Surgical management of optic disc pit maculopathy with a fovea sparing internal limiting membrane flap

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Optic disc pit (ODP) is rare congenital cavitary anomaly of the optic disc. Serous detachment of macula is the most common complication of ODP and occurs in 25%–75% of these cases. Although various surgical techniques have been used for the treatment of ODP maculopathy; consensus still eludes as far as the optimal surgical approach is concerned. We herein report a case of ODP maculopathy in a young female treated successfully with vitrectomy, fovea sparing internal limiting membrane flap, and C3F8 tamponade.

Key words: Optic disc pit, optical coherence tomography, serous macular detachment

Optic disc pit (ODP) maculopathy is an established complication of ODP occurring in 25%–75% of these eyes.[1] The natural history of untreated ODP maculopathy portends a poor visual outcome with the final visual acuity of 20/200 or worse.[2] A number of treatment options have been explored which include laser photocoagulation at temporal optic disc margin, pars plana vitrectomy with or without intravitreal tamponade, internal limiting membrane (ILM) peeling, macular buckle, subretinal drainage with autologous platelets over the ODP and sealing the ODP with inverting peeled ILM or Tisseel fibrin sealant.[2]

Ho et al. proposed preserving the Epi-foveal ILM during ILM peeling for myopic foveoschisis management thus preventing the postoperative development of full-thickness macular holes (FTMH).[3] We report a case of ODP maculopathy in a young female, treated surgically with vitrectomy and fovea sparing ILM peeling technique.

Case Report

A 27-year-old woman presented with painless, progressive diminution of central vision in her left eye for 1 year. Best-corrected visual acuity (BCVA) was 20/20 and 20/50 in her right and left eye, respectively. Anterior segment examination was unremarkable in both eyes. Fundus of the right eye was normal. Left eye revealed an oval, gray-yellow craterlike depression at the superotemporal aspect of optic disc [Fig. 1a]. Left eye also had serous macular detachment measuring about 1.5 disc diameter. Spectral domain optical coherence tomography (SDOCT) of the left eye showed outer layer retinoschisis (arrow), inner layer retinoschisis (arrowhead) with an outer layer hole (star) [Fig. 1b].

Figure 1: (a) Color fundus photograph of the left eye at presentation showing a superotemporal optic disc pit (arrow) and neurosensory detachment (arrowhead). (b) Spectral domain optical coherence tomography across the macula shows outer layer retinoschisis (arrow), inner layer retinoschisis (arrowhead) with an outer layer hole (star).

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eye showed outer layer retinoschisis, inner layer retinoschisis, and outer layer hole (OLH) with central macular thickness of 737 μm [Fig. 1b]. SDOCT of the right eye was normal.

The patient was treated with diode laser temporal to the optic disc with burns in two to three rows. Four months later, the patient returned with no improvement in vision (BCVA of 20/125 in the left eye). SDOCT revealed the persistence of the schitic retina with an OLH. Subsequently, she underwent three ports 25-guage pars plana vitrectomy with fluid-air exchange using 14% C3F8.

Core vitrectomy was done and then intravitreal triamcinolone acetonide assisted posterior vitreous detachment was done. Instead of the conventional ILM peeling technique; a fovea sparing ILM flap was fashioned and left in place over the fovea [Fig. 2: intraoperative photograph; Brief surgical technique is highlighted in Video 1]. Postoperative prone positioning for 12 h a day for 2 weeks was maintained. One month after the surgery, BCVA in left eye improved to 20/80. Fundus showed reduction in the schisis which was documented both clinically and by SDOCT (central macular thickness of 481 μm) [Fig. 3a]. At 7 months postsurgery, SDOCT showed near complete regression of the schitic retina with the restoration of the foveal contour (central macular thickness measuring 261 μm) [Fig. 3b].

**Discussion**

ODP is a rare and typically unilateral congenital cavitary anomaly of the optic disc. Although uncomplicated ODP remains asymptomatic; an ODP complicated with maculopathy can cause severe visual impairment necessitating aggressive treatment approach.[5] Nature and source of fluid that is found intra- and sub-retinally in these eyes is controversial. Vitreous, cerebrospinal fluid, and blood from ODP or choroid have been postulated as the likely source of the fluid.[2,4] Further, the pathogenesis of this entity also remains unclear. Our group has reported that the involvement of the ORL was the first step in ODP maculopathy. Thereafter, the fluid could seep be bi-directionally into subretinal space or the inner retinal layers.[3]

There is very little clarity regarding the combination of treatment procedures for ODP maculopathy that would maximize the surgical success while minimizing side effects and possible complications.[6]

ILM has been suggested to be an important component of ODP maculopathy. Tangential and anteroposterior tractions are believed to facilitate the passage of fluid from the optic pit into the macula and ILM peeling eliminates this tangential traction.[5] Shukla et al. have performed vitrectomy with ILM peeling, barrage laser photocoagulation and gas tamponade in their study. Good surgical outcomes were recorded by the authors in terms of restoration of macular anatomy and visual improvement. However, they report that four out of seven patients developed FTMH postoperatively. The authors attributed the high incidence of FTMH to the peeling of ILM over thinned out retina.[5] Mohammed and Pai achieved satisfactory anatomical and functional outcomes by using an inverted pedicled ILM flap to close the ODP. The authors reported that obtaining such a flap could be technically challenging along with the possibility of postoperative displacement of the flap from the ODP.[6] Since there have been many theories put forward regarding the source of fluid in ODP maculopathy, we believe that sealing the ODP solely may not prevent movement of fluid into the retina.

In our case, in view of the extremely thinned out retina and the possible risk of developing postoperative macular hole following ILM delamination, we performed a fovea sparing ILM flap which was left in place. There was good functional and dramatic anatomical improvement during follow-up. To the best of our knowledge, treatment of ODP maculopathy with fovea sparing ILM peeling flap in conjunction with laser photocoagulation, vitrectomy and gas tamponade has not yet been reported in the literature.

To summarize, vitrectomy with gas tamponade in conjunction with fovea sparing ILM peeling is a viable option for the surgical treatment of ODP maculopathy.

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Conflicts of interest
There are no conflicts of interest.

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