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Case Report / *Olgu Sunumu*

# A Peripapillary Staphyloma Case Presenting with Unilateral Non-Fixing Eye

## *Tek Taraflı Fiksasyon Bozukluğu İle Seyreden Peripapiller Stafilom Olgusu*

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***Summary***

Peripapillary staphyloma (PPS) is a non-hereditary congenital optic disc (OD) anomaly. Herein, we aimed to describe a case of PPS and discuss the clinical, orbital magnetic resonance imaging (MRI) and ultrasonography (USG) findings. A one-year-old healthy girl was referred to our hospital for absence of fixation in her left eye. In addition to ophthalmological examinations, MRI and ocular USG were also performed. Sciascopic examination revealed -2.0 diopters in her right and -8.0 diopters in her left eye. Slit-lamp examination showed normal findings in both eyes. At fundus examination, peripapillary staphyloma was detected in the left eye, while the right eye showed normal findings. MRI and USG also supported the diagnosis. As a result, MRI and USG findings can support the clinical observations in the diagnosis of PPS, which should be kept in mind especially in the differential diagnosis of unilateral absence of fixation in children. *(Turk J Ophthalmol 2012; 42: 397-9)*

**Key Words:** Optic nerve, optic disc, peripapillary staphyloma

***Özet***

Peripapiller stafilom (PPS), genetik geçişli olmayan, doğumsal bir optik disk anomalisidir. Çalışmamızda, bir PPS olgumuzun klinik, orbital manyetik rezonans (MRI) ve oküler ultrasonografi (USG) bulgularını sunmayı amaçladık. 1 yaşındaki sağlıklı kız çocuğu hastanemize, sol gözü ile fiksasyon yapamadığı için gönderildi. Olgumuza tam göz muayenesine ilaveten, orbital MRI ve oküler USG tetkikleri de uygulandı. Skiaskopik muayene sonucunda sağ gözde -2.0 sol gözde -8.0 dioptri kırma kusuru saptanırken, biyomikroskopik muayene her iki gözde doğaldı. Fundus muayenesinde sağ göz normalken, sol gözde PPS saptandı. Bu tanı, MRI ve USG ile doğrulandı. Sonuç olarak PPS, çocuklarda tek taraflı fiksasyon bozukluğu ayırıcı tanısında mutlaka akla gelmeli ve MRI ve USG nin klinik bulguları destekleyici tanısal önemleri unutulmamalıdır. *(Turk J Ophthalmol 2012; 42: 397-9)*

**Anahtar Kelimeler:** Optik sinir, optik disk, peripapiller stafilom

## Introduction

Peripapillary staphyloma (PPS) is a non-hereditary congenital optic disc (OD) anomaly in which a deep excavation surrounds OD with a relatively normal appearance.[1-2] Although it is generally unilateral and the visual prognosis is poor, a bilateral case with normal visual acuities has been reported.[2-3] The etiology of the disease is thought to be associated with the decrease of the support of peripapillary structure of posterior sclera due to a problem in neural crest differentiation at the fifth gestational month.[4]

Some congenital and acquired ocular pathology like cataract formation, persistent pupillary membrane, microcornea or retinal detachment can be associated with PPS, so these patients must be followed-up regularly.[5] In spite of poor visual prognosis, visual acuity rarely increases by the occlusion of the affected eye.[5]

Herein, we report our clinical observations of a 1-year-old girl with PPS in her left eye and discuss our case with her magnetic resonance imaging (MRI) and A- and B-mode ultrasonography (USG) results.

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## Case

A 1-year-old female child was referred to pediatric ophthalmology department of Ulucanlar Eye Research Hospital. Hospital for absence of fixation in her left eye. Her medical records indicated that she was the second child of her family and systemic examination including neurological was within normal limits. Also her parents and brother are healthy and her mother had completely normal gestation period. She was born at 40 weeks of gestational age with a birth weight of 3150 gr.

Ophthalmic examination of the right eye revealed normal anterior and posterior segment elements (Figure 1), with central fixation while tracking and focusing the objects and faces. Her left eye could not fixate any objects when the right eye was occluded but she was orthophoric on Hirschberg when both eyes were open. A left relative afferent pupillary defect was present. Although she was very young, complete ophthalmologic examination could be done without sedation or anesthesia. After cycloplegia with cyclopentolate eye drops, her refractive examination with retinoscope revealed -2.0 diopters in the right and -8.0 diopters in the left eye. The fundus examination of the left eye by direct and indirect ophthalmoscopy showed PPS, pigmentary changes in retinal pigment epithelium (RPE) around the excavation (Figure 2) and normal branching retinal vessels arising from OD. Also, no choroidal or OD colobomas were detected in both eyes.

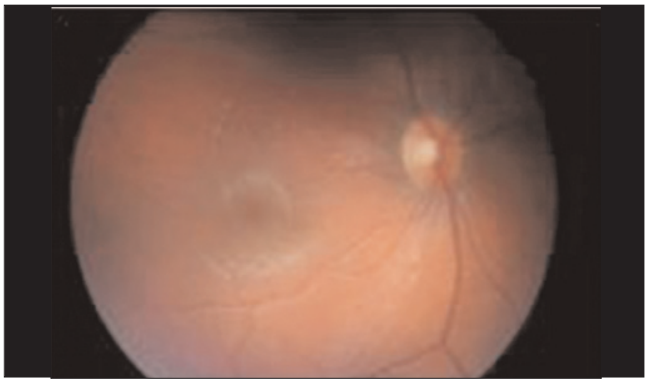

**Figure 1.** Fundus examination of the right eye shows normal findings

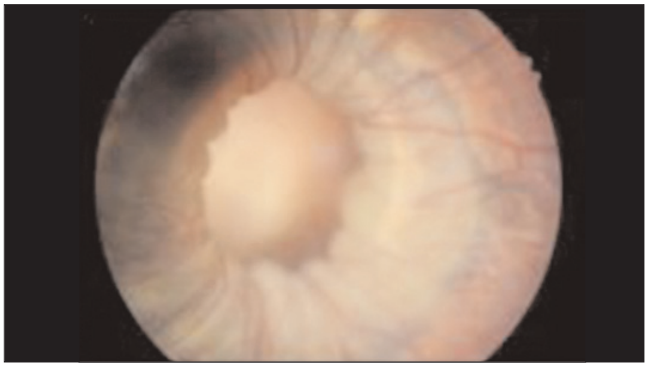

**Figure 2.** Fundus examination of the left eye shows a deep excavation around the optic disc and pigmentary changes of retinal pigment epithelium at the edge of the excavation

On sagittal and axial magnetic resonance T2-weighted images, no structural pathologies other than the continuity of the excavation on the posterior wall of the bulbus with vitreous cavity were detected (Figure 3 and 4). A- and B-mode USG showed a 10 mm long depth and 4.5 mm long width of peripapillary excavation (Figure 5) and a 29 mm long axial length.

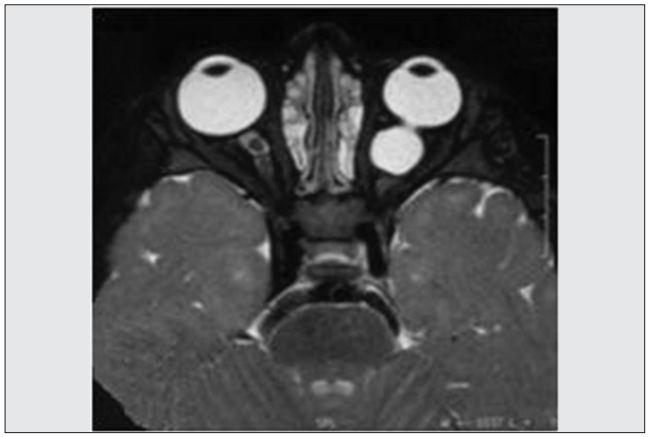

**Figure 3.** The axial magnetic resonance T2-weighted lipid-suppressed image of left orbit discloses the continuity of the excavation on the posterior wall of the bulbus with vitreous cavity

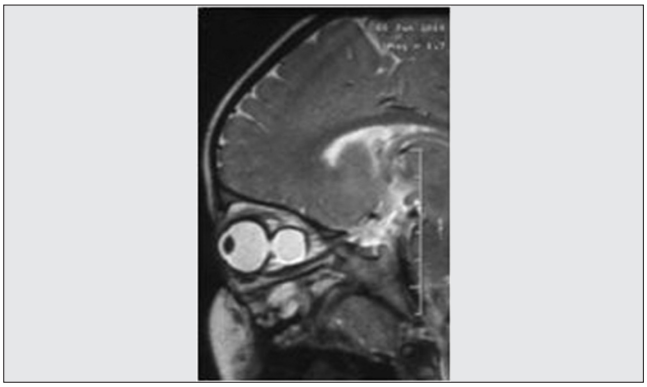

**Figure 4.** The sagittal magnetic resonance image discloses the excavation on the posterior wall of the bulbus in the left orbit

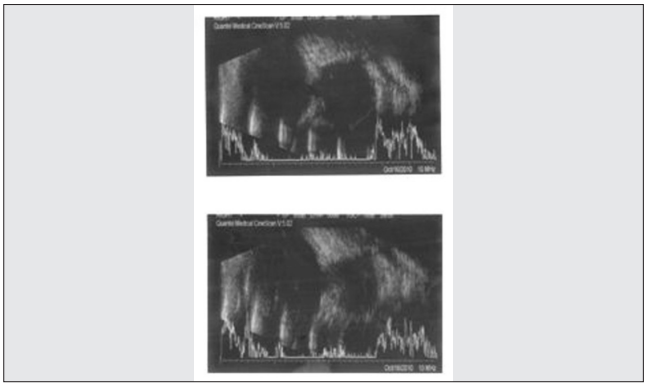

**Figure 5.** B-mode ultrasonography shows the depth and width of the excavation

## Discussion

PPS is a non-hereditary, congenital and generally unilateral disease which is characterized by an OD frequently in normal appearance in the central region of a deep excavation and normal retinal vessels with generally poor visual prognosis.[1-3,5-7] It is rarely associated with other systemic congenital diseases and choroidal or OD colobomas and is easily misdiagnosed with other excavated OD abnormalities like optic disc coloboma, morning glory syndrome, and optic pit.[1-3,5-7] The depth of staphyloma is variable and is even associated with a high level of 10mm like in our case. Also, in some cases, the fovea can be involved, such as in Kim et al.'s[5] report.

PPS can easily interfere with other congenital excavated OD abnormalities as mentioned before, and careful clinical examination is mandatory for differential diagnosis. In OD coloboma, the excavation is on the disc and is frequently associated with iris and/or choroidal colobomas and other systemic diseases. The excavation in morning glory syndrome is cone-shaped and narrower with glial remnant in OD centre.[8] In this OD anomaly, the vessels arising from large OD are in radial pattern and the number of these vessels is higher than normal levels. Also moyamoya syndrome, a disease in which certain arteries in the brain are constricted, may accompany morning glory disc anomaly.[9] Our case was referred to us for absence of fixation in her left eye and PPS was diagnosed clinically. Also MRI and USG findings supported our diagnosis and eliminated the other congenital excavated OD abnormalities.

Although visual prognosis is generally poor in PPS, Kim et al.[5] observed improvement in visual acuity by occlusion therapy. The refractive status in PPS is also paradoxical. These cases have generally emetropia or mild myopia, but Kim et al.[5] reported high myopia (-10.0 diopters) in their case series. The correlation between the depth of the excavation and refractive error or visual acuity level is not fully understood up to now. Our case had also high myopia (-8.0 diopters) in the affected eye and the depth of the excavation was 10 mm, while the width was 4.5 mm. The great dimensions of the excavation might be associated with high myopia in our case. Woo et al.[10] measured the depth of PPS by spectral-domain optical coherence tomography in their 4-year-old case and observed multiple intraretinal cystic cavities along the temporal tuft of staphyloma. Also they thought that OCT had revealed the signs of a connection between perineural space and the inner retinal layers.

The stimulation of the other eye with light may cause a contraction in PPS, but the mechanism of this action is not clear.[11-13] Farah et al.[11] mentioned a possible neuromuscular contraction mechanism for this entity. They thought that a circular, heterotopic smooth muscle situated at the posterior pole of the eye, associated with an autonomic cholinergic reflex, and innervated by a ciliary nerve might have caused these contractions. These contractile mechanisms were thought to improve in the process of time; so, we could explain the reason of the absence of these contractions in our case by her young age.

This report emphasizes the importance of careful ophthalmic examination and imaging techniques in the diagnosis of PPS. Our case had a clinically typical PPS supported by magnetic resonance imaging and ultrasonography. Early diagnosis of the disease is important because of the treatment of associated ocular problems like refractive error. In spite of poor visual prognosis in most of the cases, we must keep in mind that visual improvement can be achieved with the correction of their refractive status and occlusion therapy.

## References


1. Gottlieb JL, Prieto DM, Vander JF, Brown GC, Tasman WS. Peripapillary staphyloma. Am J Ophthalmol. 1997;124:249-51.
2. Blair MP, Blair NP, Rheinstrom SD, Jednock NJ. A case of peripapillary staphyloma. Arch Ophthalmol. 2000;118:1138-9.
3. Caldwell J B, Sears ML, Gilman M. Bilateral peripapillary staphyloma with normal vision. Am J Ophthalmol. 1971;71:423-5.
4. Pollock S. The morning glory disc anomaly: contractile movement, classification, and embryogenesis. Doc Ophthalmol. 1987;65:439-60.
5. Kim SH, Choi MY, Yu YS, Huh JW. Peripapillary staphyloma clinical features and visual outcome in 19 cases. Arch Ophthalmol. 2005;123:1371-6.
6. Sanjari MS, Falavarjani KG, Kahkouli MB.Bilateral peripapillary staphyloma, a clinicoradiological report. Br J Ophthalmol 2006;90;1326-7.
7. Brown G, Tasman W. Congenital anomalies of the optic disc. New York: Grune & Stratton; 1983:178–83.
8. Brodsky MC. Congenital optic disk anomalies. Surv Ophthalmol. 1994;39:89-112.
9. Taşkintuna I, Oz O, Teke MY, Koçak H, Firat E. Morning glory syndrome: association with moyamoya disease, midline cranial defects, central nervous system anomalies, and persistent hyaloid artery remnant. Retina. 2003; 23:400-2.
10. Woo SJ, Hwang JM. Spectral-domain optical coherence tomography of peripapillary staphyloma. Graefes Arch Clin Exp Ophthalmol. 2009;247:1573-4.
11. Farah ME, Bonomo PP,Nobrega M, Hoflin-Lima AL. Contractile peripapillary staphyloma with light stimulus to the contralateral eye. Arch Ophthalmol. 2001;119:1216-7.
12. Kral K, Svarec D. Contractile peripapillary staphyloma. Am J. Ophthalmol 1971;71:1090-2.
13. Wise JB, Maclean AL, Gass JD. Contractile peripapillary staphyloma. Arch Ophthalmol 1966;75:626-30.