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Case Report

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A Patient With Optic Nerve Pit Maculopathy Successfully Treated With Juxtapapillary Laser Photocoagulation

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ABSTRACT

Optic disc pits are rare and congenital or acquired anomalies of the optic disc, usually remaining asymptomatic. However, serous macular detachment or optic disc maculopathy is the most common complication, causing significant visual deterioration, without a current consensus about treatment. A 55-year-old woman with a past medical history of diabetes mellitus and systemic hypertension was referred for an abnormal finding in the retina. The Corrected Distance Visual Acuity (CDVA) was 20/40 in the right eye and 20/30 in the left eye. Marcus-Gunn was negative and Slit-lamp biomicroscopy revealed no pathologic findings in both eyes. Funduscopic examination showed an excavation in the inferotemporal part of the Optic Nerve Head (ONH) with serous macular detachment extending to the optic disc. Based on clinical examination and paraclinical imaging fluorescein Angiogeraphy (FAG) Optical Coherence Tomography (OCT), optic pit maculopathy was diagnosed and the patient underwent Juxtapapillary Laser Photocoagulation (JLP). After 2 years of follow-up, there were anatomical and functional improvements.

Introduction

Optic Nerve Pit (ONP) is a rare congenital or acquired disorder of the optic nerve head with an incidence of approximately 1 in 10,000 eyes that may progress to the ONP maculopathy in 25% to 75% of the patients [1, 2]. ONP can cause visual symptoms or can be incidentally found in ophthalmic examinations. ONP maculopathy can decrease final visual acuity less than 20/200 in 80% of eyes [2-4]. Various modalities have been proposed for the management of ONP maculopathy, including Pars Plana Vitrectomy (PPV). During PPV, laser photocoagulation and the injection of sulfur hexafluoride (SF6) or perfluoropropane (C3F8) as a tamponade have been tried in many studies [5-7].

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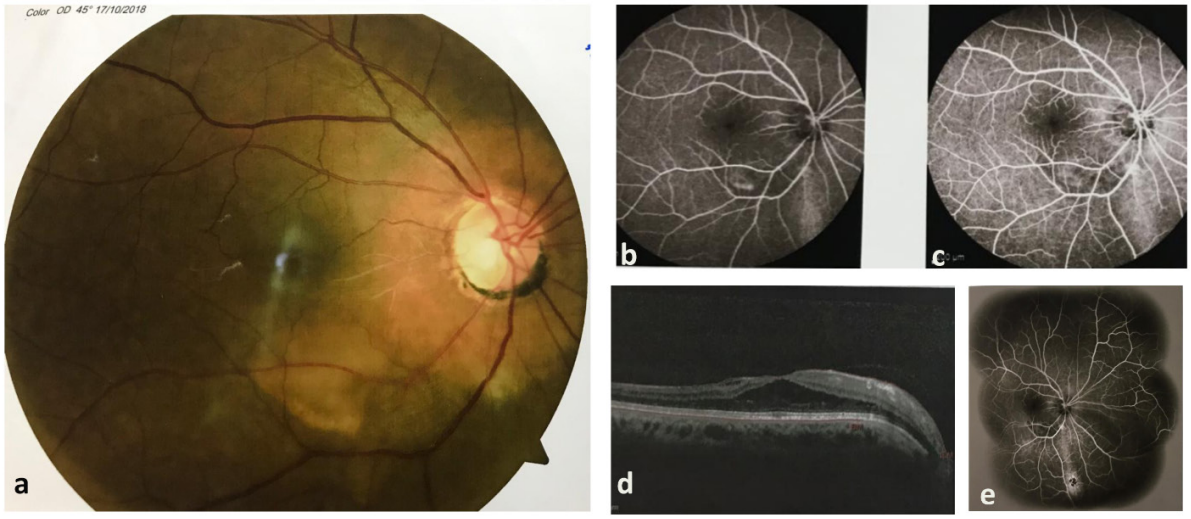

Figure 1. Color fundus (a) of the right eye shows a white area of excavation in the temporal part of the optic disc. Retinal pigmentary changes are observed along the inferotemporal arcade. Early and mid-arteriovenous phases of fluorescein angiography show a blockage area adjacent to the Retinal Pigment Epithelial (RPE) alterations compatible with chronic serous retinal detachment. Mild intraretinal leakage is also noted in the inferotemporal part. (b, c) Optical Coherence Tomography (OCT) from the macula shows schisis areas separating outer nuclear and outer plexiform layers. (d) Wide-field fluorescein angiography in the late phase shows the lack of leakage confirming no cystoid macular edema associated with diabetic retinopathy.

Considering the rare nature of ONP maculopathy, various modalities have been reported with different success rates. However, the best management protocol is still a controversial issue. In a recent systematic review, investigating the best treatment modality in patients with ONP maculopathy, the authors reported a positive publication bias toward the PPV [8].

Herein, we report a case of the ONP maculopathy treated successfully with Juxtapapillary Laser Photocoagulation (JLP). The successful standalone use of JLP has been previously but not very frequently reported.

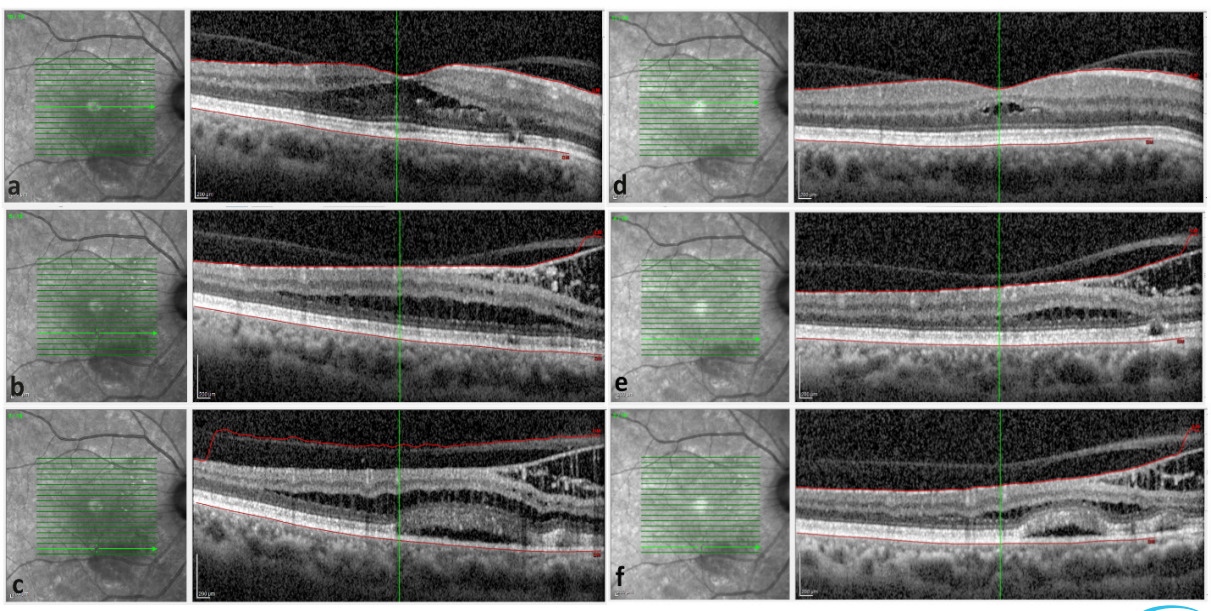

Figure 2. Optical coherence tomography before juxtapapillary laser photocoagulation (a, b, c) and after 6 months (d, e, and f)

The corresponding slabs have been chosen to demonstrate the exact changes. Laser scars at the level of retinal pigment epithelium-choroid (e and f).

Case Presentation

A 55-year-old woman was referred to our retina clinic for evaluation of an abnormal finding detected in her first ophthalmic check-up examination for Diabetes Mellitus (DM). She noted DM for 5 years, which was under control with oral agents. Systemic hypertension was also present for 5 years. No other systemic or ocular history was noted.

Using a Snellen chart, the Corrected Distance Visual Acuity (CDVA) was 20/40 in the right eye refracted with -0.75 - 1.00×130 and 20/30 refracted with -0.5 - 0.5×65 in the left eye. The pupil's reactions to light were normal in both eyes. The relative afferent pupillary defect was not present. Slit-lamp biomicroscopy revealed no pathologic findings in both eyes. In the funduscopy, an excavation was observed in the inferotemporal Optic Nerve Head (ONH). Subretinal fluid and intraretinal fluid extending from the ONH to the posterior pole dominantly distributed in the inferior part was also evident. A sclerotic cilioretinal artery was also observed (Figure 1A).

The fluorescein angiography revealed the areas of blockage and retinal pigment epithelial alterations compatible with chronic SRF, most dominantly along the inferior arcade and lack of late pooling in the cystic spaces of macula compatible with schisis (Figure 1B, C, and E). Optical coherence tomography confirmed the presence of optic pit maculopathy (Figure 1B). No microaneurysm or area of capillary non-perfusion was observed in the Fluorescein Angiography (FA). The patient underwent JLP for one session. The argon laser was used and the laser parameter setting involved the argon laser with exposure duration of 100 ms, size of 100 microns, and power of 70 mW.

The patient has been followed for 2 years. The CDVA reached 20/30 after 6 months. Anatomical improvement has also been achieved (Figure 2). Both anatomical and functional improvements remained stable during the follow-up and macular edema or diabetic retinopathy have not been progressed.

Discussion

Untreated ONP maculopathy could result in irreversible visual loss secondary to cystoid changes and Retinal Pigment Epithelial (RPE) alterations [2, 9, 10]. Macular involvement necessitates treatment to prevent visual impairment [3]. Our patient was successfully treated with the use of JLP instead of PPV. Although the reported success rates widely vary between different studies, JLP can be considered as the first treatment modality in patients with ONP maculopathy.

The origin of the fluid and the underlying mechanism of maculopathy is still unclear. The presence of mucopolysaccharide that primarily belongs to the vitreous, and transition of silicone oil or gas in the subretinal space after PPV, can support the vitreous cavity as the origin of the fluid [11-14].

Another plausible mechanism is the accumulation of fluid in intraretinal or subretinal spaces through direct communication between subarachnoid and subretinal spaces. While this hypothesis is supported by the migration of air-bubble into the subarachnoid space and also in the OCT images, various studies failed to reveal a direct communication [15-21]. Choroid and retinal vessels have also been postulated as the potential source of the fluid [2, 22]. However, these hypotheses are not strongly supported by experimental evidence and have not been observed in other retinal pathologies.

Regardless of the origin of the fluid, different modalities have been tried in the management of ONP maculopathy, such as PPV, injection of gas tamponade, JPL, macular buckling surgery, and internal limiting membrane peeling. Recently, more novel approaches have also been successful, including the use of fibrin or autologous blood sealants over the pit, ocriplasmin, radial optic neurotomy, or inner retinal fenestration [23-28]. The rare nature of the disease and the unknown origin of the fluid are among the factors leading to uncertainty about the best treatment choice. However, it seems reasonable to start with less an invasive treatment, like laser photocoagulation. The laser scar, which is applied in the temporal part of the optic disc could serve as a barrier to the fluid entrance into the macula [2, 9, 29].

A recent systematic review, investigating the surgical treatment of ONP maculopathy reported no effects for concurrent use of tamponade. The interesting point of this systematic review was the detection of publication bias toward the positive results. It means that PPV studies with extremely positive results are published more than PPV with negative results [8]. Therefore, we postulate that the higher success rates of PPV could be attributed to this bias in the literature.

In conclusion, we reported a patient with ONP maculopathy that was successfully treated with JLP. Regarding the various treatment modalities, JLP seems a reasonable initial treatment.

Ethical Considerations

Compliance with ethical guidelines

There were no ethical considerations to be consideredin this research.

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Conflict of interest

The authors declared no conflict of interest.

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