

Shaggy Photoreceptors With Small Choroidal Melanoma

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Differentiation between small choroidal melanoma (<3-mm thickness) and benign choroidal nevus is challenging due to several clinical similarities. Both lesions can appear pigmented or nonpigmented and show similar basal and thickness dimensions. The importance of differentiation lies in the risk for systemic metastasis of approximately 12% at 10 years for small melanoma¹ vs no potential for metastasis from nevus. Early detection of small choroidal melanoma is critical, as each millimeter increase in thickness potentially adds 5% increased risk for metastasis.^{2,3} In an effort to improve early detection, the mnemonic “TFSOM” was designed to represent the clinical features predictive of small melanoma (vs nevus), where T = thickness greater than 2 mm, F = subretinal fluid, S = symptoms, O = orange pigment, and M = margin touching optic disc.^{1,3,4} Lesions with 3 or more clinical features generally represent small choroidal melanoma and are treated promptly.

There is no single test to differentiate choroidal nevus from small melanoma. The differentiation lies in a spectrum of findings. Enhanced-depth imaging optical coherence tomography (EDI-OCT) is a useful tool for imaging the retina and choroid. This advanced technology provides cross-sectional imaging of the eye to a resolution of 3–4 μm ,⁵ far greater than other imaging modalities. By comparison, ultrasonography provides 50–200 μm resolution.⁶ We have observed the presence of subretinal fluid with elongated (shaggy) photoreceptors on EDI-OCT overlying some small choroidal melanomas, a feature not generally found with choroidal nevus ($P=.0001$).⁷ In fact, herein, we illustrate three cases of small choroidal melanoma with characteristic shaggy photoreceptors on EDI-OCT. In each case, the feature resolved following treatment with plaque radiotherapy.

CASE REPORTS

Case 1. A 58-year-old white man noted blurred vision in the left eye (OS) for 6 months. Corrected visual acuity was 20/30 in the right eye (OD) and 20/50 OS. Slit-lamp examination of both eyes (OU) and fundus OD were unremarkable. The fundus OS manifested a small pigmented subfoveal choroidal mass of 3 mm basal diameter with overlying subretinal fluid and lipofuscin (orange pigment) (Figure 1A). Ultrasonography depicted a thin choroidal mass of 1.9-mm thickness. EDI-OCT clearly demonstrated an elevated choroidal mass with posterior shadowing, overlying subretinal

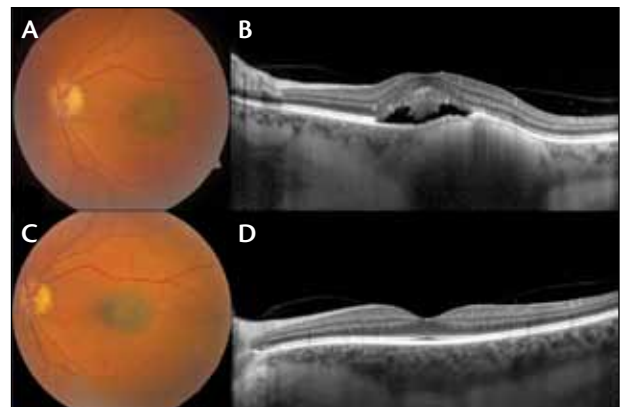


Figure 1. A 58-year-old white man with small choroidal melanoma in macular region with overlying orange pigment and subretinal fluid (A). Enhanced-depth imaging optical coherence tomography (EDI-OCT) showed elevated choroidal mass with overlying subretinal fluid and shaggy photoreceptors at presentation (B). At 4 months following plaque radiotherapy, the tumor regressed (C). At 4 months after treatment, EDI-OCT showed the choroidal mass regressed, subretinal fluid resolved and normalization of photoreceptors (D).

fluid, and shaggy, elongated photoreceptors overlying the tumor (Figure 1B).

The features were consistent with small choroidal melanoma, and plaque radiotherapy was performed. At 4 months follow-up, visual acuity had improved to 20/30 OS. The melanoma was regressed (Figure 1C) to thickness of 1.4 mm. On EDI-OCT, the choroidal mass was flattened, subretinal fluid resolved, and photoreceptors appeared normalized (Figure 1D).

Case 2. A 60-year-old white man presented with a suspicious choroidal lesion OS. Corrected visual acuity was 20/20 in each eye (OU). Slit-lamp examination OU and fundus OD were unremarkable. Fundus OS revealed a pigmented choroidal lesion of 6.5 mm basal diameter in the macular region with overlying subretinal fluid and lipofuscin (Figure 2A). Ultrasonography manifested a thin choroidal mass of 2.0 mm thickness. EDI-OCT showed elevated choroidal mass with overlying subretinal fluid and shaggy elongation of photoreceptors overlying the tumor (Figure 2B).

The features were consistent with small choroidal melanoma, and the patient underwent plaque radiotherapy. At 5 months follow-up, visual acuity had decreased to 20/40 OS. Fundus examination revealed reduction of orange pigment (Figure 2C). The melanoma regressed to a thickness of 1.8 mm. EDI-OCT revealed flattened mass, resolution of subretinal fluid, and normalization of photoreceptors (Figure 2D).

Case 3. A 71-year-old white woman was referred for evaluation of a pigmented lesion OS. Visual acuity was 20/40 OU. Slit-lamp examination revealed multiple iris

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freckles OU. Fundus examination showed extramacular fine hard drusen OU and a small choroidal melanoma measuring 5 mm in diameter and 3.8 mm in thickness with overlying subretinal fluid and orange pigment OS (Figure 3A). EDI-OCT showed minimal subretinal fluid and shaggy elongation of photoreceptors overlying the tumor (Figure 3B).

The clinical features were consistent with small choroidal melanoma, and the patient underwent plaque radiotherapy. At 8 months follow-up, visual acuity had decreased to 20/150 OS. The melanoma regressed to a thickness of 2.2 mm (Figure 3C). EDI-OCT revealed resolution of subretinal fluid and normalization of photoreceptors (Figure 3D).

DISCUSSION

Early detection of choroidal melanoma is important. In an analysis of 7256 eyes with choroidal melanoma, the mean tumor base was 11.3 mm and mean tumor thickness was 5.5 mm. Overall, 25% of patients manifested

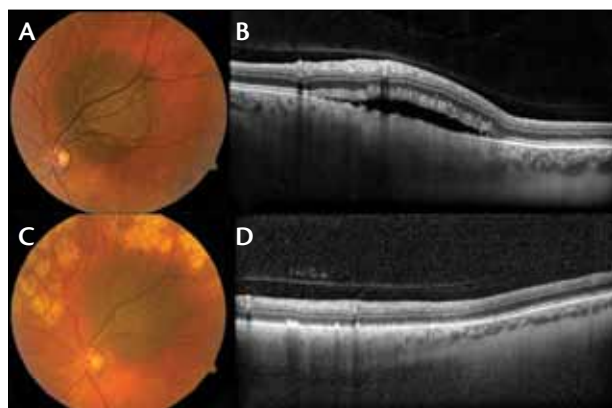


Figure 2. A 60-year-old white man with small choroidal melanoma in the macular region with overlying subretinal fluid and lipofuscin (A). EDI-OCT showed a minimally elevated choroidal mass with overlying subretinal fluid with shaggy photoreceptors (B). At 5 months after treatment, the melanoma was regressed (C), subretinal fluid had resolved, and photoreceptors were normalized (D).



Figure 3. A 71-year-old white woman with small choroidal melanoma with overlying subretinal fluid and orange pigmentation over the apex of the lesion (A). EDI-OCT showed shallow subretinal fluid and the presence of shaggy photoreceptors overlying the tumor (B). At 9 months after treatment, the melanoma was regressed (C), subretinal fluid resolved and the photoreceptors were normalized (D), but the retina was thinned.

Shaggy, elongated photoreceptors are not unique to small choroidal melanoma and have been found with other diseases.

metastasis by 10 years, and the prognosis varied based upon tumor size. For small melanoma (< 3-mm thickness), the 10-year metastatic rate was 5% (0.0-1.0-mm thickness), 12% (1.1-2.0 mm), and 12% (2.1-3.0 mm). By contrast, the 10-year metastatic rate for patients with medium-size melanoma (3-8-mm thickness) was 25%, and for large melanoma (>8 mm) was 49%.² Tumor size at the time of choroidal melanoma detection is an important factor in prognosis.

In general, the diagnosis of small choroidal melanoma is suspected based on classic clinical features and previously mentioned risk factors.^{1,3,4} Previous versions of OCT have been useful in depicting overlying subretinal fluid, but with little detail of precise neuronal changes. EDI-OCT is more revealing of intraretinal and subretinal features as well as choroidal detail.⁹ EDI-OCT can also more accurately identify, image, and measure lesions too small (<1.0 mm in thickness) to be identified by ultrasonography.¹⁰ A recent report⁷ analyzing the EDI-OCT features of small choroidal melanoma compared with choroidal nevus disclosed several significant differentiating features. Small melanoma was statistically more likely to show on EDI-OCT features of increased tumor thickness, subretinal fluid, lipofuscin deposits, retinal pigment epithelial irregularities, and the striking feature of shaggy photoreceptors. Shaggy photoreceptors were described as irregular, elongated, and presumed swollen photoreceptors due to fresh subretinal fluid. They were found overlying 49% of small choroidal melanomas and in no case overlying choroidal nevus.^{7,8}

Shaggy, elongated photoreceptors are not unique to small choroidal melanoma and have been found with other diseases. Using spectral domain optical coherence tomography (SD-OCT), the outer segments of photoreceptors have been found to show elongation in central serous chorioretinopathy.¹¹ Elongation of photoreceptors has also been reported on SD-OCT in eyes in the acute stage of sympathetic ophthalmia.¹²

In our 3 cases, shaggy photoreceptors were an important finding on EDI-OCT, suggesting that the subretinal fluid was fresh. Following treatment, the subretinal fluid resolved and the retina flattened with normalization in

anatomic appearance and visual function. This implies that shaggy photoreceptors are reversible and not permanently disabled.

In conclusion, the EDI-OCT feature of shaggy photoreceptors overlying a pigmented choroidal mass is another potential feature of small choroidal melanoma. Fortunately, visual recovery concomitant with resolution of the fluid and photoreceptor appearance occurred in all 3 cases reported here following treatment.

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