Management of double optic disc pit complicated by maculopathy

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Optic disc pit (ODP) is a rare, congenital, cavitary anomaly of the optic disc. Usually, single ODP occurs in an eye and only eleven cases of double ODP have been reported so far in the literature. In the present study, we report a case of unilateral double ODP, with both the pits present in opposite disc segments. They were associated with serous retinal detachment at the macula and retinal pigment epithelium degeneration nasal to disc. The case was managed successfully with vitrectomy, internal limiting membrane peeling, plugging the pits with homologous partial-thickness scleral flaps, and gas tamponade.

Key words: Optic disc pit maculopathy, scleral plug, vitrectomy

Optic disc pit (ODP) is one of the several congenital cavitary anomalies of the optic disc, caused due to imperfect closure of the embryonic fissure during development.[1] It is most commonly located in the inferotemporal segment of the disc.[1] Usually, single ODP occurs in an eye, with double ODP being rarely reported in the literature.[2-10]

We report the successful management of a patient presenting with unilateral, symptomatic, double ODP involving opposite segments of the disc.

Case Report

A 34-year-old male presented with a painless, gradual decrease in vision in his left eye. His best-corrected visual acuity (BCVA) was 20/20 in the right eye and 20/240 in the left eye. Intraocular pressures (IOP) and pupillary reactions were normal in both the eyes. Anterior segment examination was unremarkable bilaterally. Posterior segment examination in the left eye showed two distinct ODPs, one each in inferotemporal and inferonasal segments of the disc; serous retinal detachment (SRD) at the macula and retinal pigment epithelium (RPE) degeneration at macula; and RPE degeneration nasal to disc [Fig. 1a]. Fundus autofluorescence showed hypo-autofluorescence in the region corresponding to SRD at macula; and hypo-autofluorescence nasally in the region corresponding to RPE degeneration, which was bordered by a hyper-autofluorescence ring [Fig. 2].

Optical coherence tomography (OCT) through the disc showed two distinct hyporeflective areas, one each in inferotemporal and inferonasal segments, suggestive of two ODPs. Communication between the optic disc and retinal layers was seen in the region of the inferotemporal pit [Fig. 3a and b]. OCT through the macula showed serous retinal detachment (SRD, 1353μ) and intraretinal fluid but no outer retinal dehiscence (ORD) [Fig. 4a].

After taking informed consent from the patient, he underwent 25/23-gauge (G) pars plana vitrectomy (PPV). Posterior vitreous detachment induction was followed by internal limiting membrane (ILM) peeling 2-disc-diameter around the fovea, fluid-air exchange (FAE), and active internal subretinal fluid (SRF) drainage through the pit. Although intraoperative OCT was not used after the SRF drainage SRD was seen to decrease clinically. One of the 25G trocar-cannula systems was replaced with a 23G system. Two homologous partial-thickness scleral flaps (1 mm × 1 mm) were crafted from the inferonasal quadrant of the same eye, which was then plugged into each pit. Finally, FAE was done and tamponade was given with 20% sulfur hexafluoride gas. Endolaser was not done [Supplemental Video 1].

Forty-five days postoperative, his BCVA improved to 20/80. The scleral plugs were seen to be snugly fitting in the two pits [Fig. 1b]. OCT showed that the communication between the optic disc and retinal layers had closed [Fig. 3c]. Intraretinal fluid resolved completely while SRD reduced to 300 μ [Fig. 4b]. There was no postoperative full-thickness macular hole (FTMH) formation or resorption of the scleral plug.

Discussion

First described in 1882, ODP is a rare congenital anomaly with an estimated incidence of 1 in 11,000 population. Histopathologically, it is described as herniation of dysplastic retina into a collagen-lined pocket, which overlies the optic nerve and extends through a defect in lamina cribrosa.[1] Usually, only one ODP is seen in an eye, however, rarely two distinct pits have been reported.[2-10]

We present the management of a case with double ODP, where both the pits were present in opposite segments. As studies have shown that sealing the pit with adjuncts such as inverted ILM flap and scleral plug causes faster fluid resolution and early visual recovery, the pit is routinely plugged in

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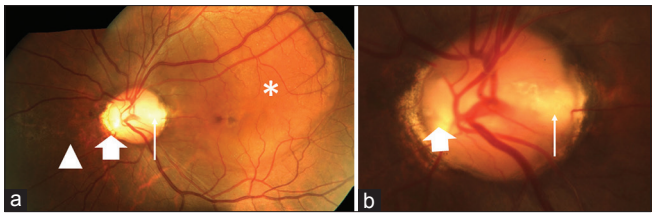

Figure 1: Colour fundus images showing (a) preoperative status of the two distinct optic disc pits. The pit in inferotemporal segment of disc (thin arrow) was associated with large serous retinal detachment (asterix) and retinal pigment epithelium (RPE) degeneration at macula, while the one at inferonasal end (solid arrow) was associated with RPE degeneration nasal to disc (arrowhead), (b) postoperative status of the two pits, each snugly fit with scleral plug

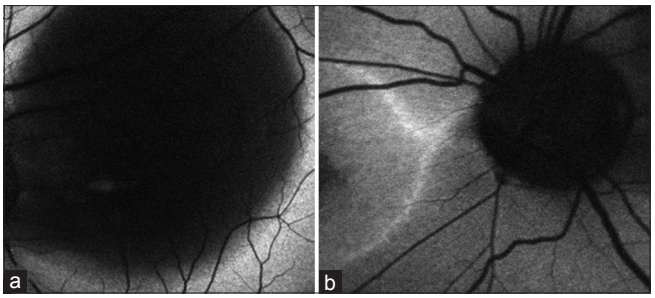

Figure 2: Fundus autofluorescence showing (a) hypo-autofluorescence in the region corresponding to SRD at macula; and (b) hypo-autofluorescence nasally in the region corresponding to RPE degeneration, which was bordered by a hyper-autofluorescence ring

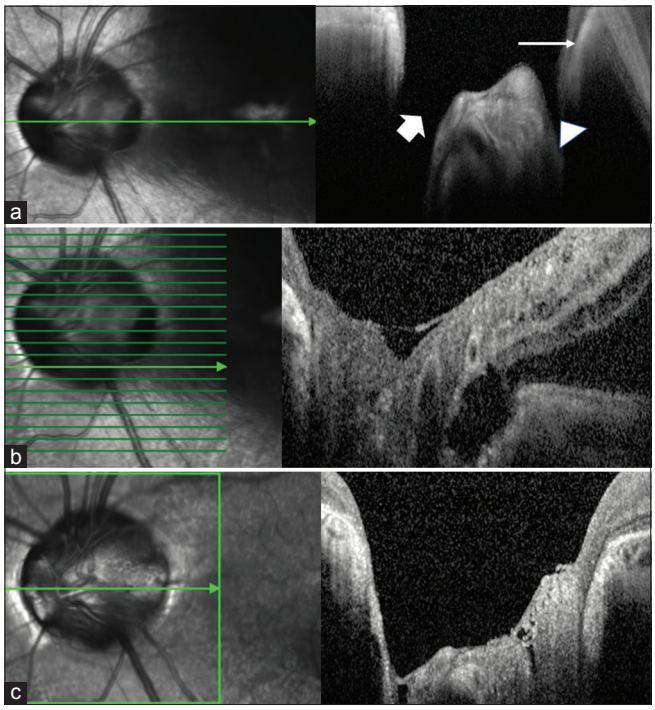

Figure 3: Optical coherence tomography images of the line scans through disc showing (a) preoperative status of optic disc showing two distinct pits, one at the inferotemporal segment of disc (solid arrow) and the other at inferonasal segment (arrowhead), and communication between the disc and retinal layers (thin arrow); (b) preoperative status of optic disc showing communication between the pit and subretinal space; (c) postoperative status showing closure of the communication

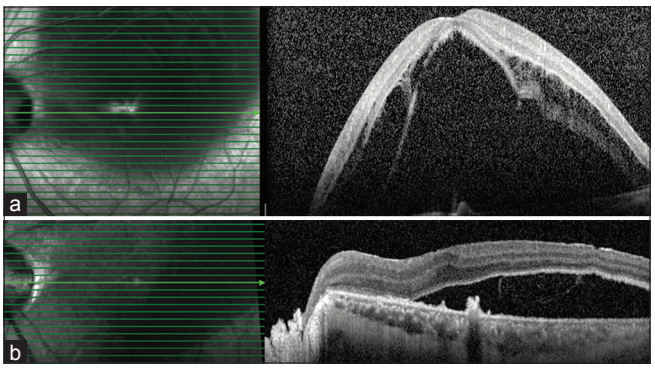

Figure 4: Optical coherence tomography images of the line scans through macula showing (a) preoperative status of macula with serous retinal detachment (SRD, 1353 μ) and intraretinal fluid; (b) 45 days postoperative status showing further reduction of SRD (300 μ). No macular hole formation was observed

all the cases of ODP-maculopathy at our center.[1] The RPE degeneration nasal to the disc could be attributed to multiple, asymptomatic, spontaneously-resolved episodes of SRD. As both the pits were causing retinal fluid accumulation, we decided to treat both the pits. The next challenge was obtaining two flaps to tuck both the pits. We decided to use the scleral plug to stuff the pits instead of ILM flaps due to relative surgical ease in obtaining two scleral flaps compared to two ILM flaps, higher stability of scleral flaps during FAE compared to ILM flaps, and absence of literature evidence to prove superiority of either modality for the treatment of ODP-maculopathy.[1] The sclera has been proved to be a good option as it has an intimate relationship with the optic nerve and remains in correct position even after 2 years.[11] The scleral flap has not only been used successfully as a rescue treatment in failed cases but also as a primary modality.[11,12] Hence, the patient was advised to undergo vitrectomy along with ILM peeling and scleral plugs for both the pits. Since it is not possible to insert a 1 mm × 1 mm scleral plug through 25G trochar-cannula system, one of the trochar-cannula systems was replaced with 23G one at the time of inserting the scleral plug inside the vitreous cavity. The scleral plugs snugly fit into the pits and ensured adequate closure of the communication between the optic disc and retinal layers. Consequently, the fluid started to resolve with notable visual gain. Studies have shown that it takes 6 months to 1 year for complete fluid resolution.[13] As there was no preoperative ORD, there was no formation of FTMH postoperatively.[13]

Till date, only eleven cases of double ODP have been reported in the literature [Table 1]. Out of these eleven cases, five eyes had pits in opposite segments of the disc i.e. nasal and temporal; while the rest six eyes had both the pits in the single segment i.e. temporal.[2-10] Out of the five eyes with nasal pit, only two eyes had tell-tale retinal signs.[2,4,5,8] Overall, two cases underwent vitrectomy along with posterior hyaloid separation, endolaser, and gas tamponade. In both cases vision improved from 20/200 to 20/50 and from 20/60 to 20/20 after a follow-up of 1 and 6 years, respectively.[5]

There is a theoretic risk of mechanical trauma to the nerve fibers during the insertion of the scleral plug, leading to visual field defects. However, visual field study was not done for this

Table 1: Overview of the cases reported in the literature with double optic disc pit

Author	No. of cases	Age/ Sex	Optic disc pit locations	BCVA	Macular involvement	Nasal retina involvement	Treat-ment
De Laey *et al.* (1979)[2]	1	NA	Temporal, Nasal	NA	Minimal	Extensive wedge-shaped area of choroidal dystrophy (unnoticed SRD)	NA
Jonas *et al.* (1987)[3]	2	NA	NA	NA	NA	NA	NA
Silva *et al* (2010)[4]	1	72/M	Temporal, Nasal	20/50	RPE atrophy	No	No
Gregory-Roberts *et al.* (2013)[5]	2	28/M	Temporal, Nasal	20/200	Intraretinal schisis, SRD	NA	Surgery
		47/M	Temporal, Nasal	20/60	Intraretinal schisis, SRD	NA	Surgery
Choudhry *et al.* (2015)[6]	1	16/M	Temporal	20/40	Intraretinal schisis, No SRD	No	No
Ali *et al.*[7] (2016)	1	51/M	Temporal	20/60	No	No	No
Koulouri *et al.* (2018)[8]	1	35/F	Temporal, Nasal	20/30	Intraretinal fluid, outer retinal schisis	Intraretinal fluid	No
Pozza *et al.* (2018)[9]	1	30/F	Temporal	CF	Large SRD	No	No
Boese *et al.* (2018)[10]	1	29/F	Temporal	20/20	No	No	No

(No.: Number, BCVA: Best-corrected visual acuity, NA: not available, RPE: retinal pigment epithelium, SRD: serous retinal detachment, CF: counting finger)

patient at any point in time. This remains as the major limitation of the study. We presented the first case of a double ODP successfully treated by plugging of the pits with scleral flap.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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