Ciliary Body Melanoma Masquerading as Chronic Uveitis

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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 involving ciliary body (CB) yields poor prognosis compared to other uveal melanomas. We report a case of unilateral CB melanoma that we believe masqueraded 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. Evaluations had been inconclusive. Uncomplicated cataract extraction with intraocular lens implantation was followed by persistent uveitis and glaucoma. Pars plana vitrectomy, RE, disclosed no evidence of infection or malignancy.

When the patient presented to us in June, 1995, he had moderate panuveitis, RE, with an intraocular pressure of 28. Non-invasive uveitic laboratory evaluations were initiated. Patient was treated with diflunisal, 500 mg. orally twice daily, and prednisolone, diclofenac, apraclonidine, and timolol, topically, RE. Two months later, the inflammation persisted. All evaluations were negative. Methotrexate therapy, 7.5 mg orally once weekly was initiated. During the next several months, the patient continued to have persistent iridocyclitis with elevated pressure.

In April, 1996, we performed pars plana vitrectomy in the right eye. During the surgery, we detected a brownish lesion in the ciliary body, at the 12 o'clock position, suggestive of a melanoma. Post-operatively, ultrasonography confirmed a lesion of moderate echo adherent to posterior iris, within the ciliary sulcus. A consultation was obtained from retina consultants. The recommendation was observation of the lesion. During the next six months, the patient continued to have persistent uveitis and elevated intraocular pressure.

In November 1996, we performed a third vitrectomy and removal of the intraocular lens implant. Continued observation of the lesion was recommended by the retina consultants. During the next six months, the patient continued to have, although diminished, uveitis on methotrexate, diflunisal, and rimexolone drops four times daily. The intraocular pressure remained elevated in the right eye, despite therapy with topical betaxolol, dorzolamide, and apraclonidine, requiring the addition of latanoprost, which intermittently controlled the pressure.

In July, 1997, 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 (gray) 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: We have presented a case of probable ciliary body melanoma masquerading as chronic uveitis. Although there was no histopathological evidence, extensive evaluations by the retinal oncologists and the clinical response of inflammation and ocular hypertension to proton beam irradiation are highly suggestive of CB melanoma.

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. But, uveitis is rarely associated with CB melanoma. There is only one published report in the recent literature of a case of ciliary body melanoma masquerading as anterior uveitis and invading the optic nerve, leading to enucleation. The necrosis of the tumor is believed to have induced the uveitis.

There are a number of uveal melanoma management options employed by ophthalmic oncologists today, including enucleation, observation, local surgical tumor resection, photocoagulation, and radiotherapy with either radioactive plaques or external beam irradiation with charged particles, and recently, the use of adjunctive interferon therapy. Observation of the tumor until there is a significant change, in this patient, is not atypical. But this case should serve as an additional caution. Malignancy should be considered in cases of chronic uveitis that do not respond to aggressive medical therapy. Ciliary body melanoma can present as refractory glaucoma and chronic uveitis. Direct treatment of the melanoma may be required to control the uveitis and glaucoma.

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