EDI-OCT of Intraocular Tumors

Brand new findings, hot off the presses.

BY CAROL L. SHIELDS, MD

nhanced-depth imaging spectral-domain optical coherence tomography (EDI-OCT) is a recently introduced modality for imaging the choroid and other far-posterior structures in the posterior pole of the eye. Since the description of this technique by Spaide and colleagues¹ a short 5 years ago, hundreds of papers exploring its implications and capabilities have been published, according to a recent review.2

Our group at the Ocular Oncology Service of Wills Eye Hospital has contributed and will continue to contribute to this burgeoning literature with a number of studies describing the characteristics of intraocular tumors as they appear on EDI-OCT. This article summarizes some of the EDI-OCT findings we have published to date and gives a foretaste of several articles that are currently in press or in manuscript form.

CHOROIDAL NEVUS

Shah and colleagues³ described the characteristics of choroidal nevus in 104 eyes imaged with EDI-OCT. Imaging was performed with the Heidelberg Spectralis HRA+OCT (Heidelberg Engineering) with a custom scan acquisition protocol.

We found that EDI-OCT imaging yielded the best results in patients who were younger than 60 years of age and who could cooperate, and for imaging tumors situated posterior to the equator. The contour of the retina overlying the nevus is that of a gentle mound. On EDI-OCT, choroidal nevus appears to originate in the outer choroid and to compress the choriocapillaris inward, causing choroidal shadowing. This choroidal shadowing and choriocapillaris compression was seen in 94% of eyes in the study. Detachment of the overlying retinal pigment epithelium (RPE) was seen in 8% of eyes, loss of photoreceptors in 43% of eyes, and irregu-

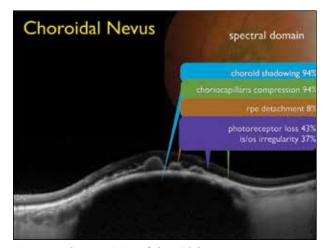


Figure 1. Characteristics of choroidal nevus as seen on EDI-OCT including choroidal shadowing, choriocapillaris compression, RPE detachment, photoreceptor loss, and IS/OS irregularity.

larity of the photoreceptor inner segment/outer segment (IS/OS) junction in 37% (Figure 1).

When subretinal fluid overlying the choroidal nevus is fresh, the photoreceptors can appear shaggy, but when the fluid is more chronic the photoreceptors can have a stalactite appearance. When the fluid has become very chronic there is loss of photoreceptors, and the photoreceptors appear retracted (Figure 2).

We are currently investigating the use of anti-VEGF therapy and photodynamic therapy (PDT) for treatment of the foveal edema and subretinal fluid associated with choroidal nevus.

The buzzwords to remember for choroidal nevus are the gentle mound contour, often with pigment epithelial detachment (PED) and with retracted photoreceptors.

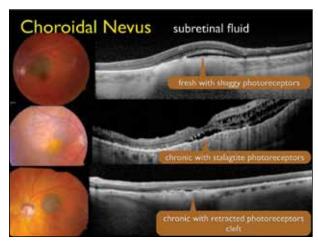


Figure 2. Depending on the chronicity of subretinal fluid overlying choroidal nevus, photoreceptors can appear shaggy, with a stalactite appearance, or retracted.

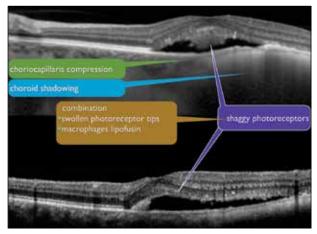


Figure 3. Characteristics of choroidal melanoma as seen on EDI-OCT.

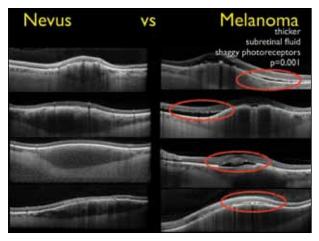


Figure 4. Side-by-side comparison of the features of choroidal nevus and choroidal melanoma on EDI-OCT.

CHOROIDAL MELANOMA

Small choroidal melanoma often shows some of the same features described above for choroidal nevus, including choroidal shadowing, choriocapillaris compression, and shaggy photoreceptors (Figure 3). The shaggy appearance of the photoreceptors, we think, represents swollen photoreceptor tips or lipofuscin-laden macrophages clinging to the underside of the photoreceptors.

Shields and colleagues⁴ examined 37 eyes with small choroidal melanoma (<3 mm thickness) using EDI-OCT and compared the characteristics of choroidal melanoma with those of choroidal nevus. Melanoma tended to be thicker, often with the presence of subretinal fluid and subretinal lipofuscin deposition, and with retinal irregularities including shaggy photoreceptors (Figure 4). Shaggy photoreceptors were overlying small choroidal melanoma in 49% of eyes but were not seen overlying choroidal nevus (P < .001); therefore, the presence of shaggy photoreceptors helps to differentiate melanoma from nevus.

The buzzwords for choroidal melanoma are a domeshaped gentle contour and shaggy photoreceptors.

CHOROIDAL METASTASIS

In 2005, Arevalo and colleagues⁵ described the characteristics of choroidal metastasis using time-domain (TD) OCT. They observed anterior displacement of the photoreceptor layer by subretinal fluid overlying a hyperreflective, thickened RPE-choriocapillaris complex. However, not much information on the metastasis itself could be gleaned with TD-OCT because of the lesion's choroidal location.

A description of the characteristics of choroidal metastasis on EDI-OCT is being prepared for publication by our group. The features observed in 31 eyes with choroidal metastasis include a lumpy-bumpy contour, choriocapillaris compression, photoreceptor loss, subretinal fluid, and subretinal debris such as macrophages with lipofuscin (Figure 5).

The buzzword for choroidal metastasis is lumpybumpy contour.

HEMANGIOMA

Hemangioma can be hard to visualize in the fundus, and it is often misdiagnosed as central serous chorioretinopathy (CSC). Ultrasound also may not provide much information. On EDI-OCT, the mass in the choroid can be clearly seen. Unlike other choroidal tumors, with hemangioma there is no compression of the choriocapillaris; the choriocapillaris may in fact be expanded. Other features include a smooth, not lumpy, contour; optical shadowing with indistinct tumor margins; and often a little wing of tumor in the periphery.

The buzzwords for hemangioma are a domed contour and no compression of the choriocapillaris.

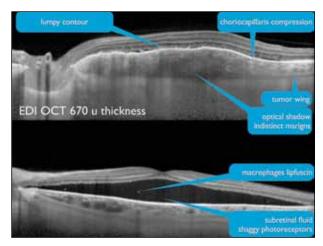


Figure 5. Characteristics of choroidal metastasis as seen on EDI-OCT.

LYMPHOMA

Choroidal lymphoma may be the most interesting tumor to image with EDI-OCT. Depending on its thickness, it can have different appearances that might be described in oceanic terms: Thin lesions, in the realm of 1.7 mm, look placid; thicker tumors of about 2.8 mm appear rippled; when the tumors become even thicker, about 4.1 mm in thickness, they have a "seasick" or wavy "rough seas" appearance (Figure 6).

Our group⁶ first described the seasick appearance of choroidal lymphoma in a case report. We are preparing a manuscript for publication describing the placid, rippled, and seasick topography as observed with EDI-OCT in 14 eyes with lymphoma.

The buzzwords for choroidal lymphoma are its undulating contour; placid, rippled, or seasick topography; and the presence of subretinal and intraretinal fluid.

TUMORS OF THE RPE

Fung, Pellegrini, and Shields have a paper in press in Ophthalmology describing EDI-OCT findings in 18 cases of congenital hypertrophy of the RPE (CHRPE). These lesions are characterized in all cases by flat contour, photoreceptor loss, and normal choroid underlying the CHRPE. No subretinal fluid is seen, and a subretinal cleft is observed in a third of cases.

There is a big difference in images of CHRPE obtained with TD-OCT compared with spectral-domain EDI-OCT. On TD-OCT, few details are visible, while spectraldomain EDI-OCT clearly shows the lesion's characteristic lacunae, photoreceptor loss, and occasional subretinal cleft (Figure 7).

The buzzword for CHRPE is flat appearance with abrupt photoreceptor loss.

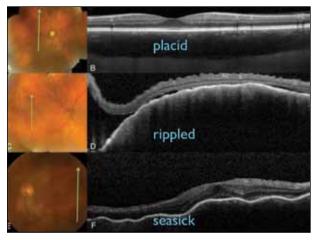


Figure 6. Thin choroidal lymphoma can appear placid, with medium thickness can have a rippled appearance, and when thicker can have a seasick appearance.

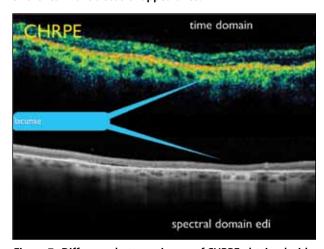


Figure 7. Difference between image of CHRPE obtained with TD-OCT compared with spectral-domain EDI-OCT.

RETINOBLASTOMA

Retinoblastoma is saved for last here because all the studies described above were performed using the Heidelberg HRA+OCT, but for these pediatric tumors we have been using the Optovue iVue portable handheld EDI-OCT. This instrument can be used in the operating room, where we can image the retinoblastoma in children under anesthesia.

The fovea is normal, but temporally the exophytic tumor can be seen (Figure 8), with normal retina lying over the tumor.

The buzzword for retinoblastoma is exophytic mass abruptly arising within the retina.

CONCLUSION

To review the buzzwords for EDI-OCT imaging of intraocular tumors:

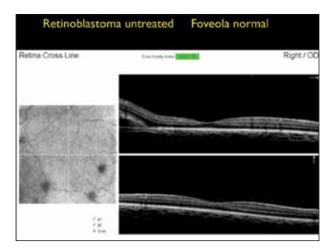


Figure 8. Patient with retinoblastoma. The fovea is normal, but temporally the exophytic tumor can be seen, with normal retina lying over the tumor.

- · Choroidal nevus: Gentle contour;
- · Small choroidal melanoma: Shaggy photoreceptors;
- Choroidal metastasis: Lumpy-bumpy contour;
- Choroidal lymphoma: Placid, rippled, or seasick contour;

- · CHRPE: Flat contour;
- · Retinoblastoma: Exophytic mass.

No doubt we are still in the early days, months, and years of learning what EDI-OCT and other imaging modalities can teach us about tumors of the choroid and other intraocular structures. We look forward to continuing to contribute to the growing literature on this subject.

Carol L. Shields, MD, is the Co-Director of the Ocular Oncology Service, Wills Eye Institute, Thomas Jefferson University. She is a Retina Today Editorial Board member. Dr. Shields states that she has no financial interest in the technologies discussed in this article. She may be reached at +1 215 928 3105; or at carolshields@gmail.com.

- 1. Spaide RF, Koizumi H, Pozzoni MC. Enhanced depth imaging spectral-domain optical coherence tomography. *Am J Ophthalmol.* 2008;146:496-500.
- 2. Mrejen S, Spaide RF. Optical coherence tomography: Imaging of the choroid and beyond. *Surv Ophthalmol.* 2013;58(5):387-429.
- 3. Shah SU, Kaliki S, Shields CL, Ferenczy SR, Harmon SA, Shields JA. Enhanced depth imaging optical coherence tomography of choroidal nevus in 104 cases. *Ophthalmology*. 2012;119(5):1066-1072.
- 4. Shields CL, Kaliki S, Rojanapom D, Ferenczy SR, Shields JA. Enhanced depth imaging optical coherence tomography of small choroidal melanoma: comparison with choroidal nevus. *Arch Ophthalmol.* 2012;130(7):850-856.

 5. Arevalo JF, Fernandez CF, Garcia RA. Optical coherence tomography characteristics of choroidal metastasis. *Ophthalmology.* 2005;112(9):1612-1619.
- Árias JD, Kumar N, Fulco EAM, et al. The seasick choroid: a finding on enhanced depth imaging spectral-domain optical coherence tomography of choroidal lymphoma. Retinal Cases & Brief Reports. 2013:7(1): 19–22