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Case report
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Sudden Ocular Pain: An Unusual Debut of a Ciliary Body Melanoma

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Abstract

The Ciliary body is an unusual location of uveal melanomas and usually these kinds of tumors appear with a reduction of vision due to the formation of sectorial cataract or retinal detachment when the tumor spread to a posterior position. We present a 57 years old woman with sudden and severe ocular pain. Ophthalmologic examination showed a hyper mature cataract and superior displacement of the lens with sectorial angular block. The intraocular pressure was 22 mmHg. After papillary dilation aciliary body tumor was observed. The ultrasound study and magnetic resonance imaging confirmed the diagnosis of uveal melanoma.

Keywords: Ocular Pain; Melanoma; Ciliary Body; Cataract

Abbreviations: BCVA: Best-Corrected Visual Acuity; NMR: Nuclear Magnetic Resonance

Introduction

Uveal melanomas are the most frequent intraocular primary tumors in adulthood. However, ciliary body location is very uncommon regarding iris or choroidal melanomas. Furthermore, ciliary body location gives symptoms later, which also gives a worse prognosis.

Materials and Methods

Biomicroscopy, dilated indirect ophthalmoloscopy, Goldmanntonomter, B-Mode echography and ultrasound biomicroscopy were performed. Subsequent RMN imaging was also obtained.

Case report

We report a case of a 57 years old woman who came to the emergency department with a sudden and severe pain in her right eye. The patient also related that she had had a decrease in her visual acuity for last two months. She was a nonsmoker, but would occasionally drink alcohol. Family history was unremarkable for malignancy. Medical history only included controlled hypertension and cholesterolemia. Best-corrected visual acuity (BCVA) at first was 20/200 in the right eye and 20/30 in the left eye.

The examination of the right eye showed a brown pigmented lesion in the lower region of iris, without satellites lesions or papillary deformity. She also presented a hyper mature cataract antero-superiorly displaced endangering the anterior chamber space and the iridocorneal angle (Figure 1). The intraocular pressure was 22 mmHg in the right eye and 16 mmHg in the left eye. After papillary dilation, we found a pigmented tumoral mass behind the iris which was situated between 4 to 6 hours and produced the dislocation of the lens (Figure 2).The retinal examination was of no value because of the presence of the mature cataract. The ophthalmoscopic examination of the left eye was normal.

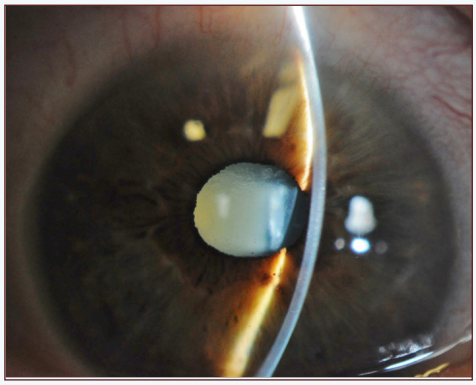

Figure 1: Hyper mature cataract antero-superiorly displaced with superior iridocorneal angle closure.

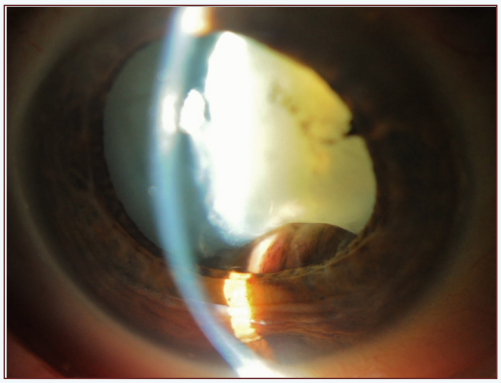

Figure 2: Pigmented tumoral mass behind iris that produces lens displacement.

Results

The study with B-Mode echography and ultrasound biomicroscopy (OTI HF 35-50 Ultrasound System ®) demonstrated a 15 X 10 mm tumor which came from ciliary body and displaced the lens (Figure 3). The nuclear magnetic resonance (NMR) with gadoline confirmed the existence of an intraocular tumor with hyper intense signal in T1 and hypo intense in T2 (Figure 4) with a size of 15 x 10 x 8 mm and heterogeneous catchment of contrast (Figure 5). With these results a ciliary body melanoma was suspiciously diagnosed. The extensive study was negative and a brachytherapy with of iodine 125 (I125) treatments was decided. Actually and after cataract surgery our patient is asymptomatic and without signs of recurrence.

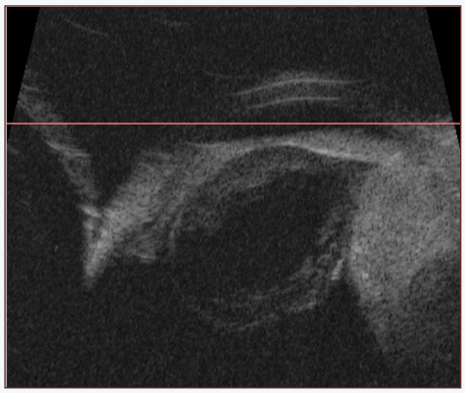

Figure 3: Ultrasound biomicroscopy: Ciliary body mass that displaces lens.

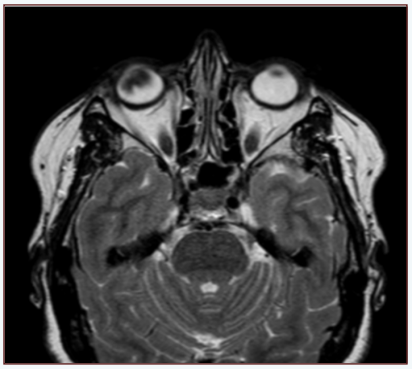

Figure 4: Nuclear magnetic resonance: Intraocular tumor with hypo intense signal in T2.

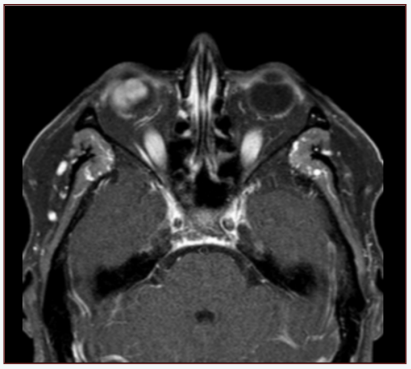

Figure 5: Nuclear magnetic resonance: Intraocular tumor with heterogeneous catchment of contrast.

Discussion

Ocular melanoma is the most common intraocular primary tumor in adulthood, which mainly affects white males between fifty and sixty years old. It represents 3.7% of cases of melanoma and it is the second most usual location after cutaneous melanoma [1]. Its incidence is nearby 4.3 cases per million populations [2]. And its most common position is the uveal tract, which is supposed the origin of the 82.5% of intraocular melanomas [1]. Although inside this rate, ciliary body melanoma only represents 12% of uveal melanomas its location is a factor of poor prognosis [3].

The majority of these tumors are asymptomatic until their growth produces a decrease of visual acuity due to the formation of a sectorial cataract or retinal detachment [2]. The presence

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of dilated episcleral vessels near the tumor (sentinel vessels) is also frequent [4]. Unusually these tumors may also produce an increase in intraocular pressure because of the angular closing by the pigment dispersion produced by myeloma [5] a uveitis due to necrosis of tumor [6] or aleukocoria in children [7]. There is no existing record of a case of ciliary body melanoma that appears with sudden and severe eye pain without a significant increase of intraocular pressure. In the case I am talking about this circumstance happens as well as other manifestations like the presence of a mature cataract, lens subluxation and a sectorial angular closure.

The diagnosis of this pathology is clinical using the slit lamp, accompanied by additional tests such as mode A ultrasound (presence of kappa angle); mode B ultrasound or ultrasonic biomicroscopy (the size, location and extent of tumor is characterized) and orbital NMR (show tumor mass as a hyper intense on T1 and hypo intense on T2) [8]. The confirmation of the diagnosis is obtained by the histological study of the lesion.

Pathological study is also important to establish the prognosis of these melanomas because there are some factors that can add to a worse prognosis for this kind of tumors like epithelioid cell line, high mitotic activity, necrosis of tumor, lymphocytic infiltration or extra sclera extension [9]. Genetic factors are other important prognostic. The mutation in genes 3 and 8 is related to poor prognosis in these tumors [10].

Regarding the treatment of these tumors, there are several alternatives such as radiotherapy, brachytherapy or external beam radiation; local excision (iridocyclectomy) or enucleation. Depending on the size of the tumor and the presence or absence of distant metastases we would choose a treatment or another. Our patient has been treated with brachytherapy due to the characteristics of the tumor and the absence of extraocular disease.

Ciliary body melanomas have a rate of distant metastasis of19% and 33% at 5 and 10 years respectively, being the most frequent liver, lung or bones metastases [1]. This rate is the highest of uveal melanomas, which makes the survival from these tumors, also lower [3].

Conclusion

In conclusion we suggest that sudden ocular pain is a very unusual presentation of already unusual ciliary body melanoma. The objective of this article is describing this way of atypical debut. These tumors are the most metastatic inside the group of intraocular melanomas. However the brachytherapy treatment can be a good choice for this kind of tumors if we detect them before extra ocular dissemination.

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