Optic disc melanocytoma is a darkly pigmented mass of the optic disc characterized histopathologically by Zimmerman and Garron in 1962 as a uniform accumulation of heavily pigmented cells with abundant cytoplasm, small nuclei, and inconspicuous nucleoli, all of which are benign cellular characteristics resembling ocular melanosis. In 2004, Shields and colleagues described the clinical features in a large cohort of 115 patients and related, that melanocytoma, although benign, has the capacity to cause significant visual morbidity. They reported that at 10 and 20 years, respectively 18% and 33% will lose two or more lines of vision by Kaplan-Meier estimates. We report a case of optic disc melanocytoma with notable visual field loss.

**CASE DESCRIPTION**

A 33 year-old white man perceived an inferior visual field defect in his right eye for 1 month. Visual acuity was 20/20 in both eyes, and intraocular pressures were 19 mm Hg in both eyes. External and anterior segment examination was unremarkable for both eyes. The patient had brown irides without ocular melanocytosis. Fundus examination of the left eye was unremarkable, but the right eye showed a pigmented lesion occupying the superior half of the optic disc from 10 to 2:30 o’clock, with extension into the juxtapapillary retina superotemporally and lacking a choroidal component (Figure 1). Optic disc edema was present but retinal vessels were not dilated or tortuous. Both fluorescein and indocyanine green angiography revealed hypofluorescence corre-
sponding to the lesion, with some mild leakage on fluorescein angiogram along the inferotemporal optic disc that correlated with disc edema (Figure 2). Visual field examination showed an inferonasal paracentral scotoma on the pattern deviation plot corresponding to the predominantly superotemporal juxtapapillary extension of the optic disc tumor (Figure 3). Magnetic resonance imaging (MRI) of the brain and orbits revealed no deep optic nerve abnormality. These findings were consistent with the diagnosis of optic disc melanocytoma. The patient was monitored yearly.

After 14 years of follow up, visual acuity remained stable at 20/20 in both eyes with no significant change in appearance. The visual field defect remained stable (Figure 3).

**DISCUSSION**

Visual field defects reflect abnormalities in the visual pathway. Much as a neurologist will localize an abnormality to the brain, spinal cord, or the peripheral nerve by correlation with clinical findings, an ophthalmologist can recognize and localize a visual field defect to either the prechiasmal, chiasmal, or post-chiasmal visual pathway. Prechiasmal defects are unilateral and ipsilateral, chiasmal defects will almost always be bitemporal, while post-chiasmal involvement cause visual defects that are contralateral and homonymous. It is important to localize the defect as each site could portend a different prognosis.

Visual field defects can be detected in up to 90% of patients with optic disc melanocytoma. In a series of 115 patients with optic disc melanocytoma studied for visual field defects, the most commonly reported visual field abnormality was enlargement of Mariotte’s blind spot, found in 32% of patients. The size of the blind spot corresponds to the extension of the lesion beyond the boundary of the optic disc. This extension causes a shadowing effect on the peripapillary retina and subsequent blind spot enlargement. Osher and colleagues highlighted this phenomenon by demonstrating that melanocytomas confined within the optic disc displayed normal visual fields.

In addition to enlargement of the blind spot, patients can also display an arcuate defect, quadrantal defect, nasal step, or paracentral scotoma representing impaired axonal flow secondary to mechanical compression directly on the nerve fiber bundles or microcirculation. Nerve fiber bundle defects should correlate to the location of the lesion. Our patient had extension beyond the superior margin of the disc that was mostly superotemporal, resulting in an inferonasal paracentral scotoma. Over 14 years, there was no demonstrable growth of the lesion, and the visual field examination remained stable. Furthermore, when both superior and inferior nerve fiber layers are involved, patients may have double arcuate defects leading to constriction of the visual field and a residual central island. When extensive nerve fiber involvement occurs, patients may also have significant retinal ganglion cell loss and a concomitant afferent pupillary defect, as seen in 9% of cases.

When anatomic findings and functional tests do not correlate, other etiologies should be sought. For example, in a case report by Rai and co-workers, a patient presented with optic disc melanocytoma, but visual fields showed an inferior arcuate defect that correlated with thinning of the superior neuroretinal rim more consistent with glaucoma than melanocytoma. In Zimmerman and Garron’s original series of 28 eyes with optic disc melanocytoma, glaucomatous visual field defects were found in three cases, which further highlights this point. These cases...
emphasize the importance of meticulous examination of the neuroretinal rim in both the involved and fellow eye and correlation with the visual field results. Intracranial meningioma can also be associated with optic disc melanocytoma and should be suspected if visual field defects respecting the vertical midline are found.

In summary, optic disc melanocytoma is a benign pigmented lesion that can lead to a myriad of visual field disturbances. Correlation of clinical and visual field findings is of utmost importance to effectively rule out other etiologies and estimate visual prognosis. Patients with optic disc melanocytoma should be monitored annually for ocular findings and particularly for transformation of the benign tumor into melanoma, a feature found in 2% of cases.

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