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Ciliary Body Melanoma Masquerading as Chronic Uveitis

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Introduction

The incidence of uveal melanoma is about six per million per year; 80% in the choroid, 12% in the ciliary body, and 8% in the iris. Malignant melanoma of ciliary body is often asymptomatic, and as a result may reach larger dimensions before they generate clinical symptoms in the patient. The symptoms that may develop in patients include visual field changes and progressive deterioration of vision. One of the most notable signs is dilation of the episcleral vessels (sentinel vessels) in the quadrant corresponding to the tumor's location. The tumor may also invade the iris or sclera. Ciliary body melanoma yields poorer prognosis compared to other uveal melanomas.

In this study, we report a case of unilateral ciliary body melanoma presenting as chronic uveitis and ocular hypertension.

Case Report

A 62-year-old man presented with a five-year history of intermittent uveitis and ocular hypertension in his right eye. His symptoms began following uneventful cataract surgery with intraocular lens implantation. He subsequently underwent pars plana vitrectomy, which showed no evidence of infection or malignancy.

When we saw the patient at the initial visit, he had moderate panuveitis with an intraocular pressure of 28 mmHg in the right eye. First, we ordered a comprehensive uveitis workup. Due to the severity of the inflammation, the patient was treated with diflunisal 500 mg orally twice daily, along with topical prednisolone, diclofenac, apraclonidine, and timolol in the right eye. All laboratory tests were negative or within normal limits; however, despite treatment, the inflammation persisted for another two months. Oral methotrexate therapy was initiated at a dose of 7.5 mg weekly. Over the next several months, the patient continued to have persistent iridocyclitis with elevated pressure in the right eye.

At this time, we decided to perform a diagnostic pars plana vitrectomy in the right eye. During the surgery, we detected a brownish lesion at 12 o'clock in the ciliary body at 12 o'clock position suggestive of ciliary body melanoma. Post-operatively, ultrasonography confirmed a lesion of moderate echo adherent to posterior iris, within the ciliary sulcus. A consultation was obtained from the retina service, and observation of the lesion was recommended. Over the next six months, the patient continued to experience persistent uveitis and elevated intraocular pressure. We then decided to remove the intraocular lens as a potential source of inflammation; however, this intervention did not resolve the condition, and the patient continued to have persistent inflammation with elevated intraocular pressure over the following six months.

A year later, retinal examination and ultrasound biomicroscopy showed that the ciliary body mass had enlarged, to approximately 7.0 x 6.0 x 4.2 mm, with extension into the limbus. Evaluation by the retinal oncologists confirmed the suspicion that the mass was probably a ciliary body melanoma. The patient elected to undergo proton beam irradiation and received a total of 70 Gy delivered in five fractions. One month after his proton beam treatment, the uveitis had dramatically improved, and the intraocular pressure had returned to normal.

Discussion

Despite the lack of pathologic confirmation, it is highly likely that the patient had a ciliary body melanoma based on the clinical course and response to treatment.

Shields and colleagues reported that secondary intraocular pressure evaluation was present in 17% of eyes with CB melanomas. The most common mechanism of elevated intraocular pressure was pigment dispersion and tumor invasion of the angle in CB melanomas; however, uveitis is rarely associated with CB melanoma. There is another published report in the literature of a case of ciliary body melanoma masquerading as anterior uveitis and invading the optic nerve, ultimately leading to enucleation. It is believed that tumor necrosis induces uveitis.

Treatment of choroidal melanoma depends on tumor size, location, presence of metastasis, and visual potential of the eye. Small to medium-sized tumors are most commonly treated with plaque brachytherapy. Other eye-preserving options include proton beam radiotherapy and, in selected cases, transpupillary thermotherapy or stereotactic radiosurgery. Large tumors, those with extrascleral extension, or eyes with no useful vision may require enucleation. In cases with metastatic disease or high risk of spread, systemic evaluation and management are essential, including liver-directed therapies, immunotherapy, or clinical trials, since the liver is the most common site of metastasis. Regular long-term surveillance is critical regardless of treatment modality.

Conclusion: intraocular malignancy should be considered in cases of chronic uveitis that do not respond to aggressive medical therapy. Ciliary body melanoma may present as refractory glaucoma and chronic uveitis. Direct treatment of the melanoma may be required to control both the uveitis and glaucoma.

References

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