CASE REPORT

Presumed Optic Disc Melanocytoma in a Young Nigerian: A Diagnostic Challenge

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ABSTRACT

Optic disc melanocytoma (ODM) is a rare, benign, deeply pigmented ocular tumor arising from melanocytes within the optic disc or from any part of the uvea. It occurs more in dark skinned individuals and females. We report a 17-year-old female who presented to our outpatient department with a history of poor distant vision from childhood, worse in the right eye. Ocular examination revealed visual acuity of 6/36 and 6/18 in the right and left eyes, respectively, which improved to 6/9 bilaterally with a pinhole. There was a relative afferent pupillary defect in the right eye, and a posterior segment examination of same eye showed a raised pigmented optic disc lesion occupying the inferior two-thirds of the optic disc and obscuring the lower disc margin. Both the anterior and posterior segments of the left eye were normal. A diagnosis of presumed ODM was made. Spectacles were prescribed, and the patient was counseled on regular follow-up to monitor progression. ODM should be considered in patients presenting with a pigmented optic disc lesion. Regular follow-up with fundus photography is advocated.

Keywords: Melanocytoma, optic disc, pigmented ocular tumor

INTRODUCTION

Optic disc melanocytoma (ODM) is a rare, benign, deeply pigmented ocular tumor which arises commonly from dendritric melanocytes within the optic disc or from any part of the uvea.[1,2]

It is usually found on the disc and may extend into the vitreous for about 2 mm.[1] It may appear on fundoscopy as a flat or slightly elevated pigmented lesion with fibrillate extensions which may extend over the edge of the disc and may involve the adjacent retina.[2]

Dark skinned races and mediterranean races[1,3] are affected more commonly with a reported female preponderance.[2,4,5]

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Diagnosis is usually incidental when the tumor is detected on routine examination. The mean age at diagnosis is 50 years.[2,6]

CASE REPORT

A 17-year-old Nigerian female presented at our out-patient department on account of painless nonprogressive visual impairment since childhood, worse in the right eye, affecting both distant and near vision. No floaters or flashes of light were reported and no constitutional symptoms. Systemic review and examination were essentially normal.

On ocular examination, unaided visual acuity was 6/36 and 6/18 in the right and left eyes, respectively. This improved to 6/9 in each eye with pinhole. There was a relative afferent pupillary defect in the right eye and posterior segment examination in the right showed a raised pigmented optic disc tumor measuring about

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1.5 disc diameters in the inferotemporal aspect of the disc obscuring the inferior disc margin with overriding normal caliber retinal vessels. There was no retinal vascular tortuosity, no sub-retinal fluid or retinal detachment was seen and macula was structurally normal [Figure 1].

There were no pigments seen in the anterior vitreous on slit lamp examination. The anterior and posterior segments of the left eye were normal [Figure 2].

Intraocular pressure on applanation tonometry was 18 mmHg and 17 mmHg in the right and left eyes, respectively.

Ocular ultrasound scan showed a well-defined homogenous soft tissue mass with broad base arising from the choroid in the optic nerve area and projecting into the vitreous cavity. No retinal detachment or sub-retinal fluid was seen.

An assessment of right presumed ODM was made. She was refracted with visual acuity improvement to 6/5 in either eye and spectacles were prescribed. A fundal picture was taken as a baseline for comparison at subsequent visits, and she was counseled on compliance with follow-up at 6 monthly intervals. On the follow-up visit, 7 months after the first visit, a fundus evaluation showed no changes in the tumor size and color [Figure 3].

DISCUSSION

ODM is seen as a dark brown or black lesion on fundoscopy. In 54% of cases, it extends to the adjacent choroid; in 30% of cases, it extends into the adjacent sensory retina while, in 15% of cases, it does not spread beyond the disc.[6] It is typically a unilateral lesion, bilateral cases have been reported, but they are quite rare.[6]

It rarely measures beyond two disc diameters,[4] and pigment dispersion may be seen in the presence of large tumors.

ODM is commonly described as a stationary lesion[2,7] but may enlarge in about 10–15% of cases.[6] It usually involves the inferotemporal aspect of the optic disc.[1]

It is often asymptomatic with good visual acuity reported in most affected persons; however, mild reduction in visual acuity may be present in about 5% of cases.[8] In a study of 37 cases of ODM at the oncology unit of Jules Gonin Hospital, visual acuity was observed to be normal in 70% and subnormal in 27% of the cases.[9]

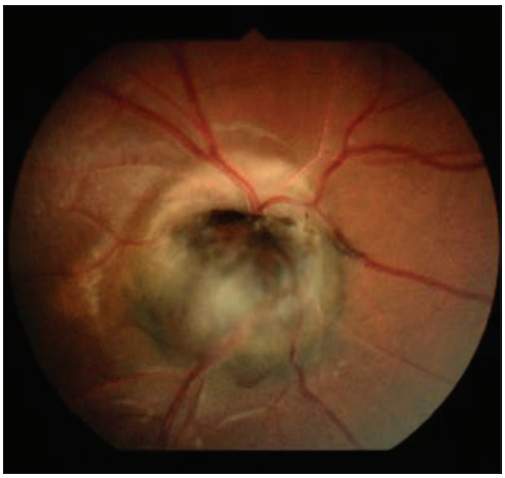

Figure 1: Fundal picture of the right eye at presentation

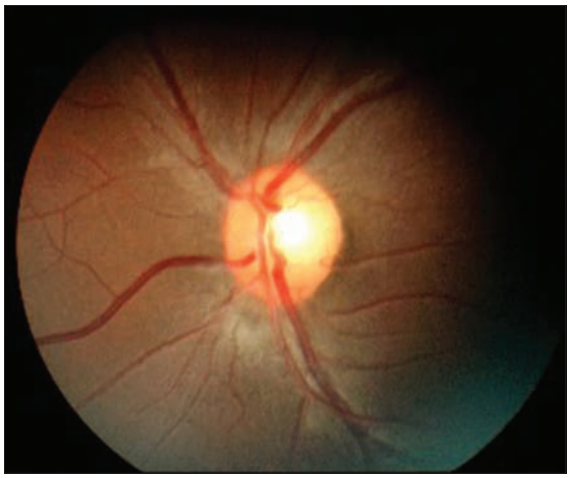

Figure 2: Fundal picture of the left eye at presentation

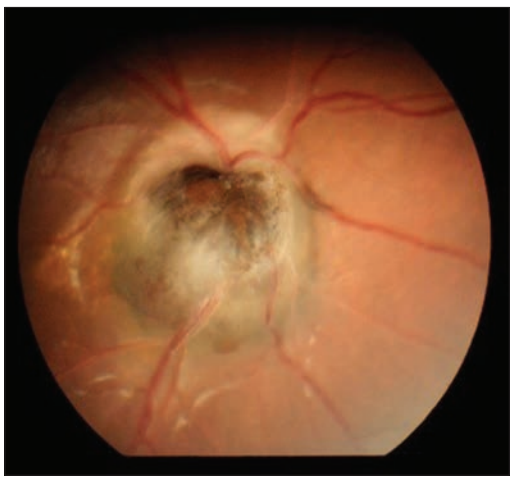

Figure 3: Fundal picture of the right eye 7 months after diagnosis

Sudden reduction in visual acuity may indicate malignant transformation, necrosis, or a hemorrhage within the lesion.[6] Malignant transformation is however rare and is reported in about 1–2% of cases.[5,10]

Afferent pupillary defect may be present, an associated finding in up to 30% of cases,[9] as seen in this patient. It is possibly a result of nerve fiber layer compression, and may co-exist with good vision.[2]

Visual field defects are commonly reported, Osher *et al*. reported up to 90% in their study.[11] These defects may include enlargement of the blind spot, peripheral or fascicular defects, and tubular vision.[11]

Fundus fluorescein angiography is the hallmark of diagnosis. There is hypofluorescence due to close compaction and deep pigmentation of the melanocytes with reduced vascularity.[6] This differentiates it from choroidal melanoma, where in addition to hypofluorescence, there may also be pinpoint leakages and double circulation.[12] We were unable to perform fluorescein angiography in our patient due to financial constraints and the lack of health insurance coverage.

Ultrasonography and computer tomography may be useful for lesions >0.5 mm;[6] however, they cannot differentiate ODM from other raised lesions involving the optic disc.

The features in our patient which are supportive of a diagnosis ODM are a relative afferent pupillary defect, raised pigmented optic disc lesion involving the inferotemporal aspect of the optic disc, absence of progression in size since diagnosis, good and stable visual acuity with spectacle correction. In addition, the patient is female and of African descent. Although our inability to perform a fundus fluorescein angiography in this patient posed some diagnostic challenge, clinically the features appear in keeping with an ODM and a follow-up schedule has been designed to ensure continuous monitoring of the mass.

The differential diagnoses we considered in this patient include Juxtapapillary choroidal nevus, adenoma of the retina pigment epithelium (RPE) and hyperplasia of the RPE.

CONCLUSION

ODM is a rare benign tumor, and its diagnosis should be considered in patients presenting with a pigmented optic disc lesion. Regular follow-up with fundus photography is advocated.

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Conflicts of interest

There are no conflicts of interest.

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