optic disc scan (D).

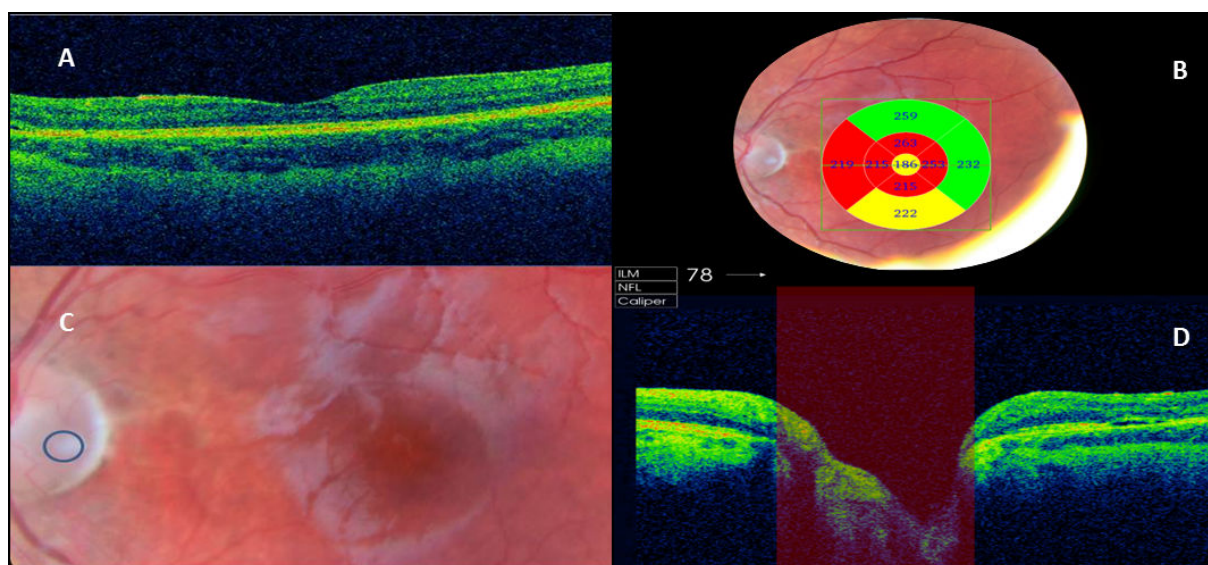


Figure 2: Post-operative FD-OCT scan showing: resolution of the macular schisis and serous detachment (A). Average central retinal thickness in the macular Grid (B). Post-operative appearance of Optic Disc Pit showing reduction in size. Highlighted with a blue circle (C) and optic disc scan (D).

Pars plana vitrectomy surgery was carried out under general anaesthesia. Triamcinolone (Kenacort[®]) was used to highlight the posterior hyaloid and cortical vitreous to ensure induction and adequate detachment of the posterior vitreous (PVD). The fibrous tissue adherent to the optic nerve head and the pit was removed with forceps and fluid/air exchange was carried out. The surgery was followed by twenty-four hours of postoperative face-down posturing.

The air tamponade resolved within the first 72 hours. The patient was followed up for one year. The serous macular detachment progressively resolved and was no longer evident at 11-months (Figure 2). At this time, not only had the ACRT diminished to 186 μm , but also the optic pit size was reduced to 430 μm by 290 μm . Despite resolution of the serous detachment, the visual acuity did not improve. We presume that this was due to pre-existing amblyopia. No corneal or lens opacities developed in the left eye during follow up.

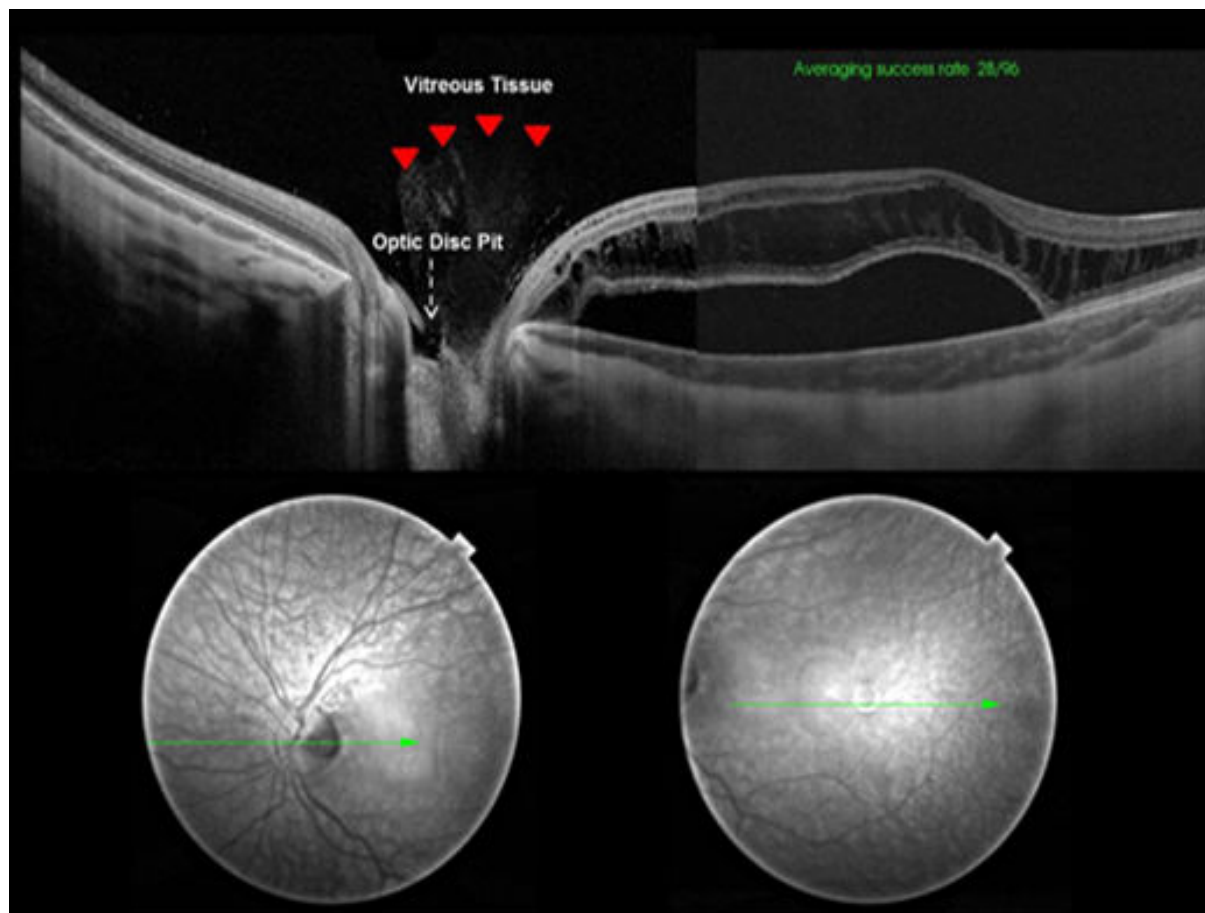


Figure 3: Composite image of two pre-operative SS-OCT scans showing the vitreous attachment to the optic nerve head and the presence of fibrous tissue attached to the optic disc pit.

Discussion

Optic disc pit (ODP) is an abnormal excavation of the optic nerve head, which can be associated with macular serous detachment and/or macular schisis leading to ODP maculopathy. Macular detachment in association with ODP was first reported by Peterson in 1958 [1].

Histological studies of ODP have shown herniation of dysplastic retina into a collagen rich disc excavation. This may extend into the subarachnoid space through a defect in the lamina cribosa. Strands of condensed vitreous terminate at the margin of the pit [2].

Swept-source Optical Coherence Tomography (SS-OCT) is a new imaging technology, which utilises a tunable laser as a light source operated at a 100,000-Hz A-scan repetition rate and with a wavelength of 1,050 nm. Reduced light scattering with deep tissue penetration and uniform image sensitivity, from the cortical vitreous to the internal surface of the sclera, are achieved [3].

Treatment modalities reported for ODP maculopathy include various combinations of PPV, gas tamponade, laser photocoagulation to the temporal juxtapapillary area, internal limiting membrane (ILM) peeling, internal drainage of submacular fluid or macular scleral buckling procedure [4-7].

There have also been reports of using fibrin glue to close the optic nerve pit when it is associated with serous macular detachment [8]. This technique involves pars plana vitrectomy, removal of posterior hyaloid, fluid-air exchange, drainage of subretinal fluid through optic disc pit, application of the fibrin sealant to the pit and air-C3F8 gas exchange and postoperative prone positioning [8,9].

The rarity of this condition makes collation of a large cohort difficult. The largest published series reported outcomes of 20 eyes that had been operated on over a period of 20 years using a variety of surgical approaches. The highest surgical success with 87% of resolution of SMD was obtained with PPV, with ILM peeling and endolaser in eight cases [10].

We have previously proposed a “minimal” surgical approach of only PPV with drainage of subretinal fluid and C3F8 tamponade. We reported a successful anatomical outcome with improvement of visual acuity from counting fingers to 6/18 [6].

We now describe the use of pars plana vitrectomy with removal of the vitreous and fibrous optic nerve head and optic pit attachments combined with air tamponade. Pre-interventional SS-OCT high resolution scans showed anatomical features previously not described which allowed for a less complex and less invasive surgical approach to be executed. Eleven months into the post-operative period, the macula

was reattached. The subsequent reduction of the diameter of the optic pit suggests potential pre-operative traction from the vitreous in maintaining the patency of the pit (Figure 2)

The pre-operative absence of a posterior vitreous detachment in our case may also have contributed to our good surgical result, as this was reported to be a positive prognostic factor by Haruta et al. [5].

Ohno-Matsui et al., first described SS OCT imaging of ODP. The authors reported a defect in the lamina cribrosa at the site of the pit with herniation of nerve tissue into the pit. However, there was no description of the absence of the Space of Martegini, the attachment of vitreous fibres to the optic nerve head nor the presence of fibrous-looking tissue attached to the optic pit, as in our case [11].

SS-OCT showed the restoration of a normal-looking anatomy at the level of the outer retinal layers anatomy (Figure 2). The lack of post-operative improvement in visual acuity may be due to the long standing maculopathy with serous retinal detachment and the associated hypermetropic amblyopia.

The formation of cataracts following gas tamponade has been reported in 24%-92% of cases [12-14]. We did not observe any lens opacity in our patient after 1 year of follow up.

In conclusion, pre-operative SS-OCT improved the visualization of the cortical vitreous and highlighted a possible role of vitreous traction in the pathogenesis of ODP maculopathy. Our modified surgical technique enabled us to minimize damage to OD and papillomacular bundle by avoiding endolaser and also minimize damage to the macular area by avoiding ILM peel and the risk of dissociated outer retina [15]. The use of air tamponade perhaps also reduced the risk of cataract formation, when compared to the risk associated to the use of gas tamponade. This case report shows that PPV with induction of posterior vitreous detachment, removal of any fibrous tissue attached to the optic pit and air tamponade can be a valuable alternative to PPV with laser photocoagulation and gas tamponade for the treatment of OPD maculopathy.

Herein, we report the presence of vitreous fibres attached to the optic nerve-head and optic pit and their possible role in the pathogenesis of this condition. These findings have only been previously described in histopathology studies. However, a larger series of cases imaged with SS-OCT is needed to show the prevalence of our findings in OPD maculopathy.

Here we report a "less invasive" surgical approach for the treatment of this condition and as a consequence of the findings on SS-OCT.

This case report may also highlight the role of vitreous traction in ODP maculopathy and a possible role for enzymatic vitreolysis combined with a pneumatic retinopexy in the treatment of ODP maculopathy.

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Proprietary Interest

None of the authors has proprietary interest in the manuscript.

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