

# Melanocytoma of Optic Disc: Clinical Manifestations, Imaging Characteristics and Follow Up

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**Abstract** Melanocytoma of the Optic nerve head, is a benign, stationary pigmented tumor present on or adjacent to the optic disc. Rarely it exhibits minor enlargement and can also undergo malignant transformation to a melanoma. Histopathologically a melanocytoma consists of intensely pigmented round to oval nevus cells with benign features. We report a case of a 77 year old lady with an Optic nerve head melanoma. The clinical features and the imaging characteristics including Ultrasound, Fluorescein angiography and Magnetic resonance imaging are presented.

**Keywords:** melanocytoma, optic nerve head, magnocellular naevus

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## 1. Introduction

Melanocytoma, also known as hyperpigmented magnocellularnevus is a variant of melanocyticnevus that has typical clinical and histopathologic features. The predominant cell type in a melanocytoma consists of plump polyhedral nevus cells that contain numerous giant melanosomes while the other cell type are the smaller spindle shaped cells which are lightly pigmented. [1] In addition to occurring in the optic nerve, melanocytoma can also be found in the uveal tract, including the iris, ciliary body, and choroid. We report a case of Optic nerve head (ONH) melanocytoma in an elderly lady.

## 2. Case Report

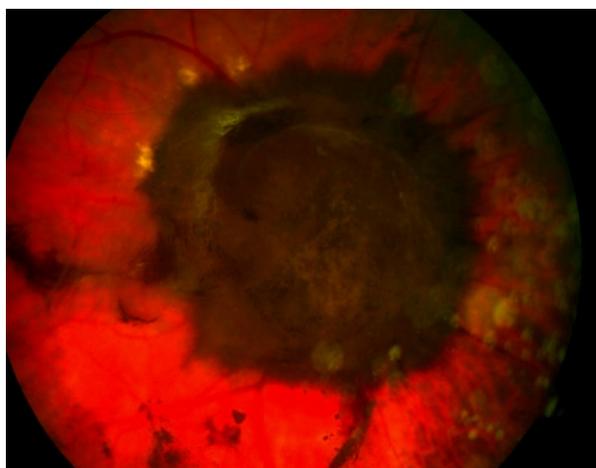


Figure 1. Optic nerve head melanocytoma RE

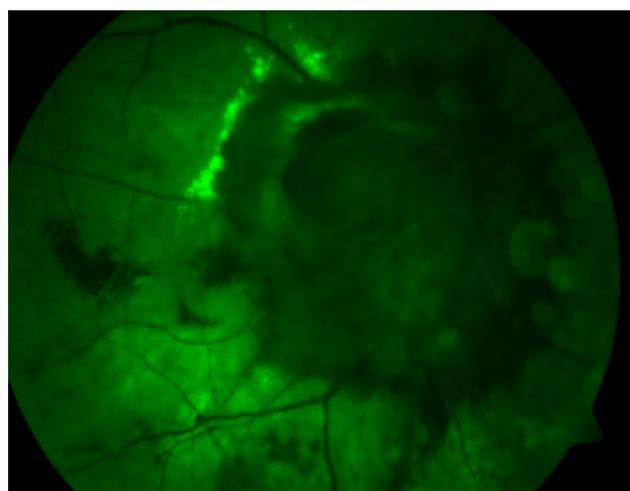


Figure 2. Optic nerve head melanocytoma RE: Red free photograph

A 77 year old lady presented in our clinic with a complaint of diminished vision right eye. She had recently moved to our area and was known to have melanocytoma of the right optic disc and had been treated by Photodynamic therapy for an intraslesional haemorrhage in another centre. Although unrelated to this she also had a history of bilateral epiphora. There was no other relevant past ocular or medical history. Her best corrected visual acuity (BCVA) RE: 6/36+1, and LE: 6/9+2. It was noted that the vision RE 3 months back was 6/18 (documented in the previous records). The lower puncta were stenosed bilaterally. Anterior segments were unremarkable and intra ocular pressures (IOP) were RE 16 mmHg, LE 16 mm Hg. Fundus examination under mydriasis revealed a large right optic nerve head melanocytoma with significant thickness, evidence of old exudates and pigment in the vitreous. There was no evidence of neovascularization. Possibility of low grade malignancy

was not excluded. A Fundus photograph, (Figure 1, Figure 2), Visual fields (Figure 3) Ultrasound (Figure 4), ONH Optical coherence tomography and Fundus fluorescein

angiography were advised. In addition she was put on the list for a bilateral 3 snip procedure.

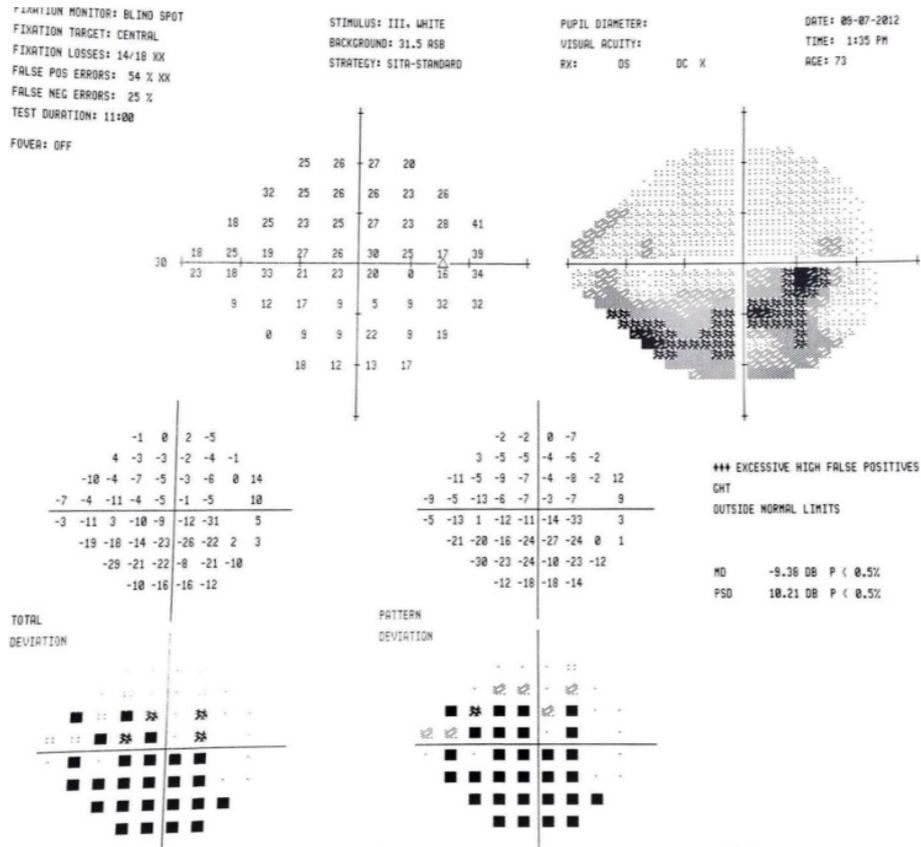


Figure 3. Visual field RE

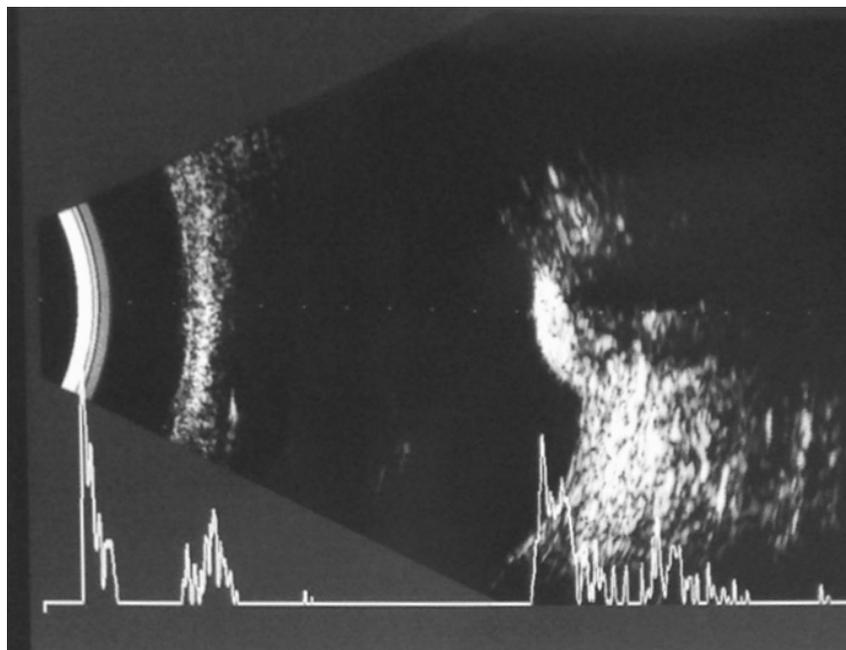


Figure 4. Ultrasound RE

FFA (Figure 5, Figure 6 ) revealed abnormal circulation in the late phase within the ONH mass. Furthermore, there was evidence that this intra papillary circulatory network was incompetent with areas of leakage. OCT revealed submacular fluid and some vitreomacular traction which was likely due to a combination of the exudative process and local laser which was applied in the past.

Comparison of the fundus photos with the old photos revealed no change in the dimensions of the lesion, except for a little more pigment in the vitreous, owing to the fact that the patient had detached her vitreous in that area.

An MRI of the orbits (Figure 7 & Figure 8) revealed no evidence of periorbital or intracranial spread of the ONH melanocytoma.



Figure 5. FFA: early phase

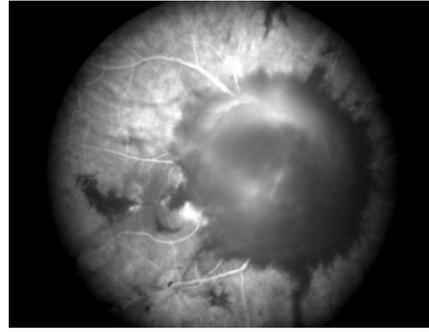


Figure 6. FFA showing abnormal intrapapillary capillary network with a 'blush' in the later phases

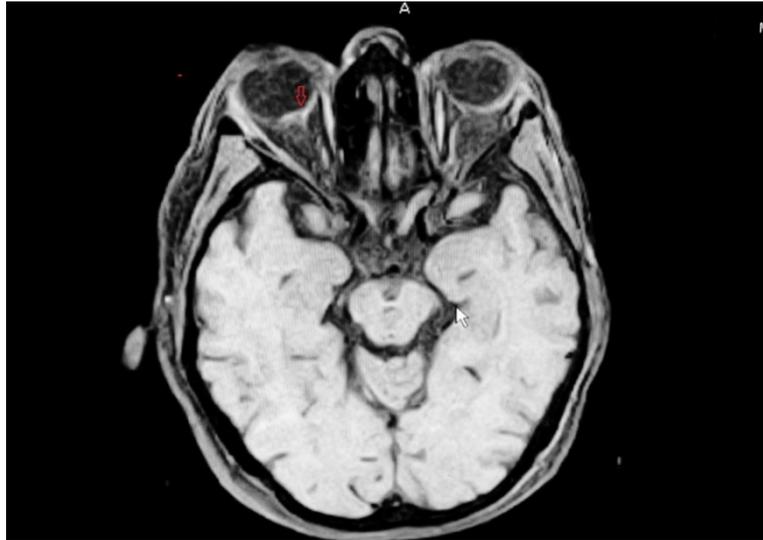


Figure 7. MRI Right Orbit with contrast: The melanocytoma is highlighted

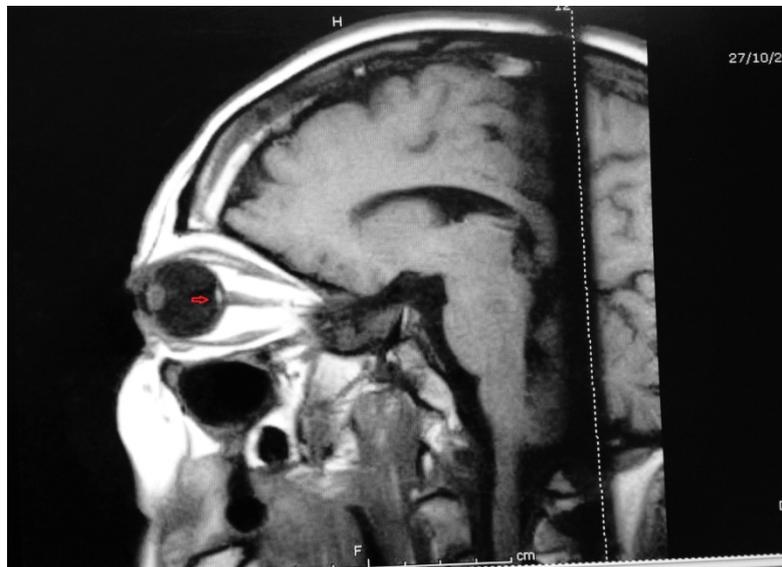


Figure 8. MRI Right Orbit : Sagittal view : No periorbital or intracranial spread.

A life long follow up has planned. She has been periodically reviewed by us for 4 years now with no evidence of extension or further visual impairment.

### 3. Discussion

ONHmelanocytomais a rare benign tumor with rare growth, malignant transformation and visual impairment.

It was believed to be a malignant neoplasm until Zimmerman in 1962, declared it to be a benign tumour and coined the term optic disc melanocytoma, [2] Cogandefined it asoptic nerve magnocellular nevus [3]

Melanocytoma of the ONH is a hamartoma which consists of large round or oval heavily pigmented melanocytes packed closely and located among axons in the optic disc, anterior optic nerve and in the peripapillary retina [4].

The lamina cribrosa of the optic nerve head, with its population of melanocytes, is the origin of these pigmented lesions on the optic nerve [5].

As seen in our case they appear as dark brown or black pigmented lesions with feathery margins due to infiltration into the adjacent retina. Some patients have afferent pupillary and nerve fibre bundle defects possibly due to nerve fibre layer compression.

Zimmerman has reported occurrence of a sudden fall in vision with papillitis and retinal hemorrhages due to ischemic necrosis in a case of melanocytoma [5]. Most melanocytomas are asymptomatic however, mild vision loss can be seen in about 25 % of cases. [6] Retinal extension of the lesion and macular subretinal fluid are risk factors for visual loss. Although rare, severe visual loss can occur as a consequence of central retinal vascular occlusion, spontaneous tumour necrosis or malignant transformation. [6,7] Visual loss by presumed anterior ischemic optic neuropathy and optic atrophy has also been reported [8,9].

Periodic ocular examination is advised in all cases. A study by Lee, Bae et al revealed tumor growth 0% at 1 year, 14% at 5 years, and 57% at 8 years. [10], in another study minor tumor enlargement occurred in 11% of patients by 5 years and in 32% of patients by 10 years. Malignant transformation was documented in 2 patients (2%) [6].

#### 4. Conclusion

Traditionally melanocytoma of the ONH was considered to be a benign, stationary tumor however, it is now known to possess potential for growth and malignant transformation.

Also, when associated with complications like ischemic tumor necrosis, growth, transformation to a malignant melanoma, central retinal vascular obstruction, retinal

extension and subretinal macular exudation, it can be a significant threat to vision.

Careful evaluation, periodic ocular examination and a long term follow up as in our case is therefore advised in all patients with melanocytoma of the ONH.

#### Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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