

Case Report

Ciliary Body Melanoma: A Case Report With A Satisfactory Clinical Evolution

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Abstract

Ciliary body melanomas are rare uveal disorders. Due to its anatomical location, it is usually incidentally discovered in advanced stages. There is no established treatment for metastatic disease and the one-year survival rate of these patients is only fifteen percent. We intend to report a case of ciliary body melanoma with good clinical outcomes.

INTRODUCTION

Ciliary body melanomas represent twelve percent of the uveal melanomas. Usually asymptomatic, it is incidentally discovered in the sixth decade of life. Owing to the anatomy of the eye, commonly the diagnosis is only made in advanced stages [1].

The present article intends to report a ciliary body melanoma case and literature review.

CASE PRESENTATION

Male, 80 years, one-eyed, right eye vision loss after a central retinal vein occlusion that evolved to neovascular glaucoma, has been attended at Federal Fluminense University Hospital Antonio Pedro for left eye (LE), cataracts surgery evaluation. At the examination, best-corrected visual acuity (BCVA), was no light perception right eye (RE), and 20/50 LE. Biomicroscopy revealed punctate keratitis and vascularized hyperpigmented nodular lesion between bulbar conjunctiva and inferior tarsus in the RE (Figure 1). IOP was 30 / 12mmHg. Fundoscopy was impossible in RE and showed mild hypertension retinopathy at LE.

After four months of follow-up, the RE conjunctival lesion evolved with ocular pain and gradual growth.

The diagnosis of ciliary body melanoma was suspected and ophthalmologic ultrasonic biomicroscopy was realized revealing a large solid mass emerging from the ciliary body with extension to practically all the orbit.

Brain and Orbit magnetic resonance images were performed unveiling heterogeneous expansive solid intraocular lesion filling the eye posterior chamber indissociable from the ciliary body. Thorax and abdomen computed tomography were also realized with no further finds.



Figure 1 Biomicroscopy illustrating vascularized hyperpigmented nodular lesion between bulbar conjunctiva and inferior tarsus in the RE.

After the evaluation of the Retina and Oculoplastic Departments, we opted for RE enucleation for diagnostic and therapeutic purposes.

Histopathological examination evidenced spindle cells uveal melanoma, with 1,7x1,2cm, with rare mitotic figures and extensive tumour necrosis. Intra and extrascleral lateral and posterior involvement, with nerves (including optic nerve) and vessels, lens, and sectors of the iris all involved.

Seven months after enucleation patient has maintained excellent clinical and ophthalmological evolution.

DISCUSSION

Ocular melanomas corresponding to five percent of all melanomas and eighty-five percent of them have its origin in the

uveal tract.¹Ciliary body melanomas are extremely rare, and the overall incidence of all uveal melanomas are of only 7/ 1,000,000 per habitant. Due to its position, they normally are not detectable until they reach great proportions and, therefore, were in a more advanced stage [2]. The expansive lesion of our patient was already voluminous on the first detection, but by diagnostic agility, we conquer a fine prognosis.

Melanoma etiology remains, mainly, unknown. Several factors have been associated with an augmented risk of developing uveal melanomas. Some phenotypic traces, as light iris, and light skin are considered risk factors. Other factors associated with the development of uveal tumors were uveal nevus, dysplastic nevus syndrome, ocular melanocytosis, neurofibromatosis type 1, and as probable risks, HIV infection, pregnancy, and use of estrogens or levodopa [3]. Chronic exposition to the sunlight is controversial in etiology of uveal melanoma. Unlike what happens with the cutaneous melanomas, the ultraviolet light was described as a minor risk factor in the uveal melanoma development [3]. Our patient has light skin and iris, hypertension as his only declared comorbidity, and he also denied intense sun exposure.

The clinical signs of this malady include exudative retinal detachment, acute anterior uveitis due to tumoral necrosis, and episcleral sentinel vessels nourishing the tumour. Curiously, these tumours can initiate with low IOP, followed by high IOP and even glaucoma. The lens or cataracts subluxation may occur as well [2]. Our patient had high IOP both by neovascular glaucoma as by the lesion presence. The ultrasound also revealed serous retinal detachment.

The differential diagnosis includes metastasis, ciliary body adenoma, ciliary body cyst, uveal effusion syndrome, and ocular medulloepithelioma [2].

In terms of treatment iridocyclectomy, brachytherapy, probe beam therapy, and as, the last resource, enucleation, may be used [2]. Once the hospital has no access to high technology, and the

BCVA and disease stage allowed, enucleation was elected as the diagnostic and therapeutic measure.

Even though the advances in diagnosis and improvement in the efficiency of the treatment, the survival rate has not evolved, as well as, the occurrence of metastasis [3]. The prognosis is not favorable with fifty percent of uveal melanoma patients developing metastatic spread. The liver is the most commonly affected organ [2].

In the case described in our article, the clinical evolution was excellent. In the last seven months, no metastatic signs were detected in the follow-up.

The literature suggests that the one-year survival rate of uveal melanoma patients is fifteen percent, this fact highlights the need for early detection [2].

Currently, several clinical trials, including chemotherapy, immunotherapy and targeted molecular therapy for the treatment of metastatic uveal melanoma are in progress [3-5].

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