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Case Report

Unusual coexistence of bilateral optic disc pits with unilateral macular hole

Lt Col Avinash Mishra ^{a,*}, Somesh Aggarwal ^b, Col Neeraj Bhargava ^c,
Col V.K. Baranwal ^d

^a Classified Specialist (Ophthalmology), Military Hospital, Ahmedabad, India

^b Associate Professor, M & J Western Regional Institute of Ophthalmology, Ahmedabad, India

^c Senior Advisor (Ophthalmology), Command Hospital (Southern Command), Pune, India

^d Senior Advisor (Ophthalmology), Command Hospital (Northern Command), Udhampur, India

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retinal detachment, central serous chorioretinopathy. However bilateral optic disc pit in association with an unilateral macular hole is so rare that to the best of our knowledge, macular hole surgery in a case of optic disc pit, has been mentioned in literature only thrice earlier.^{3–5}

Case report

A 31-year-old male patient reported with complains of blurred vision in his left eye for the past 2 weeks. He had no other symptoms, no previous ocular disease history and his general health was unremarkable. A history of trauma to the eye was specifically asked for and the patient also did mention that he had mildly hurt his left eye with his hand sometime back, which brought him to notice the problem in the affected eye. However at that point we thought that it was too insignificant. Ocular examination on presentation revealed a vision of 6/6 in his right eye while vision in the left was reduced to 1/60. Anterior segment examination was normal in both eyes. The intraocular pressure too was a normal 14 mm Hg as measured with a non contact tonometer (NCT). Amsler grid testing revealed a central scotoma in only the left eye. A dilated funduscopy revealed bilateral optic disc pits in the infero-temporal part of the optic disc. The left eye in addition revealed a full thickness macular hole. The rest of the fundus was essentially normal.

The patient was further investigated with fundus photographs [Figs. 1 and 2] as well as an optical coherence tomography (OCT) [Figs. 3 and 4]. OCT further revealed a large

Introduction

Optic disc pit, also known as ‘atypical colobomas’, is an oval, grayish-white depression in the optic disc which results due to an imperfect closure of the superior edge of the embryonic fissure.^{1,2} It's a rare clinical entity having an incidence of about 1 in 10,000, being bilateral in only 15% of the cases.² It can be either congenital or acquired. Patients with optic disc pits may be asymptomatic, being detected during routine examinations. However, many of them present with visual symptoms due to macula-related complications such as serous retinal detachment or cystoid macular oedema.

Though optic disc pit may rarely co exist with other ocular conditions like for eg keratoconus, posterior retinoschisis,

* Corresponding author. Tel.: +91 (0) 9408330655 (mobile).

E-mail address: avim27@yahoo.com (A. Mishra).

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retinoschisis along with a neurosensory detachment, a total loss of foveal contour and possibly an epi-retinal membrane (ERM) in the right eye. OCT left eye too revealed a retinal detachment along with a stage 4, macular hole. Visual Field Analysis (VFA) was normal in the right eye while in the left eye it could not be carried out due to his poor vision. He was advised macular hole surgery, under a guarded prognosis, for his left eye, however he refused. He was also advised prophylactic surgery i.e (Pars Plana Vitrectomy without internal limiting membrane peeling and photocoagulation around the pit to prevent a hole formation in his right eye). However he refused this too, perhaps because his vision was good in this eye. Presently he is being followed up on an outpatient basis and his condition has remained stable for the past 6 months of follow up. He has also been given an Amsler grid for self-monitoring and educated regarding the macular detachment symptoms, with instructions to report immediately in case they occur in the right eye.

Discussion

Wiethe, in 1882, first described optic disc pits as small, grey–white, round to oval depressions found at the optic nerve head.⁶ They occur due to incomplete closure of the superior edge of the embryonic fissure, usually during the first trimester of embryogenesis. A more severe effect of incomplete closure is a complete coloboma of the optic nerve. It usually occurs sporadically with men and women being equally affected their most common location is on the temporal and inferotemporal border of the optic nerve head with over 50% being located temporally on the nerve head, while about one-third of them are located more centrally on the disc.⁷ Optic pits range in size from 0.1 to 0.7 disc diameter along their widest dimensions and have a mean depth of about 5D. They are most commonly grey (60%), yellow (30%) or black (10%) in colour. There is usually one pit per optic disc although two or three may also occur on the same disc.

Fig. 1 – Fundus photograph (Right eye) showing the optic disc pit located infero-temporally on the disc.

Fig. 2 – Fundus photograph (Left eye) showing the optic disc pit along with a complete macular hole.

Although optic pits were described more than a century ago, the pathogenesis and pathologic nature of the associated macular lesions remain controversial. It has been suggested that the subretinal fluid could be originating from the vitreous, considering the higher incidence of posterior vitreous detachment in eyes with optic disc pits. Others believe that the cerebrospinal fluid could be the source of subretinal fluid, although no connection between the pit and the subarachnoid space has yet been demonstrated. Yet another suggestion is that it might be due to leakage from the choroid or from the vessels within the pit.⁸

Patients with congenital optic disc pit sometimes remain asymptomatic, but around 25%–75% of these cases present with visual deterioration in their 30s or 40s after developing an optic pit maculopathy.³ It is quite uncommon to have a macular hole in the presence of concomitant disease conditions such as retinitis pigmentosa (RP), choroidal coloboma, and optic disc pit.³ A PubMed based literature search revealed

Fig. 3 – Optical coherence tomography (Right eye) showing a normal macula.

Fig. 4 – Optical coherence tomography (Left eye) showing a complete macular hole.

only 3 case reports of macular hole surgery associated with an optic pit, thus signifying the rarity of the condition.^{3–5} Though both the optic pits as well as the macular hole was diagnosed clinically, however we further investigated our patient with an OCT and fundus photographs to confirm the diagnosis. We also did an VFA, as optic pits are known to be associated with several visual field defects such as centrocecal defects, para-central scotomas, arcuate defects and nasal or temporal defects out of which arcuate scotomas are the most common.⁹ But in our case the VFA was normal in the right eye. We did offer macular hole surgery to the patient for his left eye, under a guarded prognosis, but he refused. Literature too mentions the difficulties associated with such surgeries wherein in one case it was a failure,² while in another it required 3 attempts before closure could be finally achieved.³ As far as the right eye is concerned, we advised prophylactic surgery, however he refused it too and so we had to adopt a conservative approach involving a regular follow up as has been advocated by some authors too.¹⁰

Conclusion

Though optic disc abnormalities such as fibre medullares, optic disc coloboma, hypoplasia disc and optic disc drusen are not very rare, however optic disc pits are rare while bilateral pits are even more so. An asymptomatic optic pit can just be monitored with routine ocular examinations. The patient should be given a home Amsler grid and instructed on how to use it, watching for the development of any metamorphopsia. In case of any symptoms the various treatment alternatives

for optic disc pit include laser photocoagulation, macular buckling surgery, vitrectomy with or without internal limiting membrane (ILM) peeling, gas tamponade or their combination.¹¹ In case of an very rarely associated macular hole, three-port pars plana vitrectomy followed by peeling of the ILM and tamponade with octafluoropropane C3F8 (14%)/silicon oil should be tried. Though the prognosis in such cases is always grim however successful restoration of the remaining central visual acuity will not only improve their vision, but also their quality of life.

Conflicts of interest

All authors have none to declare.

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