



Case Report

Imaging signatures in optic nerve head melanocytoma

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ABSTRACT

Optic nerve head melanocytoma is a benign pigmented lesion which is generally asymptomatic, however it can sometimes be related with visual field disturbance. In most cases the tumor stays stable for the duration of the life. Rarely 1-2% of such cases undergo a malignant transformation, hence serial monitoring is necessary. We report an instance of optic nerve head melanocytoma in a 30-year-old male with characteristic imaging signatures.

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1. Introduction

Optic nerve head melanocytoma (ONM) normally shows up as dark brownish color tumors with fluffy edges situated on the optic nerve head. ONM is regularly analyzed clinically depending on its characteristic clinical features, albeit subordinate testing may help in the conclusion and diagnosis. We report an instance of optic nerve head melanocytoma with signature imaging marks in a young male.

2. Case Report

A 30-year-old man visited for routine ophthalmic checkup. On examination, corrected distant visual acuity was 20/20 in both eyes. The intraocular pressure was 15 mm Hg in both eyes. Dilated fundus evaluation in the right eye revealed a hyperpigmented lesion covering the superior half of the right optic nerve, extending past the edge of the optic nerve onto the adjacent retina. [Figure 1a] The rest of fundus did not show any abnormality in right eye. Anterior and posterior segment examinations in left eye were unremarkable. Based on the clinical appearance of

the lesion, a provisional diagnosis of optic nerve head melanocytoma was made.

A color fundus photo (Carl Zeiss, Germany) was obtained for baseline documentation. Baseline visual field (Humphrey Visual Field Analyzer, Carl Zeiss, Germany) was performed which showed an enlarged blind spot in the right eye.

ONH Angiography (Angioplex 5000, Carl Zeiss, Germany), 4.5 × 4.5 mm scan showed surface vascularity within the lesion. [Figure 1b]. OCT B-scan through the lesion revealed tumor elevation with heterogeneous reflectivity and posterior shadowing and the angiography overlay demonstrated flow over the tumor surface and slightly deeper within the lesion. [Figure 1c] Based on these findings, the diagnosis of Optic nerve head melanocytoma was established. The nature of the disease condition was explained in detail to the patient and the need for periodic monitoring was emphasized.

3. Discussion

Optic nerve head melanocytoma is a unilateral dark pigmented lesion found either on or around the optic nerve head. The normal age at finding is forth to fifth decade, and there seems to be a slight female predilection.¹

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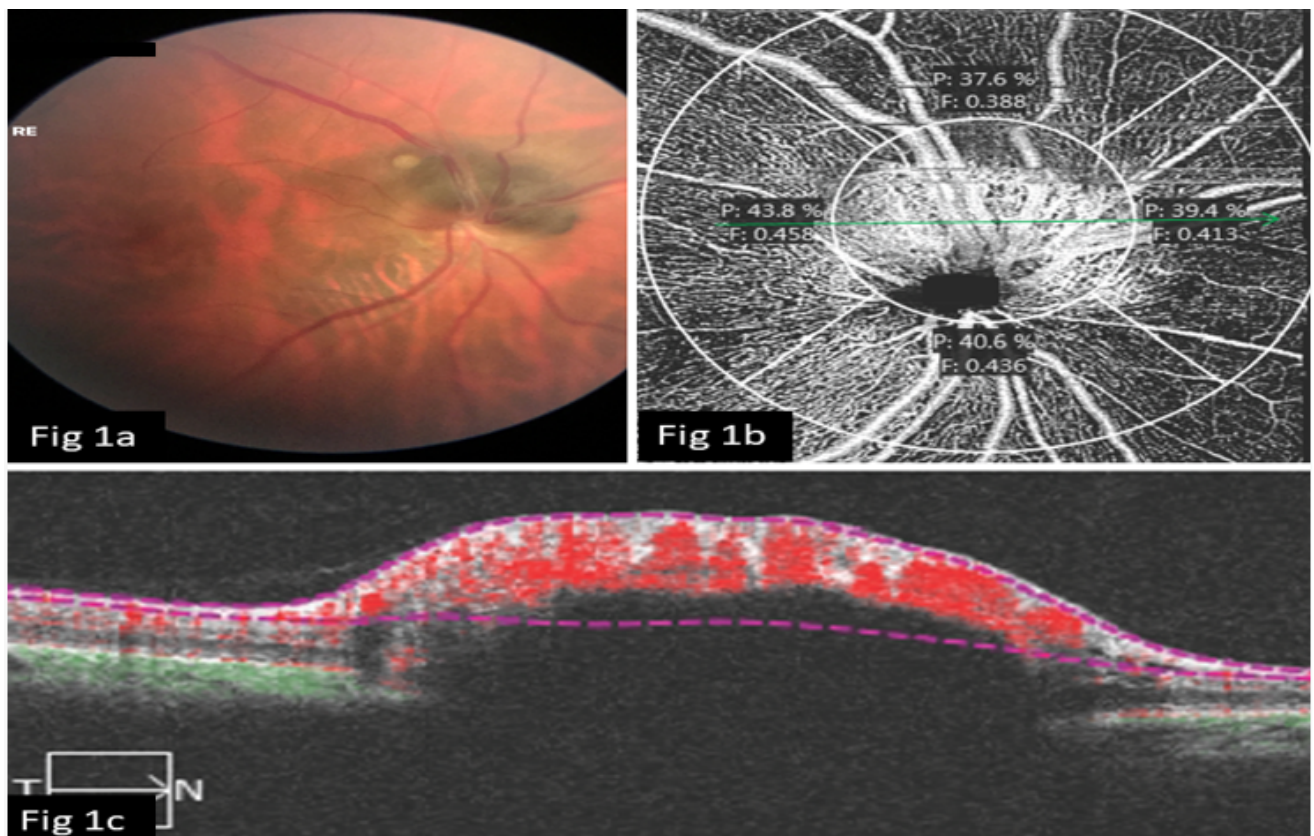


Fig. 1: (a): Color fundus photograph of right eye shows brownish- back elevated lesion with fuzzy margin along the superior aspect of optic nerve; (b): ONH Angiography 4.5 × 4.5 mm scan shows surface vascularity within the lesion in the RPC slab; (c): OCT B-scan through the lesion shows tumor elevation with heterogeneous reflectivity and posterior shadowing and the angiography overlay demonstrates flow over the tumor surface and slightly deeper within the lesion.

While some melanocytomas are bound at the optic nerve head, most cases include the adjoining retina and choroid. As per Joffe et al.,² 18% of patients present with ONH limited to the optic nerve head, 77% extend to the retina, and 47% have juxtapapillary choroid involvement.

By and large, ONM doesn't cause huge vision loss.^{1,3} Up to 26% of optic nerve melanocytomas may cause mild visual impairment, mainly due to ONH edema, retinal edema or subretinal fluid.⁴⁻⁷ Other likely reasons for vision loss include neovascularization, focal vein impediment or malignant transformation.⁴⁻⁷ An afferent pupillary deformity is accounted for in 10-30% of all ONM and has been ascribed to the pressure of optic nerve strands by the melanocytic cells.

More than 80-90% of all ONMs are associated with abnormal visual fields.¹ The commonly found visual field defect is the enlargement of the blind spot, which is accepted to be straightforwardly identified with the measure of tumor expansion and pressure on the optic nerve axons.^{1,3} Nerve fiber bundle defects also include nasal steps and arcuate defects.⁸

Differential diagnoses for ONM comprises of choroidal nevus, juxtapapillary choroidal melanoma and metastatic melanoma to the optic nerve. OCT and OCT-A are excellent apparatuses to differentiate between these two entities. OCT-A helps in visualizing the vascular flow inside the tumor, as well as its extension into the retina and choroid.

Melanocytomas more than 0.5 mm in height might be pictured with B-check. ONM might be checked over the long run with B-scan ultrasonography for malignant transformation.

As per what the established literature says, the tumor stays stable for the duration of the life and just periodic yearly observation is warranted.⁹ A 10-year longitudinal examination has shown that the tumor size could increment in up to 32% of cases and threatening change to melanoma could happen in 1-2% of cases.¹⁰ Rapid tumor growth, extensive involvement of the optic disc, lesion with atypical morphological features and profound vision loss are the risk factors for malignant transformation and such patients should be kept under close observation.

4. Conclusion

All pigmented lesions overlying the optic nerve head should be thoroughly examined and imaged to ensure correct diagnosis and appropriate management.

5. Source of Funding

None.

6. Conflicts of Interest

All contributing authors declare no conflict of interest.

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