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Case Report

Pigmented Epitheloid Melanocytoma: When a Benign Tumor Simulates Malignancy

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Abstract

Melanocytoma is a rare intraocular tumor. Some rare cases of primary melanocytomas of the choroid and ciliary body cases have been reported in the literature. We believe that most of these tumors are clinically diagnosed as nevi or melanomas and are respectively followed up or treated by surgical resection. Some clinical features can guide the diagnosis. We report the case of a 51-year-old Caucasian male who, for two months, had a gradual loss of vision in his right nasal visual field.

Upon clinical examination, a large intraocular pigmented tumor was discovered. Both the clinical characteristics and the paraclinical testing supported the diagnosis of choroid malignant melanoma. Thus, the risk of spreading prompted enucleation. The anatomical piece's macroscopic examination revealed a highly pigmented tumor. Nevertheless, a later histological analysis identified it as a benign choroid-pigmented melanocytoma. Compared to other findings, our case is peculiar due to the patient's young age, the tumor's size, and its clinical and paraclinical appearance.

Keywords: Melanocytoma; Choroid; Melanoma; Enucleation

Introduction

The pigmented epithelioid melanocytoma, a unique variation of the melanocytic naevus, was initially described by Zimmerman and Garron in 1962 [1]. It is an uncommon tumor, accounting for only 0.6% of all intraocular tumors. It is a benign tumor that seldom invades local tissues and never metastasizes [2]. The incidence of such tumors is undetermined since only those confirmed by histological examination have been reported in the literature. Certain naevuses treated by ophthalmologists and melanomas treated without surgical excision might be melanocytomas. However, most choroid and ciliary body melanocytomas are categorized as melanomas [3]. Typically, it is found near or on the optical disk. However, in rare circumstances, it can be seen anywhere along the uveal tract (iris, cilia, and choroid) or on the conjunctiva or sclerosis [4]. Melanocytoma is treated similarly to naevus, with frequent monitoring. We present a case of a large pigmented epithelioid melanocytoma on the anterior choroid, clinically identified as malignant melanoma.

Case Presentation

We report the case of a 51-year-old man, caucasian, followed up for diabetes, who consulted for a painless gradual visual loss in his right nasal visual field for two months without associated redness. The patient did not report a history of trauma or other systemic symptoms such as weight loss or headaches. The clinical examination showed visual acuity 5/10 in the Right Eye (RE) and 10/10 in the Left Eye (LE). Slit lamp biomicroscopy examination of the right eye revealed no anterior segment abnormalities. Fundus examination revealed a massive brownish mass in the temporal macular zone, occupying most of the posterior segment and masking the posterior pole, associated with localized retinal detachment (Figure 1).

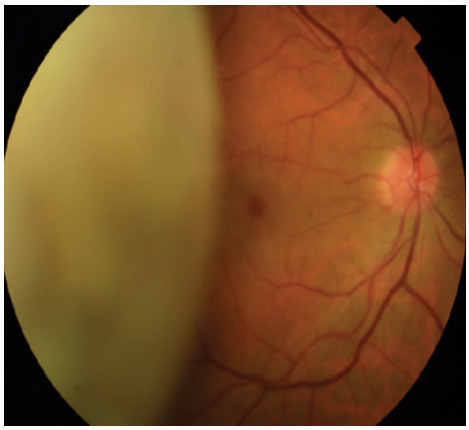

Figure 1: Image of the background of the right eye showing a large temporo-macular tumor masking the posterior pole associated with a localized retinal detachment.

Furthermore, neither pigment nor druses were visible on the tumor or the episcleral sentinel vessel. Examination of the LE was unremarkable. Both eyes showed open angles on the gonioscopy. Intraocular pressure was normal, and pupil reflex was present in both eyes. B-scan ultrasound of the RE revealed a dome-shaped hyperechogenic choroidal mass measuring 14 mm × 12 mm × 6 mm, with no internal reflectivity or calcification and accompanied by a localized retinal detachment. The tumor remained motionless while the globe moved (Figure 2).

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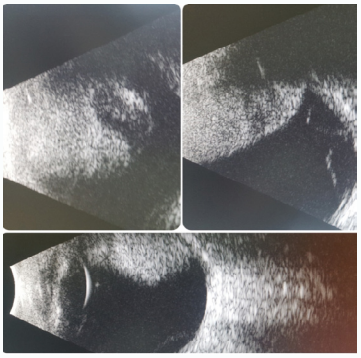

Figure 2: B mode ultrasound showing a hyper-echo-temporal biconvex mass located on the topography of the cilia and choroid of the right eye and measuring 12 mm × 6 mm associated with retinal detachment.

Orbito-cerebral Magnetic Resonance Imaging (MRI) revealed a large intraocular mass measuring 18 mm × 12 mm associated with choroidal detachment without any scleral invasion (Figure 3). Extent assessment (radiography; liver ultrasound, bone scintigraphy) revealed no signs suggestive of metastasis. Blood counts, erythrocyte sedimentation rate, and liver and kidney functions were also within normal limits. The primary diagnosis retained was choroidal melanoma. The patient agreed to enucleation to reduce the danger of spread with histological investigation. Subsequently, implantation with a silicone ball went successfully.

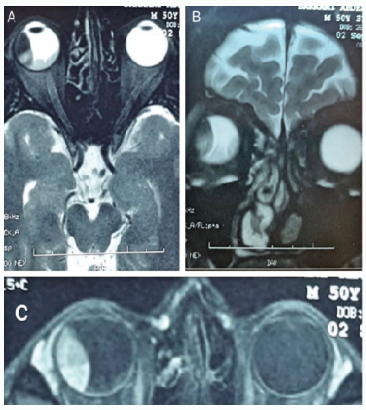

Figure 3: Cerebral orbital MRI: the intra-ocular process of the right posterior-external wall into T1 hyperintense and T2 hypointense measuring 18 mm × 12 mm heightened intensely after Gado injection, associated with a choroid detachment; evoking firstly choroidal melanoma.

Macroscopic examination revealed a globe measuring 2.3 cm × 2.4 cm × 1.9 cm. The healthy cornea measures 1.2 cm in diameter. The incision (from the pupil to the optic nerve) revealed a blackish lesion with whitish fleshy areas measuring 1.2 cm × 0.6 cm and placed 0.5 cm from the optical nerve (Figure 4).

Microscopic investigation revealed a tumor on the anterior choroid (Figure 5). The tumor was homogeneous and highly pigmented. The dark tint hid the appearance of tumor cells. These were regular fusiform with many melanophages on a melanin-rich background. Melanocytic cells had no cytonuclear a typical and were morphologically homogenous, with a tiny circular core, a normal membrane, and a low nucleo-cytoplasmic ratio. No atypical mitotic figures or conspicuous nucleoli were seen. The cytoplasm was plentiful. There was no evidence of scleral invasion, extra-scleral expansion, or anterior chamber obstruction. Furthermore, the tumor did not reach the visual nerves.

The morphological appearance was compatible with type I choroidal melanocytoma, not choroidal melanoma, as suspected on clinical examination, B-ultrasound, and MRI. The patient was diagnosed with pigmented epithelioid melanocytoma of the anterior choroid and implanted with prosthesis six weeks after enucleation. No tumor recurrence was seen after six months of follow-up.

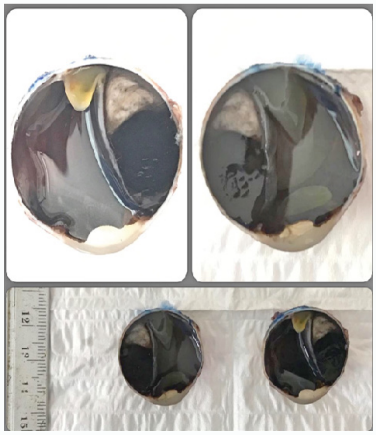

Figure 4: Section of the eyeball (pupil - optic nerve) showing a globe measuring 2.3 cm × 2.4 cm × 1.9 cm. In the healthy cornea, a blackish lesion with some fleshy whitish areas was noted, measuring 1.2 cm × 0.6 cm, located 0.5 cm from the optic nerve.

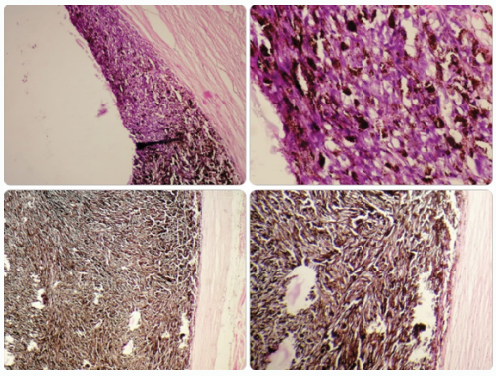

Figure 5: Highly pigmented melanocytic tumor proliferation, with spindle cell architecture. The tumor cells are elongated, with regular, roughly uniform ovoid nuclei, without atypical or mitoses.

Discussion

Intraocular melanocytomas are rare and benign tumors. Until 1999, 40 cases of uveal melanocytomas had been described [3,5]. The term "melanocytoma" was coined by Zimmerman to describe a specific type of tumor, but it has limitations as it could potentially refer to any tumor arising from melanocytes, including malignant melanoma. At a previous meeting, Cogan disputed the term "melanocytoma" and suggested a different term for a naevus located in the optic nerve. Shields later proposed the term "hyperpigmented magnocellular naevus," which seems more suitable but has not been widely adopted, leaving "melanocytoma" as the prevailing term.

In a recent study, only 5% of iris and ciliary body tumors were identified as melanocytomas. A literature review showed

approximately a dozen reported cases of choroid melanocytomas. However, there is a possibility that many melanocytomas are being misdiagnosed as naevi or melanomas, leading to treatment with laser photocoagulation or radiotherapy. The accurate diagnosis of melanocytomas typically occurs after local resection or enucleation, with confirmation through histopathological examination [6].

Shields et al. found that melanocytoma, whether located at the optic nerve head [7] or uvea [4], is more frequently observed in individuals of Caucasian descent compared to those of African descent. LoRusso, et al estimated this predominance at 80% [3]. Interestingly, melanocytoma is more prevalent in black individuals compared to melanoma, as opposed to veal melanoma. Our patient had a light skin color, which is consistent with the literature.

Often, patients are asymptomatic, and the tumor is found only accidentally during a routine ophthalmological examination. Sometimes blurred vision, eye inflammation, or pain can be part of the functional signs [3,4]. Our patient showed only a progressive amputation of visual field over two months, with preserved visual acuity. The tumor is typically diagnosed in patients between 30 and 50 years old, with an average age of 46.5 years [3]. Our patient's age aligns with the findings in the literature.

When melanocytoma invades the anterior chamber and iridocorneal angle structures, it generally does not lead to increased intraocular pressure. However, cases with necrosis are more likely to develop melanocytomalytic glaucoma. Furthermore, extra scleral invasion is rare in melanocytoma with only isolated cases reported. In our patient's case, there was no glaucoma or extra scleral invasion. When observed directly, melanocytomas are typically uniform in appearance, but cystic changes or necrosis may occur. On the other hand, melanomas are not uniform in color and may have different shades of black. Sub-retinal fluid and orange pigment with lipofuscin, present in our case, are typical markers of malignant tumors. However, recent reports suggest that these signs may not be specific to malignant tumors and could be present in both benign and malignant tumors. The histopathological examination of melanocytomas, different from the classic melanoma or uveal naevus, shows that the tumor is composed of uniformly large cells, rounded to polygonal, with a distinct cytoplasmic membrane, an abundant cyto-plasma with a rounded core, of small regular size without apparent nucleoli [8]. Nevertheless, an abundance of melanin masks the cell morphology, and special melanin removal techniques are generally needed to make the correct diagnosis [2,6].

Ultrastructural studies of melanocytoma, as reviewed by Al-Hinai et al, revealed two types of cells: type I and type II. The characteristics of type I cells are small and polyhedral, round nuclei, and a cytoplasm containing giant melanosomes. On the other hand, type II cells have an elongated cytoplasm and small melanin granules [8] and are more metabolically active than type I cells. Type II cells are responsible for tumor growth and invasion of surrounding structures. In our case, type II cells were more dominant.

In our case, the tumor measured 6 mm in height and 12 mm at the base, falling within the average size range of 3.0 mm to 15 mm, with an average of 5.7 mm [3]. These tumors typically grow slowly and rarely become malignant. Melanocytomas can undergo spontaneous necrosis and cystic degeneration due to high cell metabolism and low vascular supply. They may also lead to vaso-occlusion, especially at the papilla level, resulting in neovascular glaucoma. Other potential complications include ischemic retinopathy and melanocytic glaucoma from melanin pigment dispersion [9]. Thus, older lesions are more likely to undergo necrosis and develop cysts. Despite symptoms appearing for two months in our case, we cannot confirm the age of the lesion because no cystic changes or areas of necrosis were observed.

When melanocytoma is suspected, a retinography, a fluoresce in Angiography (AGF), a B-mode ultrasound, and a fine needle aspiration biopsy with cytological analysis are necessary to confirm the diagnosis before removing the tumor. During the AGF, the dense and relatively a vascular nature of melanocytomas usually cause the masking of choroidal fluorescence. However, some studies have shown that these tumors can also develop vascularization, though it is less common than in melanomas. Therefore, benign and malignant lesions should not be distinguished solely based on the presence or absence of vessels [10]. On B-mode ultrasound, melanocytomas appear highly internal hyper echoic compared to malignant melanoma [11]. The differential diagnosis of melanocytomas includes, in addition to malignant melanoma, several conditions such as naevus, Retinal Pigment Epithelial Hyperplasia (REP), and EPR or sensory retinal hamartomas [11]. Unfortunately, melanocytomas can feign a malignant melanoma clinically and/or radiologically, leading to drastic procedures such as nucleation [12].

Periodic checks every three months are enough to effectively monitor regular melanocytomas, particularly those on the optic disc [13]. However, if there are complications or signs of malignancy, surgical removal should be considered for small tumors by raising a partially thick area of the sclera and performing a choroidectomy. Sometimes, the diagnosis is not confirmed, and the eye is removed out of concern for a malignant growth [14]. In our case, as in many others, a melanocytoma is only confirmed after the eye is removed and examined pathologically [2].

Conclusion

Ophthalmologists should consider melanocytoma as a potential differential diagnosis for ocular tumors. Some clinical characteristics can provide valuable clues for diagnosis through a thorough ocular examination. Our case is notable due to the relatively large size of the tumor and the presence of some traits that resemble malignant melanoma. Conservative treatment could preserve the patient's vision and quality of life, but in most cases, they are treated similarly to melanomas. Conversely, pathologists should be mindful of this rare entity, as it is a specific eye tumor that may not be familiar to general pathologists.

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