Abstract

We report the case of a 26 year old lady, presenting with complaints of gradual diminution of vision in her right eye (RE). Best corrected visual acuity (BCVA) was 20/60 and N12 in the RE and 20/20 and N6 in the left eye (LE). Fundus examination and optical coherence tomography (OCT) led to a diagnosis of optic disc pit maculopathy (ODP-M) in the RE. Pars plana vitrectomy, with an attempt to make an inner retinal fenestration adjacent to the temporal margin of the pit was done. An iatrogenic break was formed, which was used for subretinal fluid drainage and sealed by diode laser endophotocoagulation. Sulphur hexafluoride (SF6) was used for internal tamponade. At 6 months follow-up, BCVA in the RE had improved to 20/30 and N8. OCT revealed no subretinal fluid and decreased fluid in the schisis cavity. This case report therefore, demonstrates the efficacy of surgical intervention for ODP-M.

Keywords: retinoschisis, optic disc pit maculopathy, surgical intervention

Introduction

Optic disc pit (ODP) is a rare congenital excavation of the optic nerve head and is considered as part of a spectrum of congenital cavitary anomalies. Optic disc pits are seen approximately 1 in 10000 eyes. They are typically unilateral, but may be bilateral in up to 15% of patients. About 70% of the pits are located on the temporal side of the disc. Optic disc pit maculopathy (ODP-M) is a term used to describe macular changes that occur in the context of an ODP, which include intraretinal and subretinal fluid accumulation, and retinal pigment epithelial changes. It occurs in 40–50% of patients with an ODP. The prognosis of ODP-M is poor if left to its natural course.

Case History

A 26 year old Indian lady presented with complaints of gradual, progressive diminution of vision in her right eye (RE) since last 6 months. On examination her best corrected visual acuity (BCVA) was 20/60 and N12 in the RE and 20/20 and N6 in the left eye (LE). Anterior segment examination and intraocular tension was within normal limits in both eyes. Fundus examination of the RE revealed an optic disc pit on the temporal side and internal limiting membrane striae and edema in the macular area (Figure 1). Optical coherence tomography (OCT) revealed retinal edema, cystoid spaces and schisis in multiple layers with central macular thickness of 563 microns (Figure 2). The diagnosis of ODP-M was made. The history of progressively decreasing vision indicated need for an intervention. The patient underwent a 23 gauge pars plana vitrectomy with posterior vitreous detachment induction (Figure 3). Internal limiting membrane was not peeled. After this an attempt was made to make an inner retinal fenestration at the temporal margin of the optic disc with a bent 25 gauge needle. This led to an iatrogenic break formation. Fluid air exchange was done and a soft tip needle was used to drain some of the subretinal fluid from the break (Figure 4). Diode laser endophotocoagulation was done to the break and the temporal margin of the optic nerve head. Internal tamponade was done with 20% Sulphur hexafluoride (SF6) gas with prone positioning for 2 weeks. The post operative period was uneventful. On 15th post operative day, OCT revealed decreased central foveal thickness to 464 microns. There was no sub-retinal fluid and the communication of the schisis cavity with the optic nerve head had been sealed by the laser scars (Figure 5). At 6 months post surgery, her BCVA in the RE was 20/30 and N8. OCT revealed a central macular thickness of 355 microns. No sub-retinal fluid was present (Figure 6). The central macular thickness had decreased by 208 microns (Figure 7).

Figure 1: Color Fundus Photograph of the right eye showing Optic disc pit at the temporal margin.
Case Report

Discussion

Lincoff described the association between ODP and schisis-like separation of the macula. He suggested that fluid initially enters the retinal stroma to form a schisis, and then enters the sub-retinal space causing a serous retinal detachment. Several possible sources have been suggested for the sub-retinal fluid, including cerebrospinal fluid, liquefied vitreous or leaky vessels at the base of the pit. The role of the vitreous in the pathogenesis of ODP-M is important and vitreous abnormalities such as vitreomacular adhesion, partial or complete PVD, and condensed vitreous strands firmly attached to the ODP area have been found in 74% of the patients. Thus, the release of vitreous traction by vitrectomy, scleral or macular buckling has been employed widely for the treatment of ODP-M. Vitrectomy combined with gas tamponade, endophotocoagulation, silicone oil, ILM peeling, or subretinal fluid drainage have also been performed with a relatively high success rate.

Inner retinal fenestration for ODP-M was first described by Spaide. He used a bent 25 gauge needle to make a partial thickness fenestration in the retina, adjacent and temporal to the optic disc pit. His hypothesis was to provide an additional pathway for the retinal fluid to drain into the
vitreous cavity. After 1 year of follow-up, the patient showed marked improvement. In a recent study, similar procedure was done on 18 eyes with ODP-M. Complete resolution of fluid in and under the fovea was achieved in 94% of cases.

Review of literature for inner retinal fenestration showed very promising results. So we decided to undertake the procedure. Although we were not able to make only a partial thickness fenestration in the inner retina, the iatrogenic break that was formed was used to drain the subretinal fluid. Further, diode endophotocoagulation applied to the break and the temporal margin of the disc sealed the communication of the macula with the optic disc pit. Once the communication was sealed, the active RPE pump started pumping out the excess fluid and the final central macular thickness decreased by 208 microns at 6 months follow-up.

A case report has been published by Jalil A et al, where they describe the draining of the sub-retinal fluid in ODP-M using a 42-gauge subretinal canula connected to a “back-flush” flute handle. In this case a 42 Gauge needle was used to make the retinotomy and retinopexy was not performed at the site of drainage. This technique resulted in very favourable outcomes. Using a 42 Gauge needle for trying an inner retinal fenestration can have an added advantage. If an iatrogenic break is formed while trying to do the fenestration, then retinopexy would not be required and the break would be self sealing. The outcome of our case can be applied in two ways in the surgical management of ODP-M. Firstly, we can plan making a small retinotomy at the temporal margin of the optic disc pit. This retinotomy can be used to drain the subretinal fluid, which because of its long standing nature is viscous and is not efficiently drained by the RPE pump. After drainage, diode laser endophotocoagulation done to the temporal margin of the disc would lead to sealing of the communication of the optic disc pit and macula. With no new fluid flowing into the retinal layers, eventually the RPE would be able to pump out the remaining residual fluid out of the macula.

Secondly, when planning an inner retinal fenestration, occasionally an iatrogenic break can form. This break should be used to drain the sub-retinal fluid followed by diode laser endophotocoagulation to seal the communication. Further, use of a 42 Gauge needle for fenestration can alleviate the need to do a retinotomy in case an iatrogenic break forms.

We have had this one case with favourable outcome. It is required that this procedure be performed on a large number of cases to better analyse and understand the outcome and complications related to this procedure.

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