

## Case Report

# Melanocytoma of the Optic Nerve- A Case Report

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### Abstract:

Melanocytoma of the optic disc is a benign stationary tumor that is usually pigmented, asymptomatic and stable in size. Even though benign, it can cause vision loss and visual field defect due to compression on the optic nerve.

A 39-year-old male presented with a black shadow in the left eye which has been present for about 4 years. Fundus photo shows a darkly pigmented lesion of the optic nerve head of the left eye with a mild hemorrhage. B-scan ultrasonography shows a slightly elevated lesion at the optic nerve head. Fluorescein angiography shows hypofluorescence of the lesion.

Optic nerve melanocytoma (ONM) may often display clinical similarities to melanoma but with adequate diagnostic tools can be distinguished from one another. ONM can either remain stable or grow progressively slowly over many years. Annual monitoring for change is highly recommended.

**Keywords: Optic Nerve Melanocytoma; Fluorescein Angiography; B scan ultrasonography**

### Introduction

Optic Nerve Melanocytoma (ONM) is a rare benign pigmented tumor and could be progressive and malignant in about 1-2% of cases<sup>1,2</sup>. Intraocular pigmented lesions can occur from melanocytes in the uvea or pigmented epithelium in the retina, ciliary body, and iris<sup>3</sup>. Sometimes it is difficult to distinguish ONM from malignant melanoma, however, most patients are asymptomatic with no vision loss<sup>1,3</sup>. Although ONM has a very low chance of malignancy, it has been previously reported that patients with intraocular melanocytic lesions have been enucleated based on suspicion of melanoma<sup>5</sup>. For those tumors that grow if followed long term, increased tumor thickness, presence of intrinsic vascularization and dome shape have been documented as risk factors for growth<sup>6</sup>.

This study highlights the clinical presentation and diagnosis of ONM using diagnostic tools in a 39 year – old male who came in for a routine eye exam.

### Case Report

A 39-year-old male came with a complaint of a black shadow in the left eye, present for the past four years. He reported no pain, floaters or flashes of light but that onset was gradual. The best corrected visual acuity was 20/20 OD and 20/25 OS. A relative afferent pupillary defect was observed in the left eye. Slit lamp examination showed a normal anterior segment in both eyes. Intraocular pressure was 20mm Hg OD and 18mm Hg OS. On Dilated Fundus Examination, the right eye appeared normal. The left eye showed a clear media with a dark pigmented elevated mass lesion, approximately two-disc diameter in size, displaced more nasally. There was the presence of disc heme located on the inferior disc region

(Figure 2). There were no scattered pigments around the disc, and no retinal edema or detachment was present.

Optical Coherence Tomography (OCT) (Figure 3) and the B scan Ultrasonography showed a dome-shaped lesion, with the B scan lesion showing a thickness of 1.88mm horizontally and 2.53mm vertically (Figure 5). The patient was referred to a retinal vitreous specialist for consultation and fundus fluorescein angiography (FA). FA illustrated hypofluorescence in all phases in areas distinctly covered by the pigmented lesion (Figure 4).

Based on the clinical features and the location, a clinical diagnosis of optic nerve melanocytoma was made. The patient is monitored periodically to keep track of any changes in the lesion.



**Figure 1: Fundus image of the right eye**

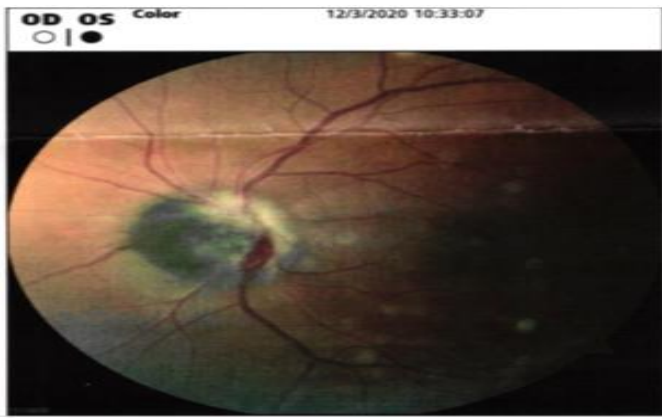


Figure 2: Optic Disc Melanocytoma with hemorrhage

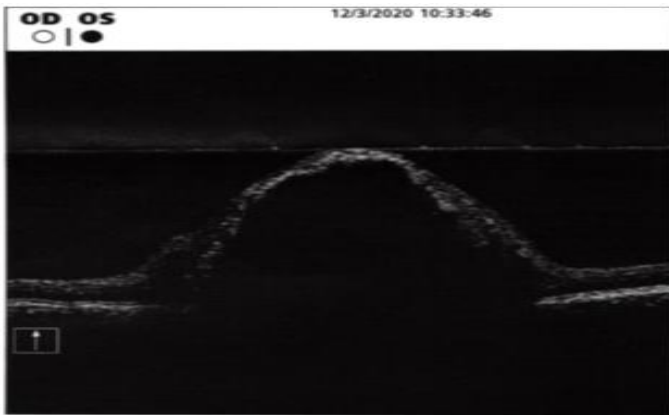


Figure 3: Optical Coherence Tomography Melanocytoma of the Optic Disc Melanocytoma



Figure 4: Fluorescein Angiography of Optic Disc

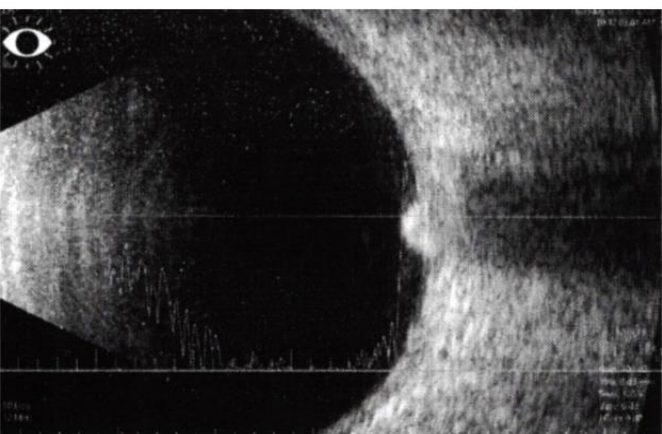


Figure 5 : B scan ultrasonography showing a dome shaped mass lesion

## Discussion

Melanocytoma in many cases presents without symptoms and is found during a routine eye exam. Impairment of visual acuity has been reported in 26% of cases when the fovea is involved<sup>7</sup>. When poor visual acuity occurs, it is usually as a result of optic nerve compression, central retinal vein obstruction, exudative retinal detachment, tumor necrosis, and malignant transformation of the tumor<sup>8</sup>.

Visual acuity defects typically occur as enlargement of the blind spot depending on the extent of extension of the tumor beyond the optic disc margin and then there is the nerve fiber bundle defect due to compression of the optic nerve<sup>7,8</sup>.

A previous review of Optical Coherence Tomography (OCT) for ONH melanocytoma has shown it to present as a dome-shaped lesion, this is consistent with the OCT in this report, as can be seen in Figure 3, occupying the anterior optic nerve and resulting in the distortion of its features<sup>4</sup>.

An important marker of tumor malignancy is vascularity, which is typically seen in intraocular malignancy<sup>9</sup>. ONH melanocytomas are relatively avascular and show hypofluorescence with fluorescein and indocyanine green<sup>8</sup>, consistent with the FA shown in figure 4 of this report. This is very important in distinguishing melanocytoma from malignant melanoma.

An Ultrasound B scan was used as a diagnostic tool in this report. A previous study has shown that the use of a 20 MHz high-resolution ultrasound B scan can be used in identifying and following patients that present with ONH melanocytoma with a high-risk characteristic for growth which can help in avoiding futile treatments<sup>6</sup>.

## Conclusion

Melanocytoma of the optic disc are rare and benign tumor and should be followed periodically with imaging for changes in size, shape and consistency as malignant transformation is possible.

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