

Unilateral Optic Nerve Hypoplasia and Ambliyopia

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ABSTRACT:

Unilateral optic nerve hypoplasia and amblyopia

Objective: To present a case who has amblyopia secondary to unilateral optic nerve hypoplasia.

Case: A 13 year-old girl presented with low visual acuity in the right eye. In her complete eye examination, her best corrected visual acuity was 2/20 right eye (OD) with -0.75 and 20/20 left eye (OS) with -0.75-0.25x88. The globe movements, intraocular pressure and anterior segment examination was normal in both eyes. Examination of posterior segment showed optic nerve hypoplasia (ONH) with hypopigmented ring surrounding the optic disc in the right eye. Retinal nerve fiber layer was thinner in the right eye than the left eye. Retinal vasculature was normal in both eyes. In addition to clinical examination, both eyes were evaluated with optical coherence tomography (OCT), fundus fluorescein angiography (FFA) and visual evoked potential (VEP).

Conclusion: Amblyopia secondary to ONH is not common but it should be kept in mind as a cause of amblyopia.

Keywords: Amblyopia, hypoplasia, optic nerve, optical coherence tomography

ÖZET:

Tek taraflı optik sinir hipoplazisi ve ambliyopi

Amaç: Tek taraflı optik sinir hipoplazisine bağlı olarak gelişen ambliyopi olgusu sunumu.

Olgu: On üç yaşında kız çocuk sağ gözünde az görme şikayeti ile başvurdu. Yapılan oftalmolojik muayenesinde sağ gözde görme keskinliği 2/20 (-0.75), sol gözde ise 20/20 (-0.75-0.25 *88) idi. Glob hareketleri, göz içi basıncı ve ön segment muayenesi her iki gözde normaldi. Arka segment muayenesinde sağ gözde optik sinir hipoplazisi (OSH) ve optik diskin çevresinde hipopigmente sarı halka görüldü. Retina sinir lifi tabakası sağ gözde sol gözden daha ince idi. Retinanın damar yapılanması normal izlendi. Klinik muayeneye ilave olarak optik koherens tomografi (OKT), fundus flöresan aniyografisi (FFA) ve görsel uyarılmış potansiyel (VEP) tetkikleri yapıldı.

Sonuç: OSH'ne bağlı ambliyopi sık olmamakla birlikte ambliyopinin nedenleri araştırılırken akıld tutulmalıdır.

Anahtar kelimeler: Ambliyopi, hipoplazi, optik sinir, optik koherens tomografi

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INTRODUCTION

The prevalence of childhood amblyopia is 2-5%, with strabismus and refractive errors constituting the most common causes, while the optic nerve hypoplasia is among the rare causes (1,2).

Optic nerve hypoplasia (ONH) is a non-progressive manifestation that may develop unilaterally or bilaterally, with reduced retinal nerve fiber number and a normal retinal tissue development (3). In this study, we aimed to present a case with a diagnosis of ONH, who was examined in a total of

4 hospitals with the complaint of unilateral low visual acuity.

CASE PRESENTATION

The 13-year old female patient whom in her statement explained that she was examined previously in 3 different hospitals with the complaint of low vision in her right eye, but couldn't be diagnosed, had a complete eye examination, visual evoked potential (VEP) test, optic coherence tomography (OCT) and fundus florescein angiography (FFA)

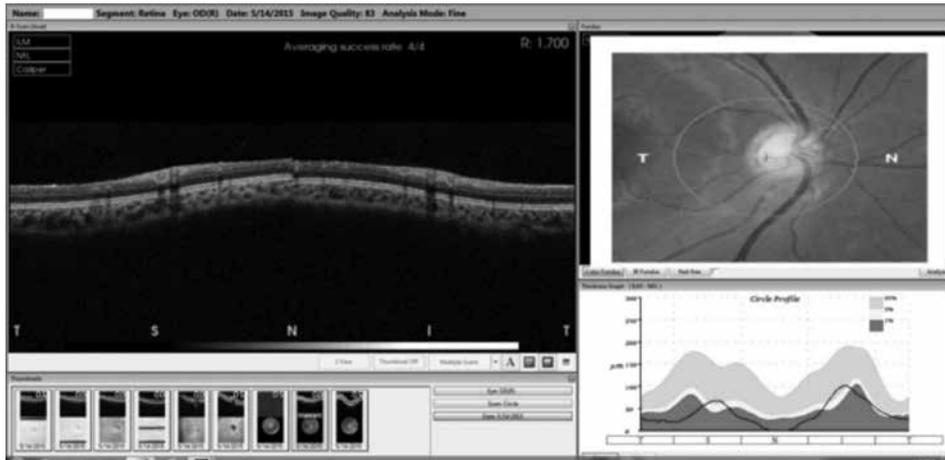


Figure-1: Hypopigmented yellow ring (double ring sign) surrounding the optic disc and the thin retinal nerve fiber layer

examinations. The patient's history was taken in terms of systemic and ophthalmologic problems. The patient's height and weight developments were evaluated. Right visual acuity was 2/20 (Snellen) with -0.75 Diopter (D), and the left visual acuity was 20/20 with -0.75-0.25x90. The cycloplegic refraction revealed -0.75 D right eye (OD) and -0.75-0.25x88 D left eye (OS). The anterior segment was normal bilaterally. Strabismus examination was observed to be orthophoric. The intraocular pressure was measured as 16 mmHg OD and 16 mmHg OS. The pachymetric measurements were 514 μm OD and 505 μm OS. The corneal endothelial cell count was 2955 OD and 2999 OS. In the right eye, the optic nerve head was observed to be smaller than in the left eye, and a yellow hypopigmented ring around the right optic disc (double ring sign) was detected, with the retinal nerve fiber layer being thinner in the hypoplastic side at OCT (Figure-1). The optic disc measurements were 1000x1200 μm OD and 1700x1500 μm OS. Although the optic disc was small, the retinal tissue and the vessel calibers were observed as normal. No pathology was detected at FFA. The patient has a normal height and weight, with no history of hormonal disorders. In her statement, she explained that her right eye vision was at the same level since she knew herself, showing no improvement or reduction. In the VEP examination,

in the right eye, she had delayed P100 latency with full-field stimuli, and with half-field stimuli, the P100 latency was lost. The latency and the amplitude was normal in the left eye with full and half-field stimuli.

DISCUSSION

Amblyopia is a frequent ophthalmological clinical manifestation at childhood, that may be due to many reasons. Strabismus and refractive errors constitute as the frequent reasons for low vision, while the ONH is among the rare causes (4). The prevalence of ONH has been reported as 7-10/100.000 (5,6). In cases with ONH, while there are predisposing factors such as a diabetic mother, excessive alcohol or anticonvulsant drug use, there was no evidence of these factors in our case's history. Our case also had no strabismus or any refractive error. In a study, ONH was detected to be at 6.3% frequency as a cause of low vision (7). It was reported that nerve fiber layer thinning accompanies unilateral ONH (8,9). Our case also showed nerve fiber layer thinning. When the ONH is bilateral, it may be associated with developmental anomalies of the central nervous system (CNS), and in 10% of cases, septo-optic dysplasia (de Morsier Syndrome) may be accompanied. No pathology related to CNS was observed in the presented case.

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