OPTIC DISC MELANOCYTOMA: A CASE REPORT

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ABSTRACT

Melanocytoma of the optic disc is a rare ophthalmic tumour that arises from melanocytes and is a variant of the melanocytic nevus. It occurs on the optic disc and often extends into the peripapillary retina and choroid. It appears as a dark brown or black lesion, typically small and is almost always unilateral. Although traditionally believed to be a relatively stationary lesion, it is now known to exhibit minor enlargement in 10–15% of cases and can cause minor visual loss by a variety of mechanisms. 1-2% cases can transform to malignant melanoma.

We present a case report of a 65-year-old female whose dilated fundus examination of the right eye revealed a circular, pigmented and elevated lesion about 1.5 mm in size covering almost whole of the optic nerve head except for the superior rim, suggestive of optic disc melanocytoma.

KEYWORDS
Melanocytoma, Optic Disc, Benign Tumour.

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INTRODUCTION: Melanocytoma of the optic disc is an ophthalmic tumour that arises from melanocytes and is a variant of the melanocytic nevus. This pigmented lesion occurs on the optic disc and often extends into the peripapillary retina and choroid.[1] In 1962, Zimmerman named this lesion melanocytoma and declared it to be a benign tumour.[2] Given that optic nerve melanocytomas are composed of intensely pigmented, round or oval nevus cells with benign features and copious quantities of cytoplasm, a more appropriate name for the lesion may be magnocellular nevus of the optic disc.[1] Although traditionally believed to be a relatively stationary lesion, it is now known to exhibit minor enlargement in 10–15% of cases and can cause minor visual loss by a variety of mechanisms. In rare instance, it can induce severe visual loss due to spontaneous necrosis of the lesion or compressive optic neuropathy. More importantly, it can exhibit malignant transformation into melanoma in 1–2% of cases. Ophthalmologists should be familiar with melanocytoma of the optic disc and affected patients should be followed periodically.[3,4]

CASE REPORT: We present a case of a 65-year-old female who presented to the eye outpatient department with the complaints of painless progressive decrease in vision in the right eye for 3 months. She had no previous history of any eye disease and her medical history was noncontributory. Her best corrected visual acuity was 20/400 right eye and 20/80 left eye. Anterior segment examination of both the eyes revealed posterior subcapsular cataract with nuclear sclerosis grade 1. The pupils were round and symmetrical. Both direct and consensual light reflexes were present and there was no afferent pupillary defect present. Intraocular pressures were 14 mmHg right eye and 16 mmHg left eye on applanation tonometry. Dilated fundus examination of the right eye revealed a circular, pigmented and elevated lesion about 1.5 mm in size covering almost whole of the optic nerve head except for the superior rim, suggestive of optic disc melanocytoma (Figure 1). The rest of the fundus examination was within normal limits. Fundus examination of the left eye was normal. Fundus fluorescein angiography (FFA) of the right eye revealed hypofluorescence in the area of the lesion with late staining of the visible portion of the optic nerve head and no evidence of leakage, optociliary shunts or double circulation (Figures 2a & b). Humphrey's automated perimetry showed corresponding superior arcuate scotoma (Figure 3). Optical coherence tomography (OCT) revealed a raised optic disc mass with no evidence of macular oedema in the right eye (Figure 4).

Magnetic Resonance Imaging (MRI) showed small enhancing isointense lesion at optic nerve head of the right globe measuring approximately 2 mm in size suggestive of right optic disc melanocytoma (Figure 5). All the investigations were normal for the left eye.

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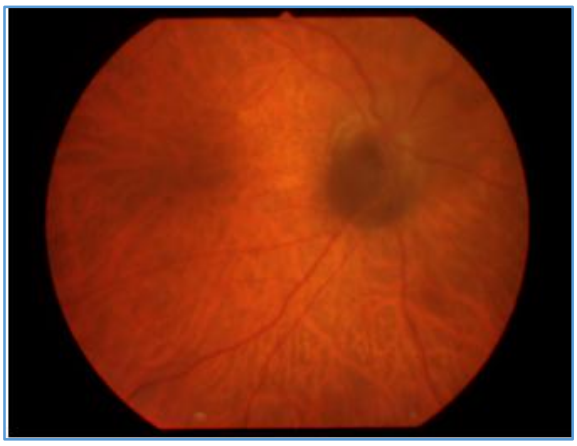

Figure 1: Coloured Fundus Photograph

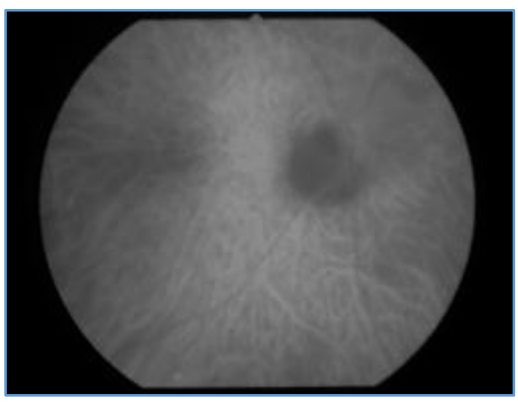

Figure 2a: Red free Fundus Photograph

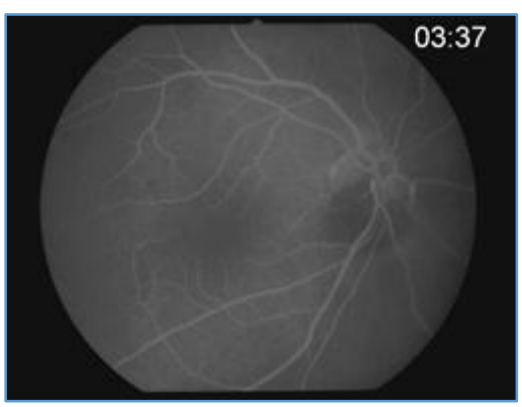

Figure 2b: Fundus Fluorescein Angiography

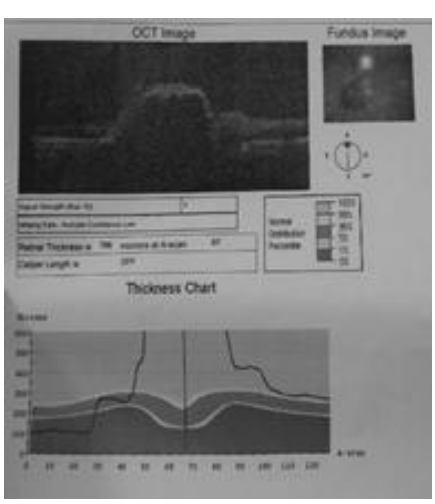

Figure 3: Optical Coherence Tomography

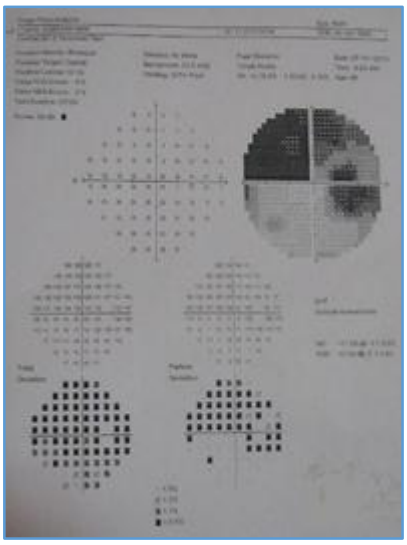

Figure 4: Automated Perimetry

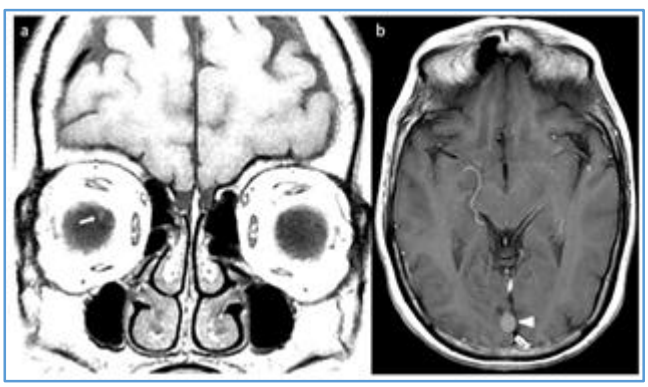

Figure 5: MRI (See the arrow pointing at the lesion)

DISCUSSION: Melanocytoma of the optic disc appears as a dark brown or black lesion. While some melanocytomas are confined to the disc, most cases involve the adjacent retina and choroid. The choroidal component looks identical to a juxtapapillary choroidal nevus, while the retinal component is usually black with a feathery margin. Melanocytomas are typically small, averaging 2 mm in diameter and 1 mm in thickness, and are almost always unilateral. Associated findings include optic nerve oedema, small retinal haemorrhages, retinal oedema, retinal exudates, subretinal fluid and vitreous seeding. The mean age at detection is 50 years and the distribution among races is equal.[1,3]

Before the diagnosis of melanocytoma is made, it is important to rule out a juxtapapillary choroidal melanoma. While melanoma can be juxtapapillary or can occur as a primary lesion at the optic disc, both these occurrences are quite rare. Features typical of choroidal melanoma are a thickness of more than 1.5 mm, subretinal fluid and the presence of orange pigment. Given the typical appearance of a melanocytoma, the diagnosis is generally straightforward and can be made with ophthalmoscopy alone. However, if an optic disc melanocytoma has atypical features, choroidal melanoma can only be ruled out after the patient has been followed over time and no changes suggestive of a melanoma have been observed. Other differential diagnoses are choroidal nevus, hyperplasia of the

retinal pigment epithelium, adenoma of the retinal pigment epithelium and metastatic melanoma of the optic disc.[1,3]

Ancillary testing for the diagnosis of melanocytoma adds little to dilated ophthalmoscopy. Considering the relatively small size of the lesion, ultrasonography, CT and MRI are all of little value. However, ancillary testing can help to quantify the lesion and diagnose any sequelae. In some cases, fluorescein angiography can be used to better demarcate the lesion, which is typically hypofluorescent in all phases unless there is associated subretinal fluid or optic disc oedema. Recently, OCT has been used to evaluate optic disc melanocytoma. Characteristic features of optic disc melanocytoma on OCT include a gradual transition from normal retina into nodular tumour, a bright anterior border layer and dense shadowing associated with no internal detail. OCT can be useful in identifying subretinal fluid associated with the lesion.[1,5]

Histologically, a melanocytoma is a magnocellular naevus with round to oval cells which occurs more commonly in dark skinned individuals.[5] Most melanocytomas do not cause loss of visual acuity but mild vision loss can be seen in about 25 percent of affected eyes. The main cause of associated vision loss is retinal exudation involving the macula. Risk factors for vision loss include retinal extension of the melanocytoma and subretinal fluid. Severe vision loss is rare but can occur secondary to central retinal vein occlusion, spontaneous tumour necrosis or even malignant transformation. While the fundus findings of central retinal vein occlusion are usually straightforward, spontaneous tumour necrosis can result in neuroretinitis or seeding of tumour cells into the vitreous.[1,3]

The transformation of optic nerve melanocytoma to malignant melanoma has been documented in 1 to 2 percent of cases.[3] If there is progressive growth or extensive involvement of the optic disc along with vision loss, then malignant transformation should be considered, although 10 to 15 percent of melanocytomas will have subtle enlargement during followup. An initial tumour thickness of 1.5 mm or more is a risk factor for growth.[3]

At the time of diagnosis, fundus photos should be taken and visual field testing should be performed. An annual followup with dilated ophthalmoscopy and fundus photography is advised. Visual field testing should be repeated if there is suspicion of tumour growth. In addition, OCT can be used both to document the extent of the lesion at diagnosis and to track any progression that may not be apparent with ophthalmoscopy alone. If at any point of time severe vision loss is accompanied by significant growth of the tumour, malignant transformation should be suspected and enucleation must be considered.[6] Extensive involvement of the disc and progressive growth of the pigmented lesion concurrent with vision loss are particularly worrisome for malignancy. However, if the vision loss is a result of ischaemic necrosis, the recovery of some vision may be possible with observation alone.[3,6]

CONCLUSION: Although melanocytoma of the optic disc generally is considered to be a benign, stationary lesion, it can produce several local complications, can cause visual loss, can grow slowly and rarely can undergo malignant transformation into melanoma. Patients with optic disc melanocytoma should undergo periodic ocular examination.[4]

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