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Unilateral Optic Disc Drusen

Perihan DİNÇ [1] Fikret UÇAR [1] Servet ÇETİNKAYA [1]

Özet

Optik disk druseni (ODD), optik disk (OD) başında, hiyalin yapısında patolojik kalsifiye materyalin birikmesi ile oluşan, konjenital bir optik disk anomalisidir. Yüzeysel yerleşimli ODD'ler fundus muayenesi sırasında teşhis edilebilirken, derin yerleşimli ODD'lerin teşhisinde; B-mod ultrasonografi (USG), fundus floresein anjiografi (FFA), bilgisayarlı tomografi (BT) gibi ek tanı yöntemlerine ihtiyaç duyulur. ODD, fundus muayenesinde OD sınırlarında papillödem benzeri bir görünüme neden olur. Bu nedenle ODD erken dönem papillödem ayırıcı tanısında yer almalıdır. Bu yazıda, papillödem ön tanısı ile kliniğimize yönlendirilen tek taraflı ODD'li olguyu sunmayı amaçladık.

Anahtar Kelimeler

Optik disk druseni
Papillödem
B-USG
BT.

Makale Hakkında

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Abstract

Optic disc drusen (ODD), which is composed of accumulation of hyalin, a calcified pathological material, is a congenital optic disc anomaly. Superficial ODDs are diagnosed easily, during fundus examination, while to diagnose the buried ones, some additional diagnostic tools such as B-scan ultrasonography (USG), fundus flourescein angiography (FFA), computerized tomography (CT) are essential. ODD may be misdiagnosed as early onset papilledema, therefore it is an important clinical case. We present here a case with unilateral ODD, who is referred to our clinics with papilledema preliminary diagnosis.

Keywords

Optic disc drusen
Papilledema
B- USG
CT

Article Info

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1. Introduction

ODDs, which are located at the anterior part of optic nerve lamina, bulging forward, composed of accumulation of hyalin and calcified material, cause indefinite appearence of optic disc edge. Migration of retinal pigment epithelial cells and hyalin degeneration of neuro-glial cells are responsible for the formation of ODD. Disorders of axoplasmic transmission of retinal nerve fibers and axonal Ca metabolism are also influential on ODD pathogenesis (Lam, Morais ve Pasol, 2008; Atmaca ve Yilmaz, 2003; Kanski, 2011). In electron microscopy, standalone and/or clusters of calcified axonal mitochondrias on lamina choroidalis were observed. Calcified axonal mitochondrias are the main sources of ODD (Ford, Biller ve Weaver, 1983; Mom, 1981).

The incidence of ODD is 0.3 - 0.4% and 27% of the cases are unilateral1. Genetic transmission is present, heritance is irregular and generally it's seen in caucasians (Atmaca ve Yilmaz, 2003; Kanski, 2011). In early childhood period, it is buried in OD plane, therefore it's difficult to determine it in

[1] Konyagoz Eye Hospital, Konya, Türkiye, dinc.perihan@gmail.com
[1] Konyagoz Eye Hospital, Konya, Türkiye, fikretucar@konyagoz.com
[1] Konyagoz Eye Hospital, Konya, Türkiye, drservet42@gmail.com

ophthalmoscopical examination. In early adolescence, it is easier to diagnose ODD. Buried ODDs are most confused with papilledema. Rarely, they are diagnosed with computerized tomography (CT) incidentally (Atmaca ve Yilmaz, 2003; Mom, 1981).

2. Case Report

Our patient was female, 27 years of age, referred to our clinics with right optic neuritis or papilledema preliminary diagnosis. In ophthalmological examination, visual acuities on both eyes were 20/20, light reflexes were positive, colour vision, light sensitivity, intraocular pressures and anterior segment structures were normal on both eyes.

In fundoscopic examination, OD borders were prominent in left eye, but indefinite and irregular especially in nasal region in right eye (Figure 1a-b). In red-free imaging superior nasal part of OD showed autofluorescence, in FFA, hyperfluorescence was observed in OD. There was no pathology related to macula in both eyes (Figure 2a-b). In B-mode USG, a hyperecogenic lesion was observed on right OD and there was no pathological finding on left OD (Figure 3a-b). In orbital CT imaging, a calcified hyperdense lesion, 2x1 mm in size was observed on right OD head (Figure 4a-b).

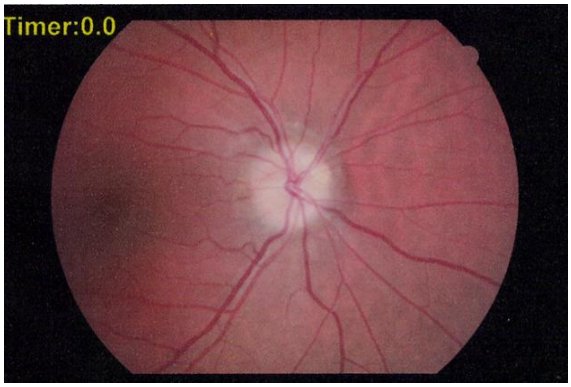


Figure 1-a Right eye optic disc appearance

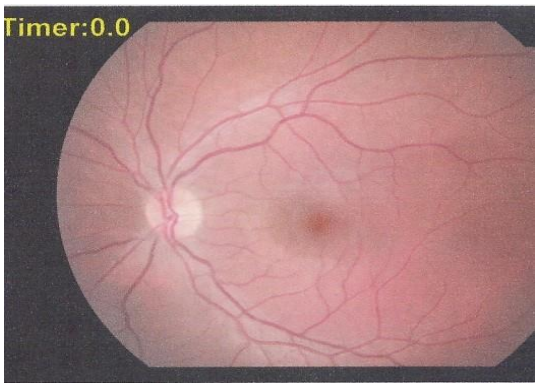


Figure 1-b Left eye optic disc appearance

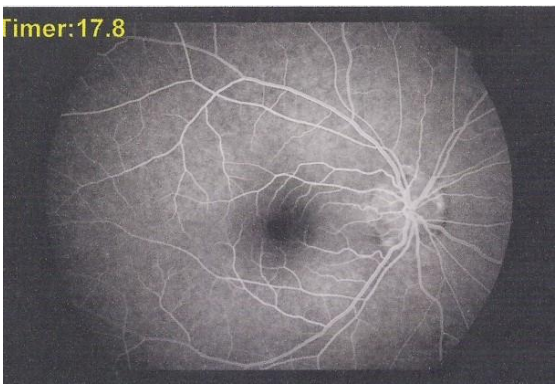


Figure 2-a Right eye FFA imaging

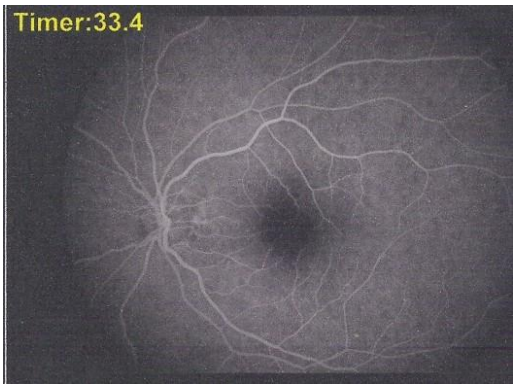


Figure 2-b Left eye FFA imaging

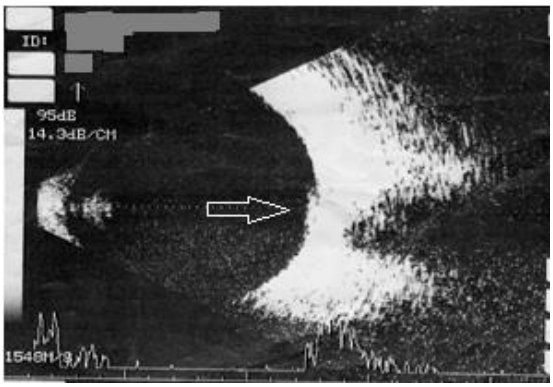


Figure 3-a Right eye B-mode USG imaging

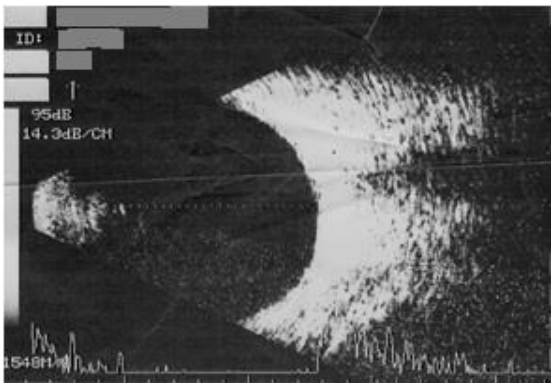


Figure 3-b Left eye B-mode USG imaging

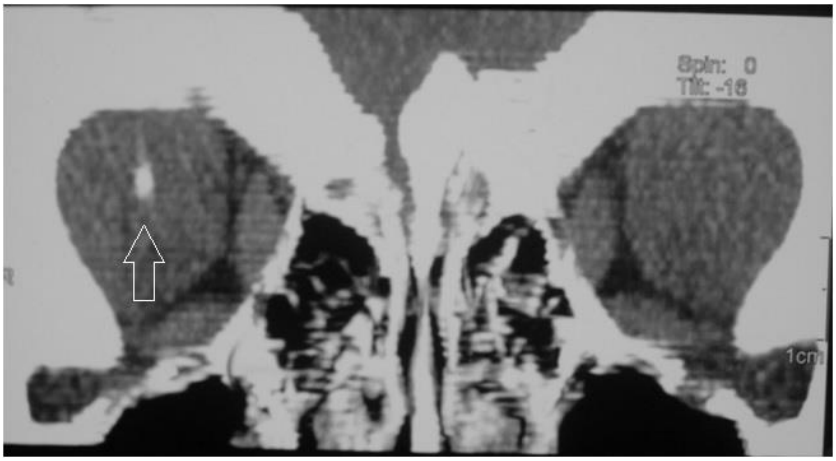
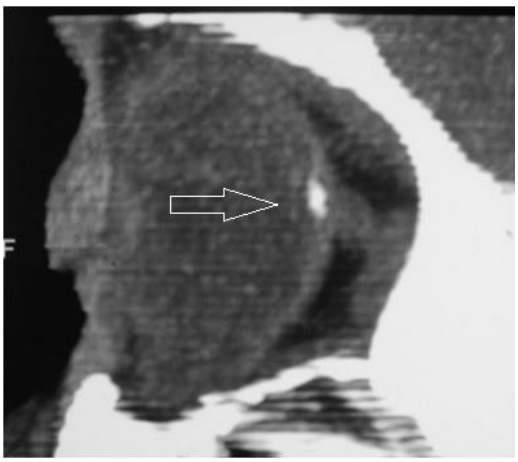


Figure 4-a Coronal orbital CT imaging



Figurel 4-b Right sagittal orbital CT imaging

In visual field analysis, general reduction of sensitivity was present in right eye and there was no problem with left eye (Figure 5a-b). In OCT imaging, OD surface area was 4.61 mm2 in right eye and 3.5 mm^2 in left eye (Figure 6a-b). There was no other ocular or systemic disorders. The case was diagnosed as right ODD.

An informed consent was obtained from the patient for this study.

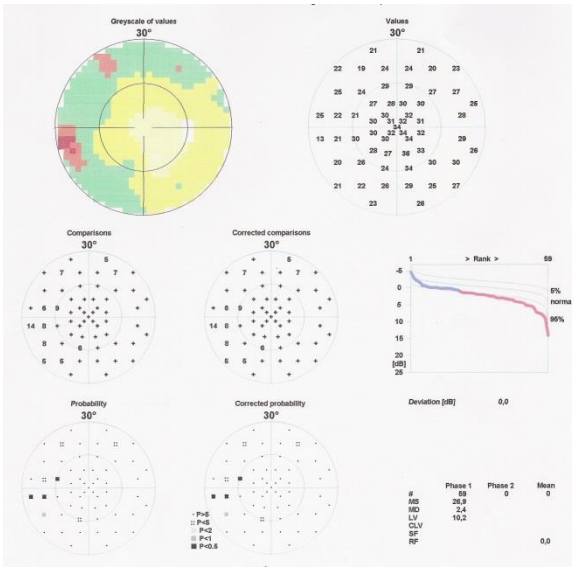


Figure 5-a Right eye visual field

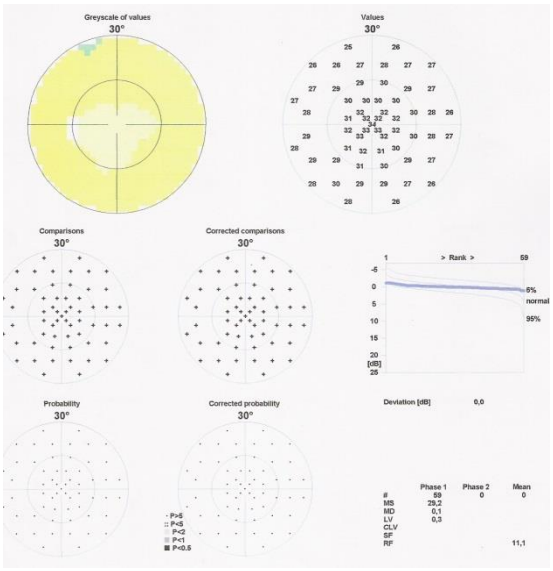


Figure 5-b Left eye visual field

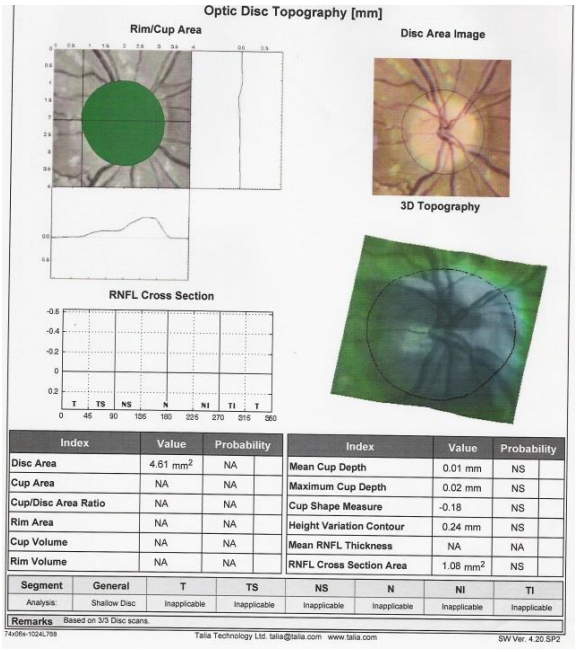


Figure 6-a Right eye optic disc topography

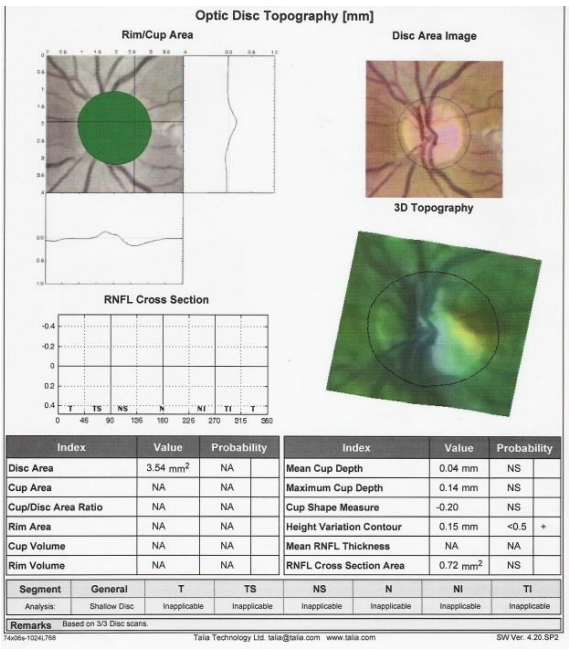


Figure 6-b Left eye optic disc topography

3. Discussion

ODD is a rarely seen optic disc anomaly, its incidence is 0.3%. Posterior embryotoxon and pigmentary retinopathy may accompany ODD. Its incidence is more in patients with retinitis pigmentosa and angioid streak. Generally it is bilateral, but also may be unilateral as in our case (Mom, 1981).

In patients with indefinite OD borders, ODD should be thought in differential diagnosis. Especially in early childhood period it may be buried and if the involvement is unilateral, ODD may easily be confused with papilledema (Kinori at al, 2013). However, there are some differences between ODD and papilledema. In ODD; OD is pink or yellow, its edges are globular, the vessels extracting from OD are branched, phisiological OD cupping is absent. In FFA, in contrast to papilledema, in early phase of ODD, hyperfluorescence is not observed. Additionally, autofluorescence in red-free imaging is not observed in papilledema in contrast to ODD (Mom, 1981; Kinori at al, 2013).

In 71% of the patients with ODD, glaucoma-like visual field defects are observed. These defects may be generalized depression, relative arcuate scotoma, enlargement of blind spot and concentric narrowing, depending on the location of ODD. These findings may be progressive. Therefore, it is difficult to determine whether these defects are originated from glaucoma or ODD. These cases may be misdiagnosed as glaucoma and treated like glaucoma (Karadeniz Uğurlu, Şefi, Maden, 2000; Ocakoglu et al, 2003; Kelbsch et al, 2019). In patients with visual field defects, OD should be examined carefully and ODD should be thought in differential diagnosis.

In patients with ODD, rarely splint hemorrhage on OD head, vitreous hemorrhage and subretinal hemorrhages may ocur (Padhy ve Behera, 2019). Very rarely, choroidal neovascular membrane around OD may occur. These membranes may spontaneously resolve or sometimes anti-VEGF therapy may be needed11,12 (Gan ve Long, 2019; Auw-Haedrich, Staubach ve Witschel, 2002).

In patients with indefinite or irregular OD borders, decreased C/D ratio, unexplained hemorrhages on OD, retina or vitreous, peripapillary choroidal membrane and nerve fiber bundle defects in visual field analysis, ODD should be thought in differential diagnosis. Especially, unilateral and buried drusens may easily be confused with early phase papilledema. Even in pediatric cases due to this confusion some investigational interventions like lumbar puncture are performed. Therefore, the patients who were diagnosed as papilledema, especially if they are young and the involvement is unilateral, ODD should be thought in differential diagnosis.

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