Optic Disc Pit

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ABSTRACT

Optic disc pits are a very rare clinical entity with a prevalence of approximately one in 11,000 people. Patients with congenital optic disc pit may remain asymptomatic, but 25% to 75% of patients in their 30s or 40s experience visual deterioration due to the most common complication of this condition “optic disc pit maculopathy”. Treatment of this complication is quite challenging. The most widely accepted treatment is surgical approach involving pars plana vitrectomy with or without internal limiting membrane peeling, with or without endolaser photocoagulation and C3F8 endotamponade. Optic disc pit associated maculopathy has better outcomes when it is plugged either with internal limiting membrane flap, autologous scleral plug or human amniotic membrane.

Keywords: Optic disc pit, optic disc, pathophysiology of optic disc.

Optic disc pit is a rare congenital abnormality that affects one out of every 11,000 people[1]. The majority of them are unilateral, while 10% to 28% are bilateral[1]. Optic disc pits, according to some researchers, are small colobomatous abnormalities of the optic disc caused by inadequate closure of the ocular foetal fissure in the optic disc region[1]. In roughly 40% to 66% of eyes with optic disc pits, simultaneous nonrhegmatogenous, serous retinal detachment, or symptoms suggesting of earlier detachment, can be identified, most commonly in the second or third decade[1].

Optic disc pit associated maculopathy develops in 25% to 75% of patients[2-4]. Maculopathy can results from subretinal fluid leading to serous macular detachment or intraretinal fluid causing serous detachment or macular schisis or both[5].

The exact origin and pathophysiology of the intra- and sub-retinal fluid, first described by Wiethe **6** in 1882, remain unknown[6,7,8]. Interestingly, there are no obvious or known triggering events for the development of ODP-M. It has been linked to posterior vitreous detachment (PVD)[7,9,10].

Vitreous traction may be linked to the development of ODP-M, given that progressive vitreous liquefaction begins in thirties. In contrast to individuals without ODP-M, Shah et al.[11] analysed data from earlier studies and discovered that most of the patients had posterior vitreous detachment. Furthermore, after PVD is completed, ODP-M can resolve spontaneously[12].

Origin of fluid: The eyes of Collie dogs are examined[13], both imaging and histology investigations revealed that the ODP defect does not allow connection between the subretinal space and the vitreous. A histopathologic examination of two eyes with ODP-M, on the other hand, revealed the presence of mucopolysaccharides, a vitreous component, inside the ODP[14].

One theory proposes that migration of fluid from the vitreous to the subretinal area is due to the pressure gradients within the eye. The intracranial pressure is conveyed to the ODP through the cerebrospinal fluid, resulting in a pressure gradient in patients with ODP. As a result, vitreous fluid might leak back to the pit in case of low intracranial pressure. When the intracranial pressure rises, the fluid is forced back into the eye, where it dissects inside or beneath the retina[7,15] This could also explain why eyes with ODP have subretinal, intraretinal, and intracranial gas or silicone oil transitions[16,17-19,20].

Regardless of the fluid source, it appears that ODP-M follows a well-known sequence of events[7,21,22,23]. Fluid from the ODP generates an inner retinal separation that looks

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like a schisis and is linked to a mild cecocentral scotoma at first. After that, a subretinal fluid dissection causes an outer retinal detachment[24,25].

According to one study, most of the patients with optic disc pit associated maculopathy have good visual outcome and may have spontaneous clearance of subretinal fluid without the need for surgery[26]. Parikakis et al.[27] presented a long-standing case of serous macular detachment with optic disc pit which resolved spontaneously with improvement in visual acuity, hypothesising that IS/OS junction integrity may be a prognostic factor for visual outcome and OCT as an indicator of final visual prognosis and the likely need for surgical management.

Vitrectomy to relieve vitreous traction on the macula and pit by inducing posterior vitreous detachment is an important part of the treatment for this condition[28, 29, 30, 31, 32]. Other methods including laser photocoagulation in peripapillary region, internal limiting membrane peeling and gas tamponade, however, are still controversial[33, 28, 29, 34-37]. The administration of a barrage along the affected disc margin to avoid fluid migration into retinal layers is the basis for laser photocoagulation in ODP maculopathy[36, 38, 39]. However, no long-term follow-up was done on laser photocoagulation complications. There is also no research comparing pre-operative visual field characteristics or macular sensitivity to surgery outcomes.

Marticorena recommended brilliant blue dye under air to delineate the intended treatment area once it pools in the submacular space as a method of selective laser application[40].

On 18 patients, with vitrectomy, hyaloid separation, and endolaser, Gregory-Roberts et al.[41] reported positive outcomes, but no air-fluid exchange.

Hirakata et al.[42] concluded that vitrectomy with posterior vitreous detachment induction without gas tamponade or laser was a successful treatment for ODP associated maculopathy. One patient did not improve in their 41 retrospective study of eight patients, and repeat vitrectomy, internal limiting membrane peeling with gas tamponade failed as well. Only the third surgical attempt, when a peripapillary laser was used, was able to minimise the retinal detachment. It's possible that laser might speed up the resolution of SRF.

In 46 patients with optic disc pit maculopathy, Abouammoh et al.[43] compared the outcomes of pars plana vitrectomy (PPV) along with juxtapapillary laser photocoagulation (JLP) and vitrectomy without JLP. They found that PPV with JLP and without JLP has similar functional and anatomic outcomes.

It has been reported that PPV with the development of a posterior vitreous detachment (PVD), gas tamponade and laser result in complete retinal reattachment and remission of the retinal schisis[44].

PPV combination with JLP and gas tamponade was advocated by Jain and Johnson[45] as their preferred treatment for ODP maculopathy. They advocated using a moderately powerful laser photocoagulation technique 1-2 hours prior to the vitrectomy procedure, and they were able to achieve 100% anatomical success. This was, however, based on unpublished data of ten eyes with no comparison group.

A laser-induced scar at the juxtapapillary area, on the other hand, may damage exiting axons and cause functional impairment. Furthermore, laser scars may expand over time, compounding the damage already done.

Rayat et al.[46] concluded in a recent case series that JLP has no further advantage for ODP associated maculopathy. They also found that internal limiting membrane peeling with gas tamponade had no effect on the final outcome. They did not include macular schisis removal rate in optic disc pit in their definition of success since they did not report on it. In these set of cases, anatomic success was ambiguous.

Onto et al.[47] described a new inner retinal fenestration approach in which a partial-thickness fenestration near the optic disc pit was used to channel fluid in the vitreous rather than retina. They recently reported complete clearance of fluid in 17 eyes (94%) in and under the fovea with no recurrence without additional treatment. Similarly, pars plana vitrectomy was combined with the construction of an inner retinal fenestration by making a partial-thickness retinotomy immediately temporal to the pit with a bent needle of 25-gauge. It was linked to improvements in retinal anatomic and functional characteristics without the need for additional therapies. These data support the concept that redirecting fluid in the vitreous rather than the retina can result in long-term improvement of the pathologic features of optic disc pit associated maculopathy[48].

Vitrectomy is the most common treatment for ODP maculopathy, and it appears to be more effective when combined with internal limiting membrane (ILM) inverted flapping to cover the ODP. If the fluid originates in the vitreous cavity, the technique will yield positive results. Another innovative treatment involves packing the ODP with rolled ILM-flap, autologous scleral tissue, or human amniotic membrane, which has shown to produce good anatomical results[49].

As adjuvants to PPV treatment of ODP maculopathy, ILM flap procedures are rational and easy. They could be effective adjuvant therapies for patients with ODP maculopathy who want to improve their BCVA[50].

The surgeon used a vitreous forceps to stuff the pit with the inverted internal limiting membrane flap, then a single row of laser was applied in the region of removed ILM along the temporal margin of the pit. The fluid/air exchange was performed at the conclusion of the surgery. Patients were advised "face down" posture for three days. In all three patients, spectral-domain OCT showed post-operative anatomical improvement, defined as full absorption of fluid 1 week after surgery[51].

Babu et al.[52] included 23 patients (23 eyes) in their retrospective analysis who had 25-gauge pars plana vitrectomy (PPV), internal limiting membrane (ILM) peeling with gas tamponade. Inverted ILM flap and autologous scleral flap were used for pit plugging in groups 2 (n=7) and 3 (n=8) respectively, but the pit was not filled in group 1 (n=8). They came to the conclusion that if the pit is plugged, OPD associated maculopathy has a better outcome. Both the inverted ILM flap and the autologous scleral plug are effective for pit plugging.

In conclusion, covering and packing the optic disc pit improves surgical results. During the first two years after surgery, the surgical intervention appears to improve vision. Because the fluid takes time to absorb, vision improvement is linked to the anatomical restoration of retinal shape, which takes 6-12 months.

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