



A Case of Multiple Optic Disc Pits: 21-Year Follow-up

Osman Melih Ceylan, Alper Can Yılmaz, Ali Hakan Durukan, Mehmet Talay Köylü,
Fatih Mehmet Mutlu

University of Health Sciences Turkey Gülhane Faculty of Medicine, Department of Ophthalmology, Ankara, Turkey

Abstract

Optic disc pits (ODP) are an uncommon congenital abnormality. Patients remain asymptomatic unless they develop maculopathy. The use of optic coherence tomography has critical benefits in the follow-up of patients who are at the amblyogenic age. The aim of this study is to present a case of double ODP in the right eye and single ODP in the left eye in a partially accommodative esotropia patient followed for 21 years. To our knowledge, multiple ODP has never been described in a patient with partially accommodative esotropia.

Keywords: Accommodative esotropia, optic disc pit, optical coherence tomography, multifocal electroretinography

Introduction

An optic disc pit (ODP) is a rare congenital defect that usually presents as an ovoid, grey-white excavation in the lamina cribrosa of the optic disc.¹ It is seen in 1 per 11,000 population and equally in both sexes, occurring singly and unilaterally in 85-90% of cases and bilaterally in 10-15% of cases.^{1,2} Serous macular detachment is estimated to affect 25-75% of patients with ODP.² Vitreomacular traction and vitreous strands over the optic disc were reported by Theodossiadis et al.³ in eyes with ODP-related maculopathy. However, there are very few studies showing that this rare, sight-threatening anomaly can sometimes be multiple. Only 12 cases of double ODP have been reported in the literature to date.^{4,5,6}

The aim of this study was to present a case of double ODP in the right eye (RE) and single ODP in the left eye (LE) of a patient with partially accommodative esotropia who was followed-up for 21 years.

Case Report

A 25-year-old female was followed-up for partially accommodative esotropia from the age of 4 years. She underwent strabismus surgery at 6 years of age for residual esotropia at distance and near with full cycloplegic refraction. Bilateral ODP was found at her first visit. Her best-corrected visual acuity (BCVA) was 20/20 (Snellen chart) in both eyes. Anterior segment examination was unremarkable bilaterally. Intraocular pressure was 16 mmHg in both eyes. Cycloplegic refraction was +2.50 (+1.00x100) in the RE and +2.00 (+0.50x90) in the LE. Dilated funduscopy revealed a double ODP in temporal and nasal rims of the right optic disc (Figure 1A) and single ODP in nasal rim of left optic disc (Figure 1B). The ODPs were also clearly visible in red-free fundus photography (Figure 1C, 1D).

Spectral-domain optical coherence tomography (SD-OCT) (Heidelberg Engineering, Heidelberg, Germany) showed subfoveal deposits in the RE (Figure 2A). This accumulation

Address for Correspondence: Osman Melih Ceylan, University of Health Sciences Turkey Gülhane Faculty of Medicine, Department of Ophthalmology, Ankara, Turkey
E-mail: drmelihceylan@yahoo.com.tr **ORCID-ID:** orcid.org/0000-0002-8832-8013

Received: 08.09.2020 **Accepted:** 24.11.2020

Cite this article as: Ceylan OM, Yılmaz AC, Durukan AH, Köylü MT, Mutlu FM. A Case of Multiple Optic Disc Pits: 21-Year Follow-up.
Turk J Ophthalmol 2021;51:123-126

probably occurred after subretinal fluid resorption and spontaneously regressed (Figure 2B). The macula was stable in the LE. Swept-source OCT (Topcon Corp, Japan) through the optic disc showed two distinct hyporefective areas in the RE suggestive of ODPs, one each in the temporal and nasal quadrants, as well as fluid accumulation under the optic nerve head and intrapapillary septum structure (Figure 2C). In the LE, a shallow ODP located nasally and associated vitreous fibers were seen (Figure 2D).

The patient's 30/2 visual field analysis (Humphrey field analyzer, Carl Zeiss Meditec, Dublin, CA) demonstrated an enlarged blind spot in the RE (Figure 3A) and no visual field defect in the LE (Figure 3B). Multifocal electroretinography (mfERG) (Vision Monitor, Monpack 3, Metrovision, France) revealed low amplitudes correlated with 2° of macula in the RE (Figure 3C) and normal results in the LE (Figure 3D). However, despite the pathological findings in the tests, the patient had no complaints. The patient was followed up annually by ophthalmic examination and OCT evaluation. Her BCVA remained stable (20/20) 21 years after the initial diagnosis.

Discussion

ODPs have typically been an incidental finding on routine dilated fundus exam. To our knowledge, multiple ODP has never been described in a patient with partially accommodative esotropia.

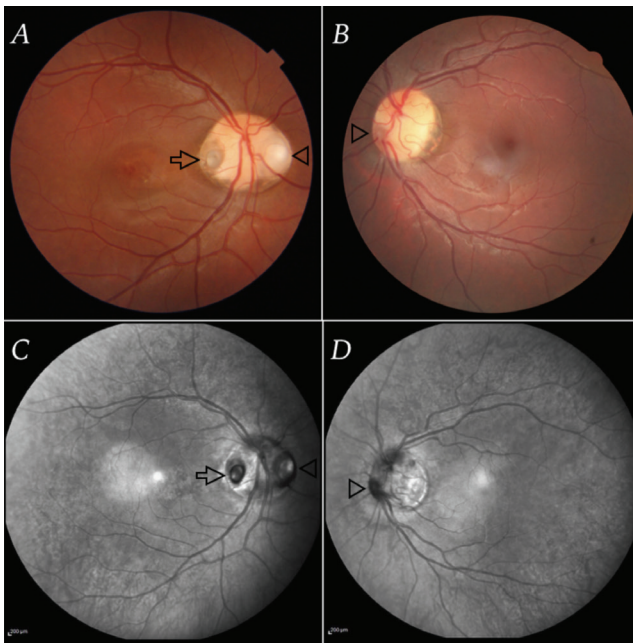


Figure 1. Color fundus and red-free photographs of the right and left eye. A, B) Two optic disc pits located temporally (arrow) and nasally (arrowhead) in the right eye and one optic disc pit located nasally (arrowhead) in the left eye. C, D) The double optic disc pit in the right eye and single optic disc pit in the left eye are clearly observed

In differential diagnosis, congenital optic disc anomalies (such as optic nerve hypoplasia, megalopapilla, morning glory syndrome, and coloboma), and acquired ODP (as in glaucoma, high myopia) should be eliminated.^{7,8} In the differential diagnosis, it is easy to distinguish congenital and acquired ODP from optic disc coloboma. Optic disc coloboma typically affects the inferior nasal rim of the optic nerve while ODP most commonly affects the inferotemporal quadrant of the optic disc.¹ Patients often remain asymptomatic until macular changes are present. Interestingly, we diagnosed this case in the first decade, and the patient was asymptomatic despite findings of maculopathy in long-term follow-up. Despite attenuated amplitudes on mfERG, her BCVA was not affected. In our opinion, her BCVA may have been preserved as a result of self-healing serous retinal detachment attacks. Although spontaneous resolution with good visual acuity was reported in about 25% of cases, pediatric patients often develop maculopathy due to traction from the formed vitreous in younger eyes.⁹

Maculopathy secondary to ODP is treated with juxtapapillary laser photocoagulation (JLP), pars plana vitrectomy (PPV), or combined treatments, but there is no consensus on the optimal surgical technique. It has been reported that the combination of PPV, gas tamponade, and JLP is more effective than PPV and gas.¹⁰ Recent studies have reported no additional benefit from JLP in long-term success rates.^{11,12} Avci et al.¹³ stated that PPV gives the best functional results in ODP maculopathy. They also emphasized that JLP may not be necessary for the success of PPV. Although there are studies showing that gas tamponade removes retinal and subretinal fluid from the macula,^{14,15} there are also studies reporting that it does not significantly contribute to the final success rate.^{10,16}

OCT is a non-invasive test to interpret the macular status, however it shows the relationship of ODP with vitreous and retina as well.^{3,17} In some cases, a hyporefective area within the optic disc excavation is visible, which may reflect accumulated fluid underneath the optic nerve head.⁹ Other important OCT findings are intrapapillary cavities, intrapapillary proliferations, septum-like structures, and subretinal precipitates which can be detected as a marker for chronicity of maculopathy.^{18,19} In our patient, one eye had fluid accumulation under the optic nerve head and an intrapapillary septum structure (Figure 2C).

Although macular and optic nerve head-related findings can be easily detected with OCT, the use of mfERG may be beneficial in patients who are uncooperative and at the amblyogenic age.

To the best of our knowledge, a total of 3 ODPs with bilateral involvement has never been described in association with partially accommodative esotropia. Therefore, in order to prevent amblyopia, patients should be closely followed from childhood. OCT and mfERG are useful tests for detecting retinal changes.

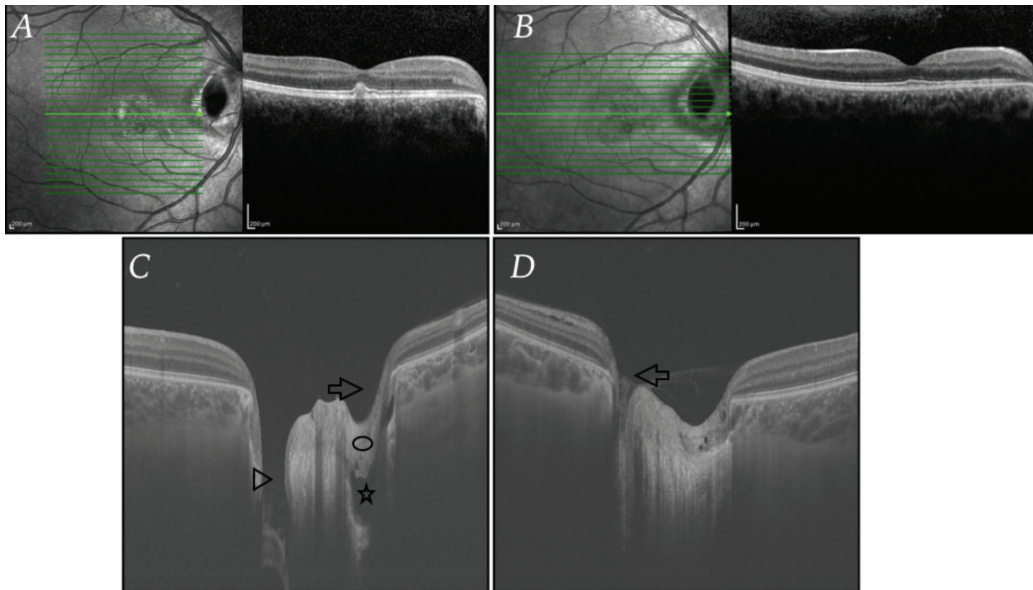


Figure 2. A) Spectral domain optical coherence tomography (SD-OCT) shows subfoveal deposits at age 19. B) Subfoveal deposits are not seen on SD-OCT at final follow-up 6 years later. C) Swept-source OCT (SS-OCT) of the right eye shows the temporal (arrowhead) and nasal (arrow) optic disc pits. Fluid under the optic nerve head appears as a hyporeflective area (star) and an intrapapillary septum structure is seen between the accumulation of fluid under the optic nerve head and the optic disc pit (circle). D) SS-OCT of the left eye shows a shallow optic disc pit (arrow) located nasally and associated vitreous fibers

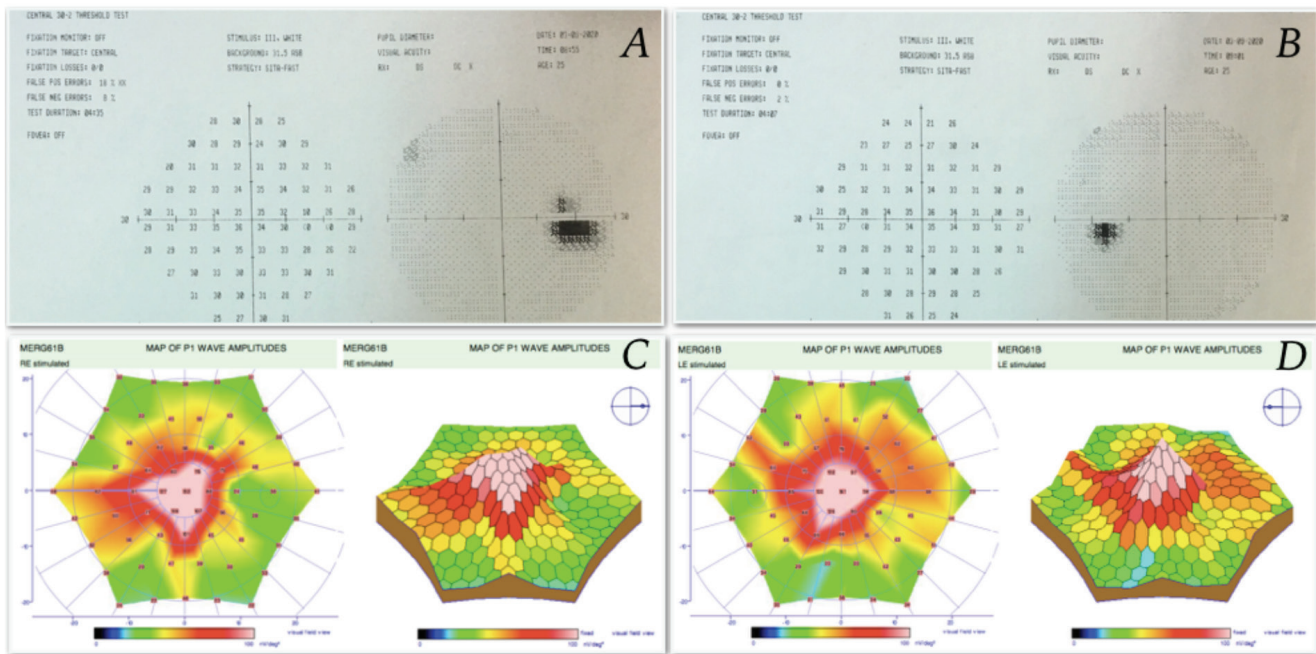


Figure 3. A) In the visual field, blind spot enlargement is observed in the right eye. B) No visual field defect is seen in the left eye. C) Multifocal electroretinography (mfERG) revealed low amplitudes correlated with 2° of the macula in the right eye. D) mfERG is normal in the left eye

Ethics

Informed Consent: The authors certify that they have obtained all appropriate patient consent forms. The patient consented to the reporting of clinical information and images under the condition of anonymity.

Peer-review: Externally and internally peer reviewed.

Authorship Contributions

Surgical and Medical Practices: O.M.C., Concept: O.M.C., F.M.M., A.H.D., Design: O.M.C., F.M.M., A.H.D., Data Collection or Processing: O.M.C., A.C.Y., M.T.K., Analysis

or Interpretation: O.M.C., A.C.Y., M.T.K., Literature Search: O.M.C., A.C.Y., M.T.K., Writing: O.M.C., F.M.M., A.H.D.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Kranenburg EW. Crater-like holes in the optic disc and central serous retinopathy. *Arch Ophthalmol*. 1960;64:912-924.
2. Georgalas I, Ladas I, Georgopoulos G, Petrou P. Optic disc pit: a review. *Graefes Arch Clin Exp Ophthalmol*. 2011;249:1113-1122.
3. Theodossiadis PG, Grigoropoulos VG, Emfietzoglou J, Theodossiadis GP. Vitreous findings in optic disc pit maculopathy based on optical coherence tomography. *Graefes Arch Clin Exp Ophthalmol*. 2007;245:1311-1318.
4. Pozza E, Postelmans L. Double optic pit and associated maculopathy. *J Fr Ophthalmol*. 2018;41:884-885.
5. Boese EA, Huang D, Tehrani S. Unilateral double optic nerve head pits. *Ophthalmology*. 2018;125:458.
6. Babu N, Baliga G, Kohli P, Ramasamy K. Management of double optic disc pit complicated by maculopathy. *Indian J Ophthalmol*. 2020;68:663-665.
7. You JY, Park SC, Su D, Teng CC, Liebmann JM, Ritch R. Focal lamina cribrosa defects associated with glaucomatous rim thinning and acquired pits. *JAMA Ophthalmol*. 2013;131:314-320.
8. Ohno-Matsui K, Akiba M, Moriyama M, Shimada N, Ishibashi T, Tokoro T, Spaide RE. Acquired optic nerve and peripapillary pits in pathologic myopia. *Ophthalmology*. 2012;119:1685-1692.
9. Yuen CH, Kaye SB. Spontaneous resolution of serous maculopathy associated with optic disc pit in a child: a case report. *J AAPOS*. 2002;6:330-331.
10. Moisseiev E, Moisseiev J, Loewenstein A. Optic disc pit maculopathy: when and how to treat? A review of the pathogenesis and treatment options. *Int J Retin Vitreous*. 2015;1:13.
11. Steel DH, Williamson TH, Laidlaw DA, Sharma P, Matthews C, Rees J, Petrou P, Charteris DG, Charles SJ, Patel CK, Crama N, Herbert E, Jackson TL. Extent and location of intraretinal and subretinal fluid as prognostic factors for the outcome of patients with optic disc pit maculopathy. *Retina*. 2016;36:110-118.
12. Rayat JS, Rudnisky CJ, Waite C, Huang P, Sheidow TG, Kherani A, Tennant MT. Long-Term outcomes for optic disc pit maculopathy after vitrectomy. *Retina*. 2015;35:2011-2017.
13. Avcı R, Kapran Z, Ozdek Ş, Teke MY, Oz O, Guven D, Yilmaz S, Kaderli B, Durukan AH, Sobacı G, Unver YB, Akduman L, Kaynak S, Dogan I, Inan UU. Multicenter study of pars plana vitrectomy for optic disc pit maculopathy: MACPIT study. *Eye (Lond)*. 2017;31:1266-1273.
14. Lincoff H, Yannuzzi L, Singerman L, Kreissig I, Fisher Y. Improvement in visual function after displacement of the retinal elevations emanating from optic pits. *Arch Ophthalmol*. 1993;111:1071-1079.
15. Johnson TM, Johnson MW. Pathogenic implications of subretinal gas migration through pits and atypical colobomas of the optic nerve. *Arch Ophthalmol*. 2004;122:1793-1800.
16. Teke MY, Citirik M. 23 Gauge vitrectomy, endolaser, and gas tamponade versus vitrectomy alone for serous macular detachment associated with optic disc pit. *Am J Ophthalmol*. 2015;160:779-785.
17. Tzu JH, Flynn HW Jr, Berrocal AM, Smiddy WE, Murray TG, Fisher YL. Clinical manifestations of optic pit maculopathy as demonstrated by spectral domain optical coherence tomography. *Clin Ophthalmol*. 2013;7:167-172.
18. Maertz J, Mohler KJ, Kolb JP, Kein T, Neubauer A, Kampik A, Priglinger S, Wieser W, Huber R, Wolf A. INTRAPAPILLARY PROLIFERATION IN OPTIC DISK PITS: Clinical Findings and Time-Related Changes. *Retina*. 2017;37:906-914.
19. Chatziralli I, Theodossiadis G, Brouzas D, Theodossiadis P. Incidence and Evolution of Subretinal Precipitates in Optic Disc Pit Maculopathy. *Eur J Ophthalmol*. 2017;27:769-773.