

Optic nerve melanocytoma: a case report.

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Abstract:

Melanocytoma is a rare, pigmented tumor, similar to choroidal nevi and most often located in the papilla. Optic nerve melanocytoma usually does not cause significant visual loss and rarely it can transform into a malignant melanoma. The diagnosis is typically clinical, however ancillary testing may aid in the diagnosis and monitoring the lesions. We present a case of optic nerve melanocytoma that was discovered on routine examination in a 42-year-old patient and describe the clinical findings, management and prognosis associated with this type of lesion.

Keywords: Melanocytoma. Pigmented lesion. Ocular tumor. Case report. Optic nerve.

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I. Introduction:

Optic nerve melanocytoma (ONM) is a rare benign pigmented neoplasm on or adjacent to the optic nerve head. Clinically these patients are usually asymptomatic. The ONM is characterized as a dark brown to black lesion with feathery margins in the optic disk. Rarely, these tumors may undergo malignant transformation. Diagnostic tools, such as ultrasonography, visual field analysis, fundus fluorescein angiography (FFA), and optical coherence tomography (OCT) can be used with different diagnostic values.

II. Case Report:

A 42-year-old male, with no relevant past ocular or medical history, presented for a routine examination with no visual complaints. The patient's best-corrected visual acuities were 20/20 OD and 20/20 OS. Anterior segment assessment of both eyes showed no associated conjunctival melanocytosis. Pupils were equally round and reactive to light with no afferent pupillary defect. Extraocular muscle testing was normal. Intraocular pressure was measured at 16 mmHg in both eyes by Goldmann applanation tonometry.

In fundus examination, an elevated dark brown lesion was found, involving upper and lower nasal quadrants of the optic nerve head of the right eye extending to nasal juxta-papillary chorio-retina (Figure 1). No significant findings were present in the left eye. Color fundus photography was performed for baseline documentation.

FFA showed hypofluorescence corresponding to the blocked choroidal and retinal fluorescence by the pigmented lesion in both early and late phases with absence of leakage around the mass (Figure 1). The lesion height was determined to be 1.1 mm by ultrasonography B (Figure 2).

Optical coherence tomography (OCT) scans showed a dome-shaped, elevated optic nerve head lesion with highly reflective layer and optical shadowing behind it, obscuring most of the optic nerve head details without subretinal fluid or cystoid retinal edema (Figure 3).

The management to be taken was an annual monitoring of this papillary lesion made of a complete ophthalmologic examination with fundus retinography and papillary OCT. Over 2 years the visual acuity remained stable and the lesion has not changed in size or shape. This confirmed the benign origin.

III. Discussion:

Melanocytoma is a rare, benign, pigmented tumor, similar to choroidal nevi and most often located in the papilla. It is thought to derive from dendritic uveal melanocytes of the lamina cribrosa [1]. Histologically, They consist of pigmented cells with abundant cytoplasm and small, uniform bland nuclei [2]. They are generally unilateral, the mean age of diagnosis is between the 4th and 5th decade of life with a slight female predominance [3].

ONM usually does not cause significant vision loss like our case reported and the tumor is discovered incidentally. Lee et al. reported that 93% of patients with ONM had vision of 20/40 or better[4].

The ophthalmoscopic appearance of the tumor is still characteristic with a dark “uniform black or brown” generally eccentric staining and overhangs the surrounding chorioretin of the papilla. Their margins are filamentous in "duck feather", due to the infiltration of the layer of optical fibers[2].

On fluorescein angiography, the part of melanocytomas herniating into the vitreous cavity remains hypofluorescent in all phases of the examination and, occasionally, a fine network of stretched retinal capillaries and telangiectasias can be observed on the surface[5].

At optical coherence tomography, melanocytomas appear in the form of an irregular hyperreflectivity of the tumor surface surmounted by a disorganized and hyperreflective retina and a shading of the underlying structures and above all it allows the monitoring as well as the early detection of a possible transformation malignant[6]. B-mode ultrasound can measure melanocytoma thickness and monitor the growth of the lesion[7].

The visual field may show an enlargement of increased blind spot, other visual field abnormalities have been described, such as arcuate defect, nasal step, or paracentral scotoma caused by mechanical compression on the nerve fiber bundles or its microcirculation [8].

The differential diagnosis of melanocytoma should be done first with the malignant melanoma due to its morbidity and mortality, malignant melanoma has as typical characteristics: more than 1.5 mm of thickness, subretinal fluid, presence of an orange pigment, presence of vascularization in the echography and hyperfluorescence of the lesion in fluorescein angiography [8]. Others differential diagnosis are choroidal naevus, hyperplasia of the retinal pigment epithelium, metastatic melanoma to the optic disc and juxtapapillary choroidal melanoma.

Local complications are rare, but can lead to reduced visual acuity: papillary edema, intraretinal edema, macular edema, venous occlusion, tumor growth, tumor necrosis. A malignant transformation is possible but rare, estimated to occur in 1–2% of cases [9]. It should be considered as a malignant risk if there is progressive growth or extensive involvement of the optic disc with vision loss[10]. Therefore, these lesions should be measured and documented at the initial visit and monitored annually to assess any growth.

IV. Conclusion:

Melanocytomas remains stable or grow poorly over a long period of time. On the other hand, the possibility of malignant transformation is controversial, it will be suspected in the face of a increase in lesion size. Their course can sometimes be complicated by severe or even total loss of visual function. This is why it is recommended to monitor their development annually and to document it using papillary OCT or, failing that, retinography.

Statement of Ethics:

All details of this case were reproduced after obtaining consent from the patient and the patient’s next of kin. Any treatments undertaken by the patient were without deviation from accepted standards of care.

Disclosure Statement:

The authors declare no actual or potential conflicts of interest.

Figures:



Figure 1: Fundus photograph and Fluorescein angiography of the right optic nerve showed blockage of fluorescence in the topography of melanocytoma at all times.

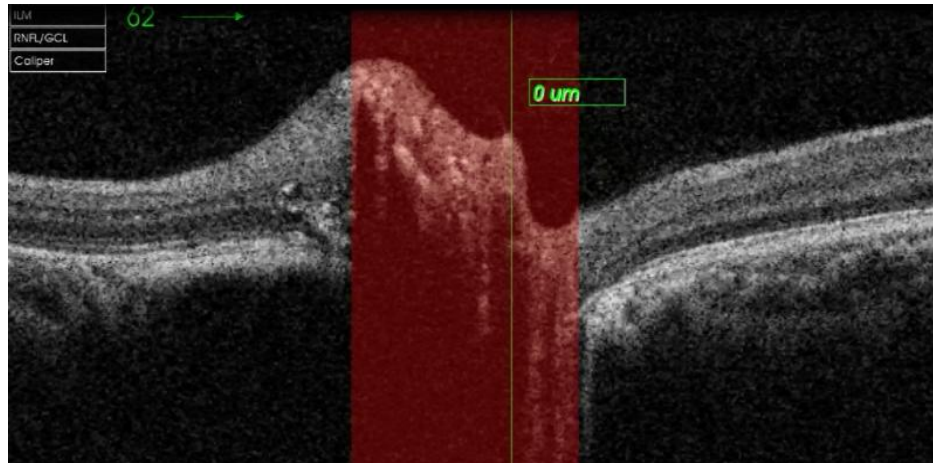


Figure 3: OCT showing elevated lesion in the optic nerve location with high reflectivity and shading.

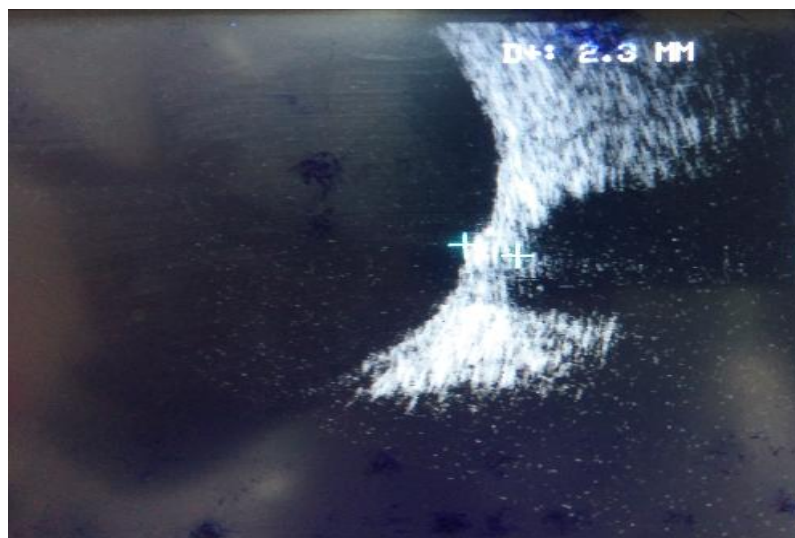


Figure 2: B scan of optic disc melanocytoma.

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