The Detection of an Occult Choroidal Macrovessel With Enhanced Depth and En-Face Imagings in Optical Coherence Tomography

Optik Koherens Tomografide Artırılmış Derinlik ve Enface Görüntülemeler ile Gizli Koroidal Makrodamarın Saptanması

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ABSTRACT

We aimed to report a rare case with the occult choroidal macrovessel (CM) detected by enhanced depth imaging (EDI) mode of spectral domain optical coherence tomography (OCT). The case is 12 years old girl regularly ongoing follow-ups in the retina section of our clinic for atypical retinitis pigmentosa. Her visual acuities were 1/10 in the right eye and 8/10 in the left eye. Biomicroscopical examination revealed aphakia, diffuse corneal opacity, pupillary irregularity and mild phitisis bulbi in the right eye. Any pathology in the biomicroscopy of the left eye was not observed. Fundus examination revealed widespread retina pigment epithelium (RPE) alterations in posterior pole in both eyes. Fundus fluorescein angiography of the left eye showed widespread RPE window defects around optic disc and in the whole macula at all phases without any retinal vascular filling defect. EDI-OCT and enface-OCT revealed a worm-like or tubular choroidal hyporeflective lesion beneath the fovea. The occult CM was diagnosed based on OCT findings. EDI-OCT and en face OCT might reveal an occult CM in the patients which fundus angiography and examination could not show it.

Key Words: Occult, choroidal macrovessel, enhanced depth imaging, enface imaging, optical coherence tomography.

ÖZ

Spektral domain optic koherens tomografi (OKT)'nin artırılmış derinlikte görüntüleme ve en-face görüntüleme modu ile saptanmış gizli koroidal makrodamarlı nadir bir olgunun rapor edilmesi-amaçlandı. Olgu atipik retinitis pigmentoza nedeniyle kliniğimizin retina biriminde düzenli takipleri devam eden 12 yaşındaki kız çocuğudur. Görme keskinlikleri sağda 1/10 ve solda 8/10 idi. Biyomikroskobik muayenesi sağ gözde afaki, diffüz korneal opasite, pupiller düzensizlik ve hafif fitizis bulbiyi gösterdi. Sol gözün biyomikroskopisinde herhangi bir patoloji izlenmedi. Fundus muayenesi heriki gözde arka kutupta yaygın retina pigment epitel (RPE) değişikliklerini gösterdi. Fundus floresan anjiyografi sol gözde optic disk etrafında ve tüm makulada herhangi bir retinal damarsal dolum defekti olmaksızın yaygın RPE pencere defektlerini açığa çıkardı. Artırılmış derinlikte görüntülemeli ve enface OKT foveanın altında solucanvari/tünel benzeri koroidal hiporeflektif lezyonu gösterdi. OKT bulgularına dayanarak gizli koroidal makrodamar tanısı konuldu. Artırılmış derinlik ve enface OKT fundus anjiyografisi ve muayenesinin göstermediği hastalarda gizli bir koroidal makrodamarı açığa çıkarabilir.

Anahtar Sözcükler: Gizli, koroidal makrodamar, artırılmış derinlik görüntüleme, enface görüntüleme, optic koherens tomografi.

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INTRODUCTION

Choroidal macrovessel (CM was firstly described by Lima et al using indocyanine green angiography (ICGA).[1] Recently, Enhanced depth imaging (EDI) and en face (EF)-optical coherence tomography (OCT) of the CM have been also reported.[2, 3] However, CMs in both reports were clearly observed in the examination, imaging, and angiography of the fundus. EDI-OCT is an imaging mode of OCT providing high-resolution cross-sectional and detailed imaging of the choroid.[4, 5] EF-OCT is a visualization mode based on software reconstruction of OCT images. EF-OCT has capable of achieving depth resolution of 3 mm. It provides a detailed and in-depth retinochoroidal visualization because it gains a series of transverse scans through the entire retina and choroid unlike widely spaced cross-sectional scans in conventional OCT.[6] Here, we report a case with an occult CM that can only be detected by OCT in EDI and EF modes.

CASE REPORT

A 12 years old girl has been regularly ongoing follow-ups in retina section of our clinic for atypical retinitis pigmentosa. In the first presentation, she had no complaint of metamorphopsia or visual loss. However, she has sensorineural deafness. Clinically she is a patient with USHER syndrome. Her best-corrected visual acuities were 1/10 in the right eye and 8/10 in the left eye. She had a medical story including right cataract surgery without intraocular lens implantation. The right eye had 20° exotropia. Biomicroscopical examination revealed aphakia, diffuse corneal opacity, pupillary irregularity and mild phitisis bulbi in the right eye. The left eye had no any pathology in the biomicroscopy. Fundus examination revealed widespread retina pigment epithelium (RPE) alterations posterior pole in the right and left eyes. Fundus fluorescein angiography (FFA) (Zeiss FF450 plus IR;. Carl Zeiss Meditec, Jena, Germany) could not provide appropriate clear image because of corneal opacity in the right eye of the patient. The FFA of the left eye showed widespread RPE window defects around optic disc and the whole macula without any change at early and late phases without any retinal vascular filling defect (Figure 1 and 2). However, any vessel-like lesion or image either retina or choroidea was not observed in both color fundus imaging and FFA. The ICGA could not perform because of the lack of dye. EDI-OCT images, which were obtained from fovea at the horizontal plane, revealed a hyporeflective worm-like or tubular image shadowing on outer choroidea and the sclera (Figure 3). Choroidal thickness at the central fovea and lesion region were 317 and 303 micrometer, respectively (Figure 4). It was detected that choroidal thickness in the lesion region was thicker than that in the temporal region despite to the literature.[7]

Additionally, an EF-OCT image from the level of the choroidal shadowing demonstrated a worm-like macrovessel (Figure 5). Occult CM was diagnosed based on the findings in EDI-OCT and EF-OCT (Zeiss Cirrus HD-OCT 5000, Carl Zeiss Meditec, Jena, Germany).

CONCLUSION

Choroidal macrovessel is an abnormal choroidal vessel firstly described and demonstrated in ICGA by Lima et al. Currently, it is not clear whether CM is congenital or acquired and, whether it has potential systemic associations. In ICGA, it has hyperfluorescent in the early phase and hypo fluorescent in the late phase, and it does not leak the dye.[1-3] Although ICGA is the best method to image the choroidal vessels, we considered that it can not provide correct information about the choroidal circulation because of pigmentary alterations in the RPE in this case. Additionally, we could not perform ICGA because of the commercial lack of dye.

Choroidal macrovessel is located at inner choroid and it has

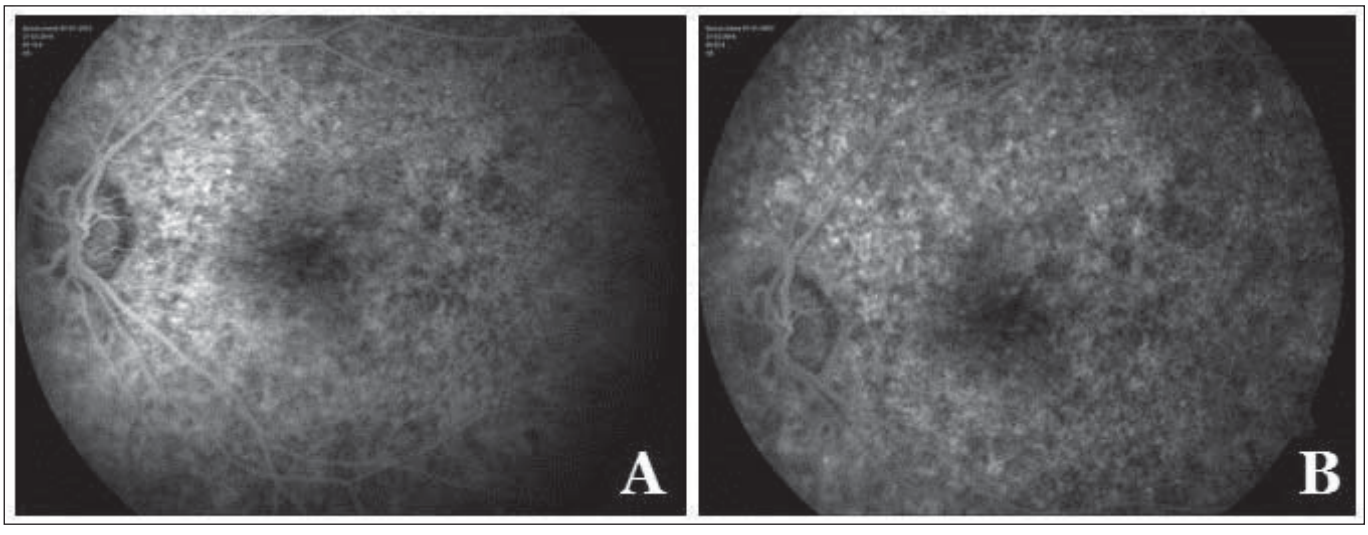

Figure 1. *The images from early (A) and late (B) phases in fundus fluorescein angiography of the left eye. They show the widespread RPE window defects in the whole macula.*

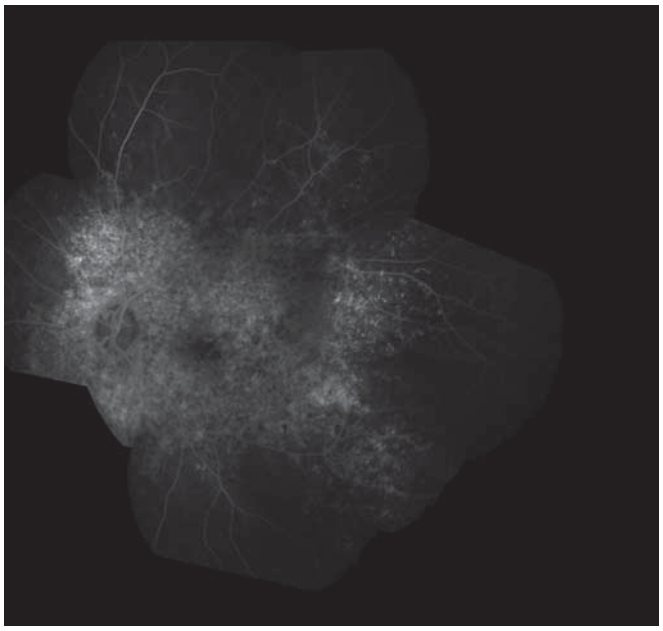

Figure 2. *The image of composite fundus fluorescein angiogram. There is no any finding suspecting choroidal macrovessel.*

a tortuous configuration. It may be associated no retinal abnormality or hyperpigmentation of the RPE, debris in the subretinal space, and changes in the thickness of outer nuclear layer.[1-3] In our case, CM was not associated with any retinal pathology based on the OCT findings.

In healthy children, the choroidal thickness is the thickest at the subfoveal area, and temporal choroid is thicker than nasal choroid[7] We observed that nasal choroidal thickness, in the other words, in the region where CM is observed in EDI-OCT, was thicker than that in the temporal region.

The differential diagnosis of a CM should be performed from the choroidal vascular lesions,¬choroidal hemangioma, subretinal parasite, vortex varix, retinochoroidal anastomosis, anomalous posterior ciliary vessel,¬ocular trauma and inflammatory choroidal disorder.[1-3, 8] Our case had no story and findings attributed to these diseases or conditions. Valsalva maneuver did not reveal choroidal varix. Systemic examination did not reveal any disorder associated with a choroidal hemangioma. We could not also detect whether this shadowing tortuous structure belonged to a retinocho-

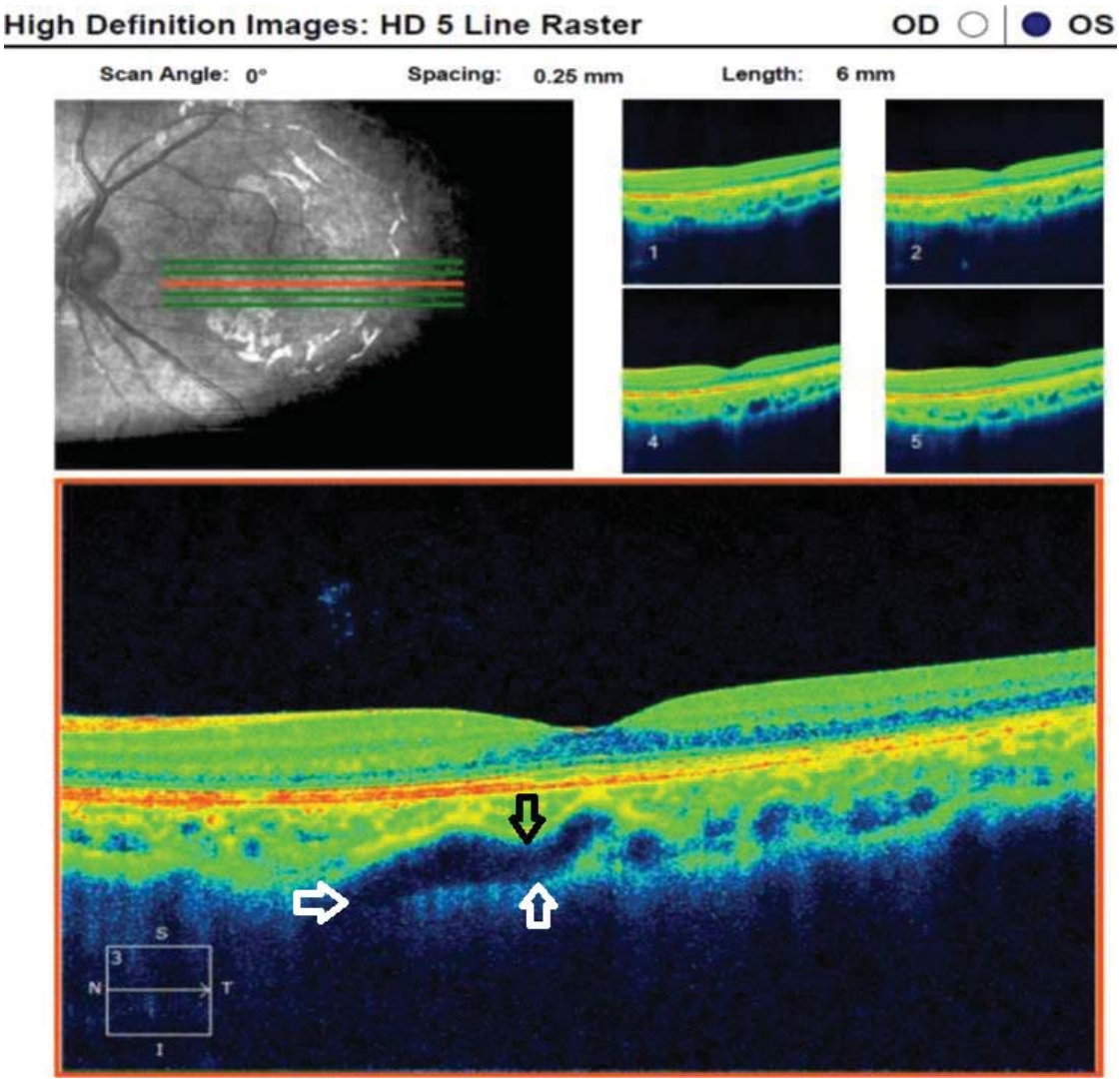

Figure 3. *OCT scans crossing the occult choroidal macrovessel. Arrows point out the occult choroidal macrovessel.*

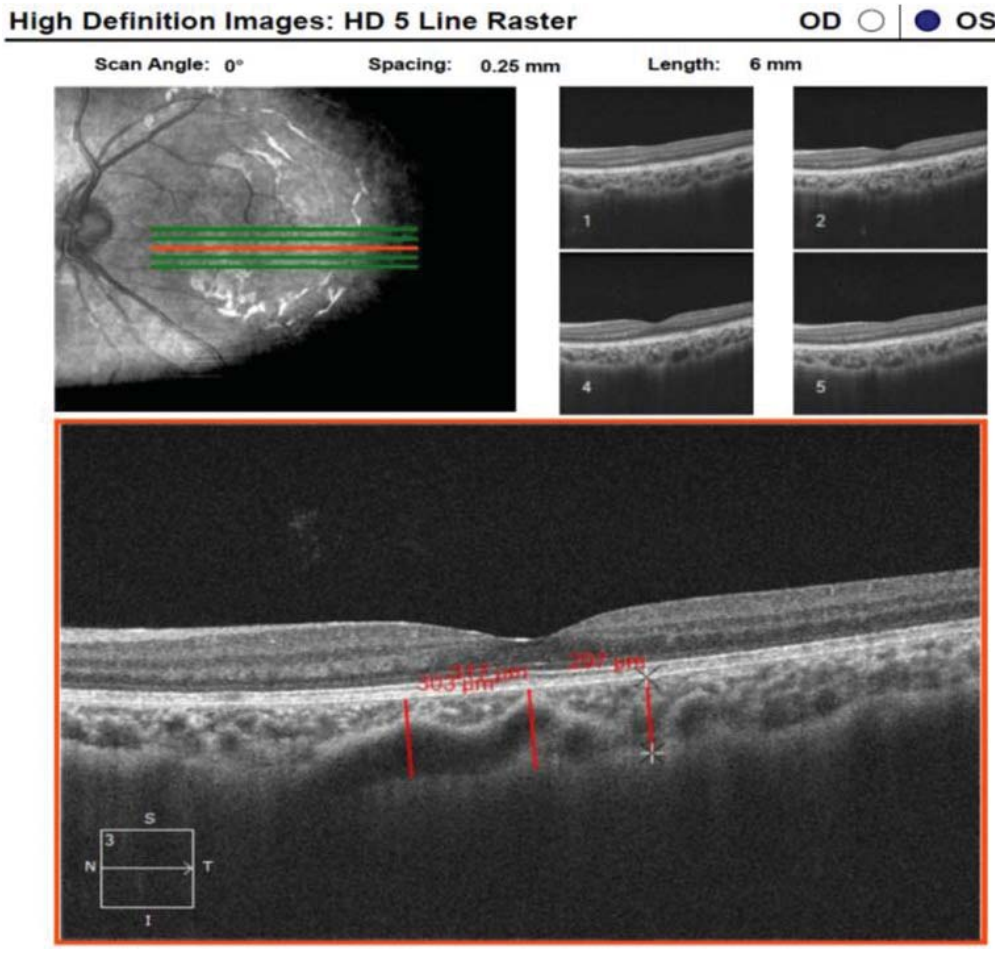

Figure 4. *The choroidal thickness measurements in subfoveal and at lesion region in the left eye using enhanced depth imaging*

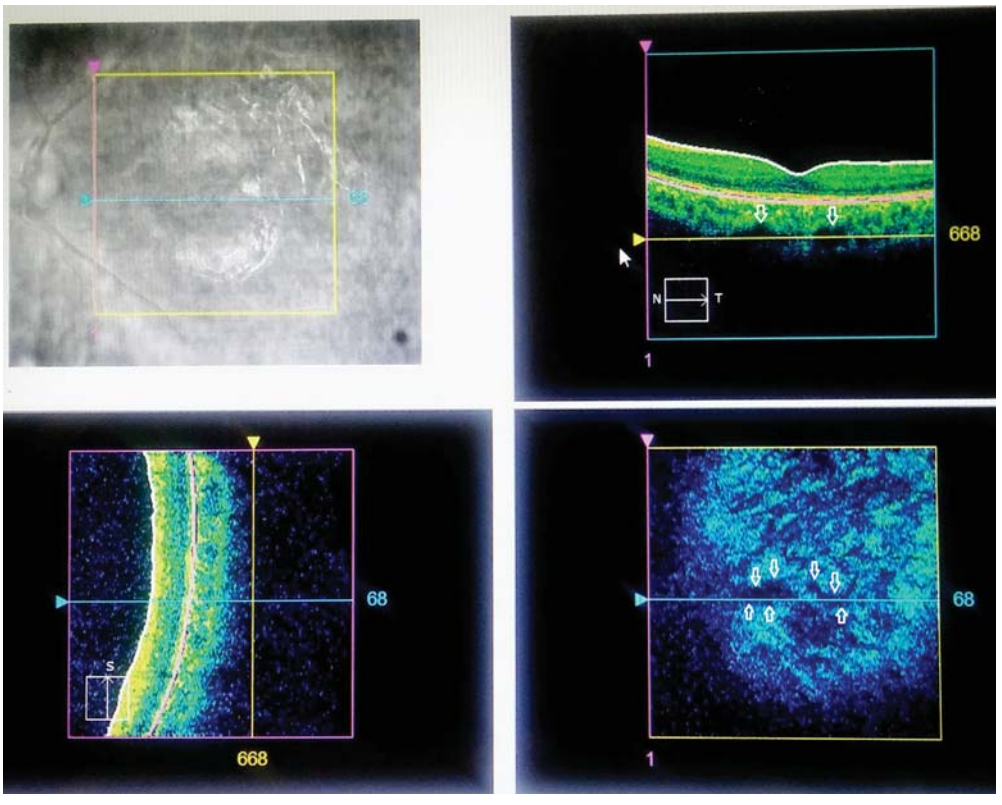

Figure 5. *En face mode imaging of choroidal macrovessel. Arrows point out the worm-like choroidal macrovessel.*

roidal anastomosis, arterial branch, aberrant posterior ciliary artery, aberrant vortex vein or vortex varix.

In this report, we presented a rare clinical entity of an occult CM using EDI-OCT and EF-OCT. In our case, CM is located at outer choroid (Haller layer) and it has a tortuous and tubular configuration. In the previous reports, it was considered that a similar configuration in ophthalmoscopy and angiography might belong to the‒CM because of the tortuous or worm-like appearance at the macula. However, ophthalmoscopy and FFA could not show it because our case has diffuse RPE window defects. We choice to name this entity as ‘‘occult choroidal macrovessel’’ because it was incidentally detected by OCT in our case during the routine follow-up examination, and fundus angiography and examination could not show it. It might suggest that the term ‘‘Occult Choroidal Macrovessel’’ might be used in the cases in which clear choroidal imaging could not be gained. Large case series to be used EDI-OCT, EF-OCT or multimodal imaging might reveal the potential importance and systemic associations of this rare clinical entity.

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