Case Report

Crystalline Lens Resorption Caused by Ciliary Body Melanoma

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Keywords

Uveal melanoma · Ciliary body melanoma · Cataract · Lens · Ultrasound biomicroscopy

Abstract

We report a case of a 51-year-old Caucasian female who presented with a ciliary body melanoma of the right eye leading to focal resorption of the crystalline lens and inducing posterior subcapsular cataract. She underwent successful enucleation, and histopathology demonstrated a ciliary body melanoma with a predominance of epithelioid cells and focal scleral extension. Genetic testing revealed a heterozygous, pathogenic mutation of BAP1 (c.1717delC, p.Leu573fs). Crystalline lens resorption is a rare but potentially important finding in ciliary body melanoma, as early detection of malignancy can be lifesaving.

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Case Report

A 51-year-old Caucasian female presented for blurry vision in her right eye. She had a past medical history significant for cutaneous basal cell carcinoma and uterine fibroids. She reported several months of blurry vision, as well as dull pain of the right eye with associated periorbital headache. Her family history was notable for mesothelioma and cutaneous melanoma in her mother, ovarian cancer in her eldest sister and cutaneous melanoma and basal cell carcinoma in her second-eldest sister.

On examination, her visual acuity was count fingers at three feet in the right eye (OD) and 20/25 in the left eye (OS). Intraocular pressure was 12 and 15 mm Hg for the OD and OS, respectively. Her pupils were equally round and reactive without a relative afferent pupillary defect. Motility was full in both eyes. Nasal visual field loss was noted to confrontation OD. On

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Fig. 1. **a** Slit-lamp photograph demonstrating a ciliary body melanoma associated with focal lens resorption and subcapsular cataract. **b** Ultrasound biomicroscopy demonstrating a solid tumor involving the ciliary body and impinging on the crystalline lens.

slit-lamp examination, an inferotemporal retrolental mass was observed, with impingement upon and focal resorption of the crystalline lens, as well as 2+ nuclear sclerotic and 2+ central posterior subcapsular cataract OD (Fig. 1). Her fundus exam was otherwise unremarkable in both eyes. B-scan ultrasonography demonstrated a round, solid lesion of the ciliary body with a thickness of 8 mm and low internal reflectivity.

The patient elected for enucleation rather than plaque brachytherapy. Histopathology demonstrated a ciliary body melanoma, stage pT2b, with a thickness of 8 mm and a diameter of 11 mm and with a predominance of epithelioid cells. Focal intra-scleral was detected without extraocular extension. Genetic testing revealed a heterozygous, pathogenic mutation in BAP1 (c.1717delC, p.Leu573fs). Two years after enucleation she was doing well with no evidence of metastasis.

Discussion

Ciliary body melanoma represents 6% of uveal melanomas and often remains hidden behind the iris, resulting in delayed presentation until the lesion has grown to a large size [1]. This is significant, as tumor size is strongly correlated with eventual tumor metastasis [1].

Among uveal melanoma, ciliary body melanoma is particularly likely to be associated with high-risk cytogenetics, while the ciliary body provides rich microvasculature which may encourage tumor growth and metastasis [2].

Mutations of the tumor suppressor BAP1 are associated with ciliary body melanoma, mesothelioma, renal cell carcinoma, and other malignancies. Amongst uveal melanomas, BAP1 mutation is associated with a higher likelihood of metastasis, ciliary body involvement, and larger tumor size [3]. The particular mutation identified in this patient (ClinVar ID: 221278) has been previously described in families with a history of malignancies and is predicted to cause a premature stop codon that truncates the nuclear localization signal of BAP1, resulting in loss of protein function [4]. While prior reports have described an increased metastatic risk in uveal melanomas harboring BAP1 mutations, there was not an increased risk of local invasion in these tumors [3].

Secondary cataract associated with ciliary body melanoma is not uncommon and may be an important early finding suggestive of an occult tumor [5]. Crystalline lens indentation has been reported to occur with ring and non-ring ciliary body melanoma [6]. However, crystalline lens resorption is a rare finding with non-ring ciliary body melanoma. Despite this rarity, it is important to identify it on examination, as early detection of occult ciliary body melanoma may be lifesaving.

Conclusion

We report a rare case of a patient with ciliary body melanoma leading to focal resorption of the crystalline lens and posterior subcapsular cataract.

Statement of Ethics

Written consent for publication (including images) was obtained from the patient described in this report.

Disclosure Statement

The authors have no conflicts of interest to declare.

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Author Contributions

W.M.C. prepared the manuscript. M.R.D. prepared the figure and assisted in editing the manuscript. M.I.S. cared for this patient, edited this manuscript and gave the final approval for publication.

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