

OPTIC DISC MELANOCYTOMA: IMAGING WITH OCTA



A case demonstrates how imaging can be used to verify a suspected diagnosis and monitor related vascular features.

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The presence of a benign melanocytic nevus of the optic disc, termed melanocytoma, was first proposed in 1959 by Zimmerman.¹ Melanocytoma is described as a benign, small, round, pigmented mass, composed of heavily pigmented oval or round cells with abundant cytoplasm and small nuclei.²⁻⁶ Although melanocytoma can be located anywhere within the uveal tract, it is most commonly found within the optic disc.²⁻⁶ Zimmerman recognized that melanocytoma cells appeared on histopathology to be similar to cells found in ocular melanocytosis.²

Optic disc melanocytoma is usually asymptomatic, but it can occasionally be associated with visual field defect, relative afferent pupil defect, visual loss, and pain.^{3,4} Important ancillary tests for this condition include visual field, ultrasonography, occasional fluorescein angiography, optical coherence tomography (OCT), and possibly OCT angiography (OCTA).³ Herein, we report a patient with optic disc melanocytoma imaged with OCTA.

CASE REPORT

A 52-year-old black man reported a history of floaters in his right eye (OD) for several decades. On examination, best corrected visual acuity was 20/30 in each eye (OU), and intraocular pressure was 13 mm Hg OD and 14 mm Hg in his left eye (OS). Anterior

and posterior segment examinations OU were unremarkable, except for the optic disc region OD. There was a small, hyperpigmented mass within the optic disc measuring 3.0 mm by 2.0 mm in diameter (Figure 1A). There was no optic nerve edema, retinal exudation, subretinal fluid, vitreous tumor seeding, or branch or central retinal vein occlusion (BRVO, CRVO).

B-scan ultrasonography demonstrated a dome-shaped lesion 2.2 mm thick and with high internal reflectivity. OCTA revealed intact retinal vasculature including the radial peripapillary capillary (RPC) network overlying the mass (Figure 1B). In comparison with the contralateral normal optic disc (Figure 2B), the

RPC appeared intact with central sparing at the optic cup, even though the melanocytoma occupied the cup region. The major retinal veins were slightly more dilated in the affected eye, and the inferonasal vein and artery were observed to be twining and twisting around each other.⁷

On OCT, the melanocytoma revealed intrinsic vascularity, best noted on OCT B-scan overlay (Figure 1D), with flow on the surface and slightly deeper within the mass. In addition, there was elevation of the optic nerve head with heterogeneous reflectivity, posterior shadowing, and no optic cup, much different from the normal, contralateral eye (Figures 1 and 2).

AT A GLANCE

- ▶ Melanocytoma can be located anywhere within the uveal tract but is most commonly found within the optic disc.
- ▶ Although usually asymptomatic, optic disc melanocytoma can occasionally be associated with visual field defect, relative afferent pupil defect, visual loss, and pain.
- ▶ Optic disc melanocytoma and uveal melanoma can share common features and must be differentiated. Melanocytoma of the optic disc has a low risk for transformation into melanoma.

Based on these findings, optic disc melanocytoma with preservation of the RPC was documented, and observation was advised. This lesion has been stable for more than 2 years of follow-up.

DISCUSSION

Optic disc melanocytoma and uveal melanoma can share common features and therefore must be differentiated.⁸ Uveal melanoma tends to occur more commonly among white

individuals (98%),⁹ whereas optic disc melanocytoma can manifest in both whites (65%) and nonwhites (35%).⁶ In a comprehensive analysis of 115 cases of optic disc melanocytoma, this mass was found more commonly in women (62%) than in men (38%) and more often in whites (65%) than in blacks (29%) or Hispanics (6%). The mean age at presentation was 50 years (median 52, range 1-91 years). Kaplan-Meier survival curves showed related visual acuity loss in 33% of eyes in 20 years, and slight tumor enlargement in 38% of eyes in 20 years. Malignant transformation into melanoma was documented in 2% of eyes.⁶

Previously, due to a misunderstanding of the histopathology and a lack of adequate ancillary testing, there was great difficulty in differentiating optic disc melanocytoma from melanoma. Consequently, enucleation was often performed as a safety measure.⁴ Optic disc melanocytoma has now been well described and found to be certainly benign, with rare cases demonstrating malignant transformation to melanoma.⁴⁻⁶ Management of optic disc melanocytoma involves annual observation with confirmatory tests including fundus photography, ultrasonography, visual field, and OCT. In the case presented here, we found OCTA useful for demonstrating preservation of the RPC plexus.

MELANOCYTOMA OR MELANOMA?

OCTA is useful in these cases to noninvasively document choroidal neovascularization (CNV), a finding associated with melanocytoma in 1% of cases, particularly if there is adjacent exudation, subretinal fluid, or subretinal hemorrhage.⁶ Furthermore, OCTA can be useful to noninvasively aid in the detection of secondary issues such as BRVO and branch retinal artery occlusion.¹¹ In our case, the patient's retinal veins were more dilated in the affected eye, presumably from limited outflow obstruction. Optic disc melanocytoma can lead to findings of BRVO in 1% and CRVO in 2%.

On a similar note, in 2017, Pellegrini et al used OCTA to detect subtle CNV

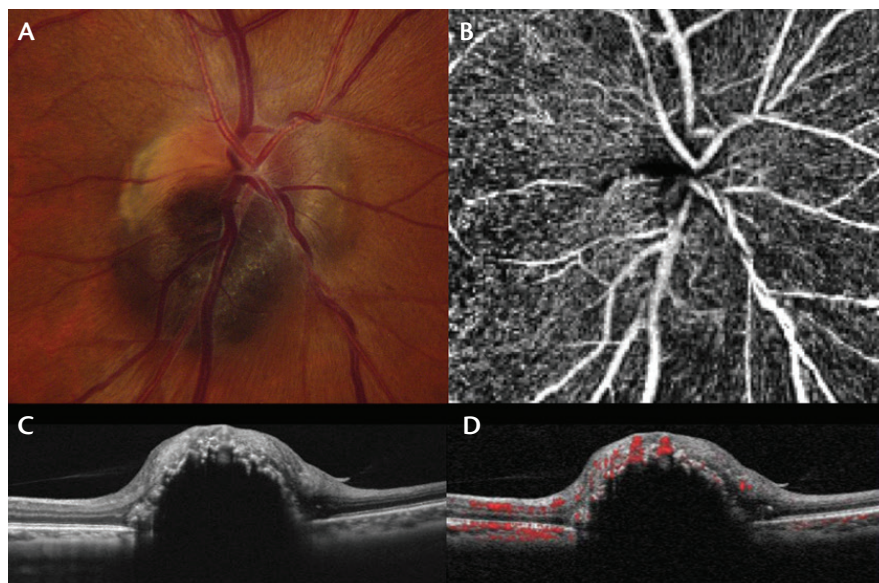


Figure 1. Optic disc melanocytoma measuring 3.0 mm by 2.0 mm in diameter (A). OCTA reveals intact RPC network with sparing at the site of anatomic optic cup (B). OCT B-scan through the lesion reveals tumor elevation with heterogeneous reflectivity and posterior shadowing (C). OCTA B-scan with angiography overlay demonstrates flow over the tumor surface and slightly deeper within the lesion (D).

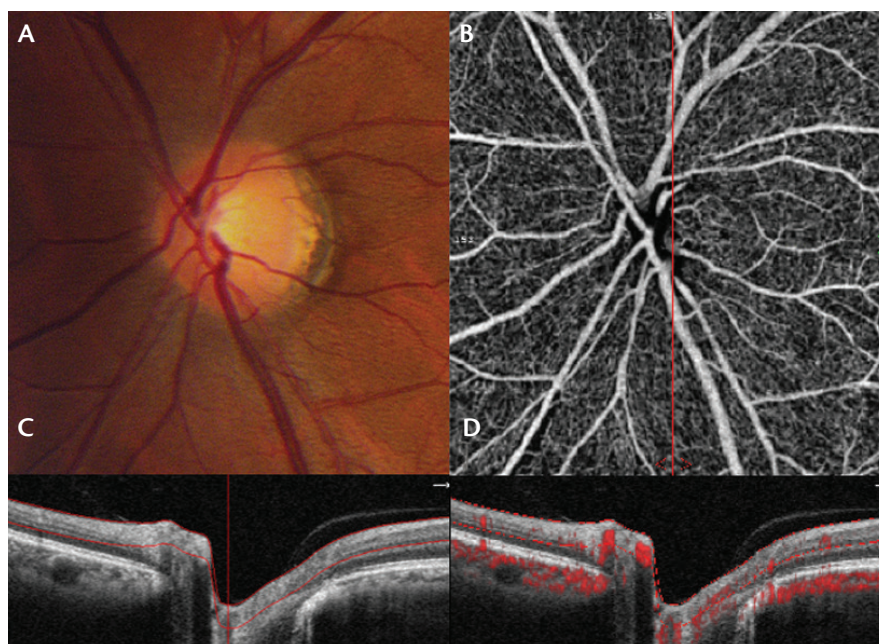


Figure 2. Normal healthy optic disc and optic cup in the contralateral eye (A). OCTA shows intact RPC network with sparing at the optic cup (B). OCT B-scan shows configuration of normal disc with optic cup (C). OCTA B-scan with angiography overlay demonstrates normal flow within retina and choroid (D).

overlying chronic choroidal nevi and document two vascular patterns including a sea fan pattern (73%) and long filamentous linear vessels (27%).¹² Also in 2017, Pointdujour-Lim et al evaluated choroidal nevus with CNV managed with photodynamic therapy and found successful partial or complete resolution of related subretinal fluid in 87% of eyes.¹³

In summary, melanocytoma of the optic disc is a benign tumor with low risk for transformation into melanoma. Related vascular features such as BRVO, CRVO, and CNV can be monitored by clinical examination and imaged with OCTA. ■

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No conflicting relationship exists for any author.

Support provided by the Eye Tumor Research Foundation, Philadelphia, Pa. (CLS). The funders had no role in the design and conduct of the study, in the collection, analysis, and interpretation of the data, or in the preparation, review, or approval of the manuscript. Carol L. Shields, MD, has had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.