

Bilateral Optic Pit Maculopathy: Case Report

İki Taraflı Optik Çukur Makülopatisi

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ABSTRACT In this paper, we present an unusual case of bilateral optic pit maculopathy. Optic disc pits occur in less than 1 in 10000 and are bilateral in 10% to 15% of cases. Approximately 50% of cases with congenital optic disc pits are associated with serous macular detachment. Long-standing serous detachments are usually associated with cystic degeneration of macula and pigment loss from the underlying retinal pigment epithelium. Hence, most eyes with serous macular detachment associated with an optic disc pit have a relatively poor visual prognosis. Here, we emphasized the clinical importance of assessment of bilateral optic disc pit.

Key Words: Macular degeneration; retinal detachment; coloboma; optic nerve

ÖZET Bu yazıda nadir görülen iki taraflı optik çukur makülopatisi ile ilgili bir olgu sunuyoruz. Optik disk çukurları 10000'de 1'den daha az görülür ve olguların %10-15'inde iki taraflıdır. Doğumsal optik disk çukuru olgularının yaklaşık %50'si seröz makula dekolmanı ile ilişkilidir. Uzun süre devam eden seröz dekolmanlar genellikle makulanın kistik dejenerasyonu ve alttaki retinal pigment epitelinden pigment kaybıyla ilişkilidir. Bundan dolayı, seröz makula dekolmanı olan gözlerin çoğunda optik disk çukuru oluşumuyla birlikte göreceli olarak kötü görsel prognoz vardır. Burada iki taraflı optik disk çukuru olan olgunun değerlendirilmesinin klinik önemini vurguladık.

Anahtar Kelimeler: Makula dejenerasyonu; retina dekolmanı; kolobom; optik sinir

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Optic disc pit (ODP), also known as atypical coloboma is attributed to incomplete closure of the fetal fissure. Optic disc pit usually occurs during the first trimester of embryogenesis.¹ It occurs in less than one in 10 000 people.² An ODP appears as an oval or round, gray, white, or yellowish crater-like depression in the optic disc. Optic disc pit is frequently situated in the temporal or infra-temporal region of the optic disc.³ Optic disc pit varies from one-fourth of the disc diameter to one-half or more of the disc diameter in size and depth, and is unilateral in 85-90% of cases.⁴ Visual acuity usually remains unaffected unless the patient develops a serous retinal detachment of the macula. Although ODP is a rare condition of the optic disc, most patients with optic pits develop serous macular detachments.¹ Here we present a patient who developed bilateral optic pit maculopathy, which is a rare abnormality.

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CASE REPORT

A 33-year-old woman presented with a complaint of blurred vision in her left eye which had started 2 months previously. Her best-corrected visual acuity was counting fingers at four meters bilaterally. Anterior segment examination was unremarkable. Her intraocular pressures were recorded as 12 mm Hg OD and 11 mm Hg OS. Indirect ophthalmoscopy and slit lamp biomicroscopy showed optic disc pit with retinal pigment epithelium (RPE) changes in the right eye, and optic disc pit along with associated serous macular detachment in the left eye (Figure 1). Both optic nerve pits were located temporally and were one-half of the disc diameter. She underwent fundus fluorescein angiography (FFA), which showed hypofluorescence at temporal sides of the optic discs due to lack of vascularization bilaterally, and hyperfluorescence at the temporal retina and macula on the right eye due to diffuse RPE changes from early stages of FFA (Figure 2, 3). The left macula showed hyperfluorescence due to RPE changes and serous detachment which started at early stage and became evident at later stages (Figure 3).

The patient was informed about the treatment and prognosis. She disapproved the recommended treatment alternatives and was followed up closely.

DISCUSSION

Optic disc pit is a rare congenital abnormality of the optic disc which does not cause complaints un-

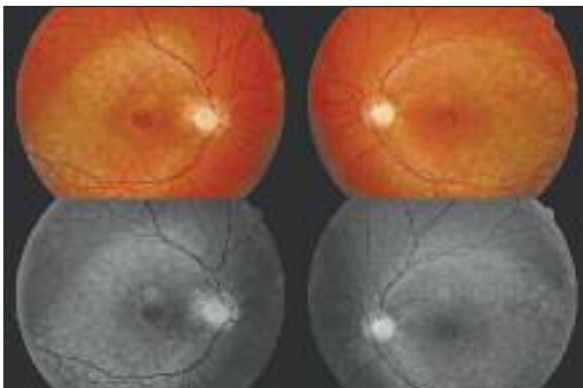


FIGURE 1: Color and red free photographs showing optic disc pit with chronic cystoid changes (OD) and serous detachment of macula (OS).

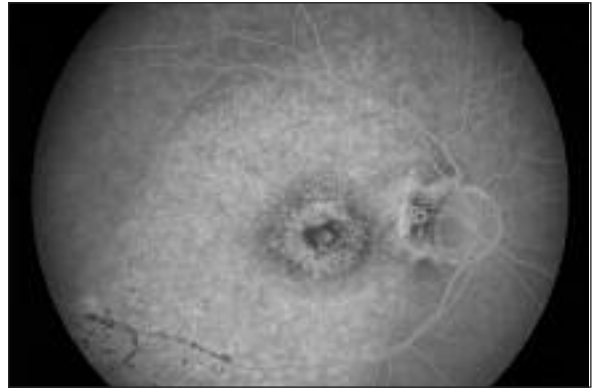


FIGURE 2: Fundus fluorescein angiography image OD.

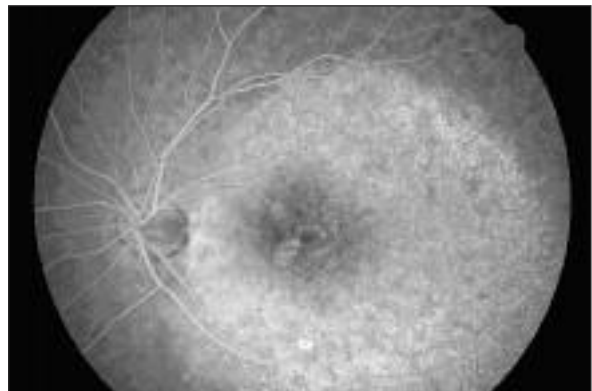


FIGURE 3: Fundus fluorescein angiography image OS.

less macular involvement occurs. In most cases, optic pits are incidentally discovered on routine fundus examination. It has been known that serous maculopathy may develop which causes serious vision loss in most patients with optic pits.⁵ Serous macular detachments that affect visual acuity are usually related to pits at the temporal region of the optic disc.⁶ The mechanism of production of the subretinal fluid associated with a serous retinal detachment is still unclear. The most popular theory suggests that fluid from the vitreous leaks through the optic pit and fills the sub-retinal space.⁷ The macular involvement occurs in 25% to 75% of eyes with an optic disc pit.¹

Although the mechanism is still unclear, it has been confirmed that the optic pit maculopathy consists of retinal detachment and inner layer retinal separation (retinoschisis). This double layered maculopathy has been attributed to the failure of

closure of the superior end of the embryonic fissure.^{8,9} A cilioretinal artery can be identified arising from the periphery of the pit. Our case, who had bilateral optic pit maculopathy, had cilioretinal artery only in the left eye.

It has been demonstrated that the damage to retina and retinal pigment epithelium is due to chronic maculopathy in ODP maculopathy, and this damage inhibits the recovery of visual acuity.⁸ No further treatment is applied, because our case had an irreversible and degenerative RPE changes due to ODP maculopathy in her right eye, and she did not accept the recommended treatment for her left eye.

Treatment includes argon laser to the peripapillary region, pneumatic displacement of the sub-

macular fluid, macular buckling surgery, vitrectomy combined with laser and/or gas injection.^{5,10,11} Attempts to repair the macular detachment and improve visual acuity should be considered before prolonged detachment resulting in irreversible degenerative changes.¹⁰

Although bilateral optic disc pit is a rare clinical condition, ophthalmologists should be aware of the lifetime-risk of maculopathy in patients with ODP and take the possibility of central visual acuity loss into consideration with this complication. They should also remember that maculopathy may develop in the eyes at different times. The patients should be informed about the vision loss and followed up closely.

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