

Neovascularized Optic Disc Melanocytoma Revealed by Progressive Visual Disturbance: A Multimodal Imaging Case Report

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Abstract

Optic disc melanocytoma (ODM) is a benign pigmented tumor originating from melanocytes of the optic nerve head. Although usually stable, it may occasionally lead to visual impairment due to complications such as optic neuropathy, ischemia, or secondary neovascularization.

A patient presented with progressive visual disturbance in the right eye over three months. Best-corrected visual acuity was 5/10 OD and 10/10 OS. Fundus examination revealed a pigmented optic disc mass with adjacent exudates. Optical coherence tomography showed optic disc edema with perilesional cystoid spaces and localized outer retinal atrophy. Fluorescein angiography demonstrated nasal peripapillary hyperfluorescence suggestive of neovascular activity. B-scan ultrasonography revealed a 2.5-mm oval hyper-echogenic lesion centered on the optic disc without calcification but with mild Doppler vascularization extending through the lamina cribrosa.

Intravitreal anti-VEGF therapy was initiated with a planned series of three injections. Multimodal imaging is essential for diagnosing optic disc melanocytoma and detecting rare vascular complications. Anti-VEGF therapy may help control neovascular activity and stabilize vision.

Keywords: Optic Disc Melanocytoma, Optic Nerve Head Tumor, Neovascularization, Oct, Fluorescein Angiography, Anti-VEGF.

Introduction

Optic disc melanocytoma is a benign melanocytic tumor arising from melanocytes located in the optic nerve head and, according to Shelds, is generally considered a variant of melanocytic nevus characterized by dense pigmentation and usually slow growth [1]. Most lesions remain stable over time and are discovered incidentally during routine ophthalmologic examination; however, visual field defects, ischemic optic neuropathy, tumor necrosis, retinal edema, vascular occlusions, and choroidal or juxtapapillary neovascularization have all been described by Shiels and Tsai [1,2].

Recent advances in multimodal imaging, including optical coherence tomography (OCT), OCT angiography (OCTA), fluorescein angiography, ultrasonography, and magnetic resonance imaging, have greatly improved the diagnostic evaluation and follow-up of optic nerve head tumors according to Zhou [3]. We report a case of optic disc melanocytoma complicated by suspected neovascularization identified using multimodal imaging and managed with intravitreal anti-VEGF therapy.

Case Presentation

A patient presented with progressive visual disturbance in the right eye over a period of three months. At examination, best-corrected visual acuity was 5/10 in the right eye and 10/10 in the left eye. Intraocular pressure measured 14 mmHg OD and 15 mmHg OS. Slit-lamp examination of the anterior segment was unremarkable in both eyes. Fundus examination revealed a dark pigmented mass involving the optic nerve head associated with adjacent exudates. Spectral-domain OCT demonstrated optic disc edema with hyperreflective areas, perilesional cystoid spaces, and localized outer retinal atrophy in the papillomacular bundle. Small macular microcystic spaces were also observed.

Fluorescein angiography revealed a hyperfluorescent area nasal to the optic disc suggestive of abnormal vascular activity compatible with neovascularization. B-scan ultrasonography revealed an oval hyper-echogenic lesion centered on the optic disc measuring approximately 2.5 mm in its largest axis. The lesion elevated the adjacent wall and did not appear calcified. Color Doppler

imaging demonstrated mild internal vascularization. The lesion appeared to extend through the lamina cribrosa. Based on clinical and imaging findings, the diagnosis of neovascularized optic disc melanocytoma was considered. Intravitreal anti-VEGF therapy was initiated. Three intravitreal injections were planned with clinical and imaging reassessment at each visit. Treatment goals included suppression of neovascular activity, reduction of retinal edema, and stabilization of visual acuity. Long-term follow-up was scheduled to monitor tumor stability and detect recurrence of vascular complications.



Figure 1: Ultra-Widefield Color Fundus Photography (Optos) of the Right Eye Showing a Deeply Pigmented Optic Disc Mass (White Arrow) Associated with Peripapillary Exudation (Yellow Arrow).

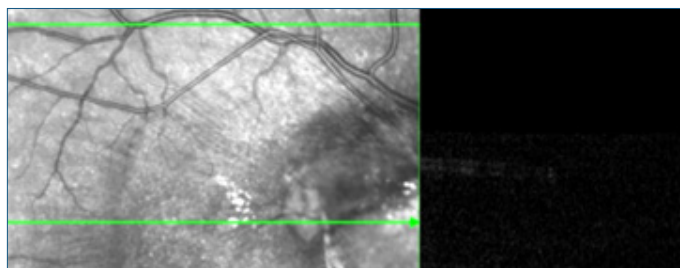


Figure 2: Macular Optical Coherence Tomography (OCT) Demonstrating Perilesional Cystoid Spaces (Yellow Arrow) and Outer Retinal Atrophy (Red Arrow) in the Papillomacular Bundle.

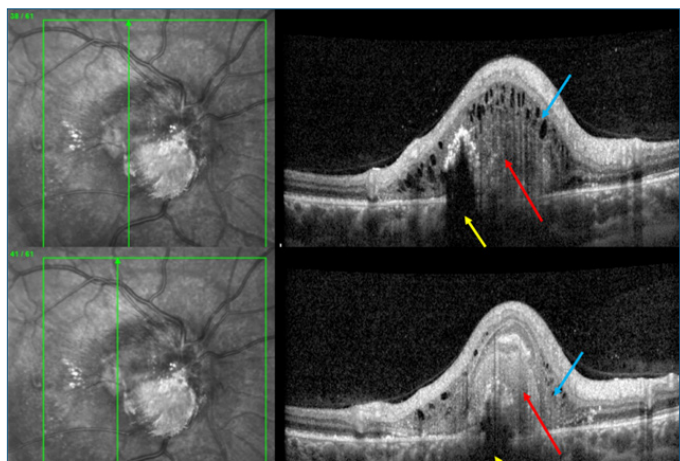


Figure 3: Optical Coherence Tomography (OCT) Scan Through the Optic Nerve Head Lesion Showing an Elevated Hyperreflective

Mass (Red Arrow) with Posterior Shadowing (Yellow Arrow) and Associated Optic Disc Edema (Blue Arrow).



Figure 4: Fluorescein Angiography of the Right Eye Showing a Focal Area of Early Hyperfluorescence Nasal to the Optic Disc (Red Arrow), with Progressive Late Leakage, Consistent with Neovascular Activity Associated with Optic Disc Melanocytoma.

Discussion

Optic disc melanocytoma is generally considered a benign lesion with stable long-term behavior, although slow enlargement and very rare malignant transformation have been reported. Visual dysfunction may result from compression of the optic nerve causing axoplasmic flow stasis, ischemic changes, or tumor necrosis, as detailed by Shields [1]. Vascular complications such as retinal vein occlusion, retinal artery occlusion, disc neovascularization, and choroidal neovascularization have also been described by Tsai, Kamisasanuki and also Al Halafi [2,4,5]. Multimodal imaging plays a crucial role in diagnosis. OCT, described by Zhou, often demonstrates an elevated lesion with posterior shadowing due to dense pigmentation [3], whereas ultrasonography, detailed by Carnevali et al and Zhang et al., typically shows a small elevated lesion with high internal reflectivity and absence of calcification, helping differentiate melanocytoma from other optic nerve tumors [6,7]. Fluorescein angiography and OCTA allow detection of abnormal vascular networks and leakage associated with neovascular complications, reported by Zhou and Kamisasanuki [3,4].

Intravitreal anti-VEGF therapy has shown favorable outcomes in reported cases of melanocytoma-associated neovascularization, by Hamza HS and Okonkwo, with regression of neovascular membranes or disc neovascularization and stabilization or improvement of visual acuity after bevacizumab or aflibercept injections [8,9].

Conclusion

Optic disc melanocytoma is typically benign but may occasionally lead to visual impairment due to secondary vascular complications. This case highlights the importance of multimodal imaging in diagnosing optic nerve head tumors and identifying associated neovascular activity. Early treatment with intravitreal anti-VEGF therapy may help preserve visual function.

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