

# Spectral Domain Optical Coherence Tomography in Simple Retina Pigment Epithelium Hamartoma

## *Basit Retina Pigment Epitel Hamartomunda Spektral Domain Optik Koherans Tomografisi*

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### **Summary**

To report the spectral-domain optical coherence tomographic findings in a case of a simple hamartoma of the retinal pigment epithelium. A 55-year-old female patient with simple hamartoma of the retinal pigment epithelium was evaluated with ultrasonography and spectral-domain optical coherence tomography. The patient was found to have a pitch-black lesion measuring one-half of the disk diameter, located at about 1 disk diameter temporal to the foveola. B-mode ultrasonogram revealed a 0.8-mm thick retinal lesion with high internal reflectivity which was consistent with simple hamartoma of the retinal pigment epithelium. Spectral-domain optical coherence tomography demonstrated an elevated lesion extending from the retinal layer toward the vitreous. The high resolution of spectral-domain optical coherence tomography allows detailed observation of the retinal pigment epithelium hamartomas and is useful in the differential diagnosis and management of pigmented fundus lesions. (*Turk J Ophthalmol* 2012; 42: 157-8)

**Key Words:** Hamartoma, retinal pigment epithelium, spectral-domain optical coherence tomography, ultrasonography

### **Özet**

Basit pigment epitel hamartomunda spektral domain optik koherans tomografi bulgularını rapor etmek. Basit retina pigment epitel hamartomu olan 55 yaşında bayan olgu ultrasonografi ve spektral domain optik koherans tomografi ile değerlendirildi. Olguda foveolanın bir disk çapı temporalinde, yarım disk çapında koyu siyah renkli lezyon saptandı. B-mod ultrasonografide yüksek iç yansıtıcılıkta 0,8 mm kalınlıkta, basit retina pigment epitel hamartomu ile uyumlu retinal lezyon gözlemlendi. Spektral domain optik koherans tomografi ile retinadan vitreus boşluğuna doğru elevasyon gösteren kitle belirlendi. Yüksek çözünürlüklü spektral domain optik koherans tomografi retina pigment epitel hamartomlarının daha iyi gözlenmesi imkanını sunar ve pigmentli fundus lezyonlarının ayırıcı tanı ve tedavisinde yararlıdır. (*Turk J Ophthalmol* 2012; 42: 157-8)

**Anahtar Kelimeler:** Hamartom, retina pigment epiteli, spektral domain optik koherans tomografi, ultrasonografi

## Introduction

Simple retina pigment epithelium (RPE) hamartoma is a rare entity, usually incidentally diagnosed in asymptomatic eyes.<sup>1,2</sup> The lesion is believed to be congenital and termed "simple" due to the assumption that it is composed of solely proliferating RPE cells. Optical coherence tomography (OCT) findings has been described in selected retinal and RPE tumors.<sup>2-4</sup> Spectral-domain (SD) OCT is a relatively new technique which is also used in the diagnosis of pigmented retina and RPE tumors.<sup>5</sup> Herein we describe the SD OCT findings of a case with unilateral simple RPE hamartoma.

## Case Description

A 55-year-old woman was found to have a pitch-black, well-defined lesion measuring half disk diameter, located temporal to the foveola in right eye (Figure 1A). The fundus examination of the left eye was normal. Distant visual acuity was 20/20 in both eyes. B-mode ultrasonogram revealed a 0.8-mm thick retinal lesion with high internal reflectivity (Figure 1B). The SD OCT revealed a full-thickness RPE replacement, causing optical shadowing in the right eye (Figure 1C). The 3 Dimensional SD OCT images showed the regular elevated lesion from the retinal layer toward the vitreous with accompanying shadowing. The visual field and Amsler grid testings were normal in both eyes.

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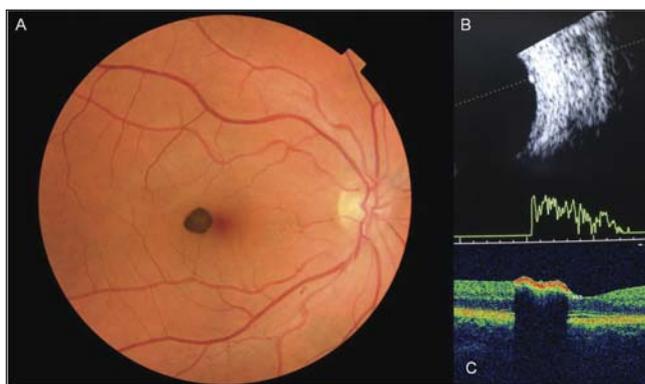
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## Comment

Gass named this entity “hamartoma”, based on superficial, full-thickness retinal involvement.<sup>1</sup> The hamartoma is presumably congenital and usually located in the macular region. Because of the stationary nature, the lesions located at the extrafoveal area are asymptomatic and usually diagnosed incidentally.<sup>1,3</sup> The parafoveal location of the lesion in our case did not cause visual loss as mentioned in earlier reports.<sup>1,3</sup>

Most reported lesions have been less than 1.0 mm in diameter and 1.0 to 2.5 mm in thickness.<sup>1</sup> Shields and associates observed dilated retinal feeding artery or draining vein (100%), adjacent mild retinal traction (80%), yellow retinal exudation (20%), and vitreous pigmented cells (20%).<sup>1</sup> Although we observed feeding and draining vessels in our RPE hamartoma we could not identify any retinal traction, exudation or pigmented cells in the vitreous.

The described differential diagnosis includes combined hamartoma of retina and RPE, congenital hyperplasia of RPE, choroidal melanoma, choroidal nevus, and reactive RPE hyperplasia. In contrast to simple RPE hamartomas combined RPE hamartomas are greyish lesions that contain glial tissue on their surfaces which causes mild to severe retinal traction.<sup>5</sup> Congenital hyperplasia of RPE lesions are well-demarcated, flat to minimally elevated fundus plaques that can range from black to completely depigmented lesions.<sup>5</sup> Most of them are located in the midperipheral or peripheral fundus, but occasionally at the macular area. Choroidal nevus and choroidal melanoma usually



**Figure 1.** **A.** The pitch-black RPE hamartoma adjacent to the fovea in the right eye. **B.** The B-scan ultrasonogram of the lesion with high internal reflectivity. **C.** The 2D-SD OCT image showing the anterior replacement of the RPE causing optical shadowing

have less prominent borders, and their color is usually are dark brown - not pitch black. Although choroidal nevus has high internal reflectivity similar to simple RPE hamartoma, choroidal melanoma has low to medium internal reflectivity on B-scan USG.<sup>5</sup> Reactive RPE hyperplasia forms as a result of ocular irritations, such as inflammation or trauma which can be detected by history, and additional symptoms.<sup>5</sup> All these lesions could be ruled out by history, clinical appearance, angiography, ultrasonography, and OCT, especially by an experienced observer.

As the lesion is usually small, it can not always be detected by ultrasonography. OCT is an effective, practical method to demonstrate these small lesions as elevated masses with solid appearance due to reflection of light by the lesion.<sup>3</sup> When compared with OCT, fluorescein angiography is a relatively invasive technique for diagnosis. As described by Huot et al. in a combined hamartoma of the retina and RPE, SD OCT allows detailed observation of the vitreoretinal interface as well as the disorganisations of retina and RPE.<sup>6</sup>

Simple RPE hamartomas are easily diagnosed solely with fundoscopic examination, and they do not usually need treatment. However, instruments such as fluorescein angiography and OCT are helpful in documentation of these lesions. Although we hold with the OCT findings of simple RPE hamartoma described by other authors, we aimed to demonstrate the high-resolution SD-OCT images which seems to be promising for follow-up of both combined and simple hamartomas as well as other intraocular tumors

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