

# Clinical image: Rare photographic documentation of uveal melanoma progression

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## ABSTRACT

**Purpose:** To present clinical images of a patient with neovascular glaucoma and hyper mature cataract masking orbital extension of a uveal melanoma. **Observations:** A 67-year-old female was referred for neovascular glaucoma and found to have an intraocular tumor with massive orbital extension. She refused surgery and returned one year later with progression of the tumor with metastases and expired seven months later. **Conclusions and importance:** Unexplained glaucoma and cataract should be investigated for harboring underlying intraocular tumors to prevent delays in diagnosis. Providers also should obtain greater understanding of psychosocial and socioeconomic barriers to healthcare.

## 1. Case report

A 67-year-old female was referred for surgical management of neovascular glaucoma with a 2-year history of right eye vision loss. Examination revealed count fingers vision, iris neovascularization, hyper mature cataract (Fig. 1A), an intraocular pressure of 63 mmHg, and 4 mm of proptosis (Fig. 1B). The finding of progressive proptosis was overlooked by several outside providers previously. Magnetic resonance imaging (Fig. 2) was performed and demonstrated an orbital mass involving the posterior globe and optic nerve. Despite discussing the possibility of melanoma, the patient refused surgery and any type of systemic workup due to socioeconomic reasons and fear. One year later the patient returned due to discomfort from her significant proptosis (Fig. 3). She underwent palliative exenteration. Histopathology confirmed uveal melanoma (Fig. 4) with systemic workup revealing pulmonary and liver metastases. She expired within seven months.

## 2. Discussion

Although the majority of uveal melanomas are identified when confined within the globe, 5.8% (123/2135) can be found with extrascleral extension.<sup>1</sup> Massive orbital extension of uveal melanoma is extremely rare: of the 123 patients, 5 (0.2%) had advanced orbital extension at the time of diagnosis.<sup>1</sup> Unrecognized intraocular tumors continue to be missed by clinicians.<sup>1,2</sup> Although intraocular tumors are an established cause of glaucoma, cataract, and retinal detachment,

inadvertent surgery continues to be done on eyes with unrecognized intraocular tumors which can lead to extrascleral extension and orbital spread.<sup>1,2</sup> This emphasizes that intraocular melanoma continues to be undetected and the delay in diagnosis can lead to dire consequences. Prognosis of uveal melanoma with extrascleral extension is poor and correlated with metastatic death.<sup>1,3</sup> Exenteration, chemotherapy, and immunotherapy are ultimately often palliative.<sup>1</sup> The medical literature has shown psychosocial factors such as barriers to healthcare, financial resources, feelings of denial, fear and anxiety can contribute to delays in seeking care, however, the exact factors have not been elucidated due to a lack of reliable tools to quantify behavior. Providers and further studies should attempt to understand socioeconomic and psychosocial barriers to prevent malignancies such as uveal melanoma from reaching a poor prognostic stage.

## 3. Conclusion

Uveal melanoma is the most common primary intraocular malignancy in adults and patients can be asymptomatic for a long period. Uveal melanoma can be missed or misdiagnosed.<sup>1-3</sup> These clinical images serve as a reminder to ophthalmologists to always consider an intraocular tumor on the differential diagnosis of unexplained glaucoma and cataract. When there is a poor fundoscopic view, imaging modalities (ultrasound, magnetic resonance imaging, computed tomography) should be considered to prevent extraocular extension by inadvertent surgery or a significantly delayed or missed diagnosis of an occult tumor.

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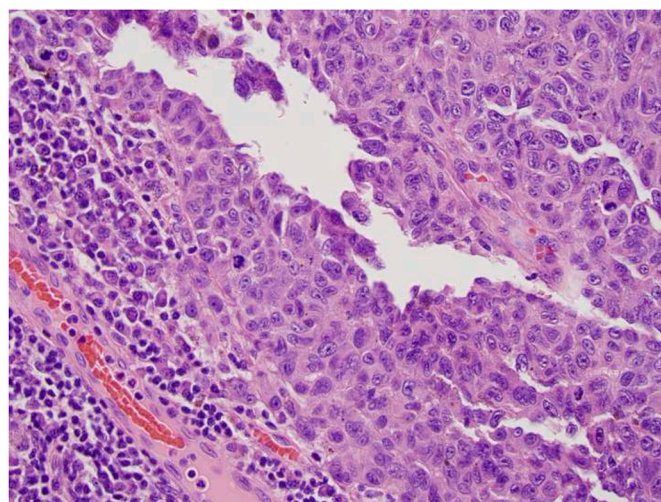
**Fig. 1A.** Slit lamp photograph demonstrating dense cataract (red star) and iris neovascularization (red arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)



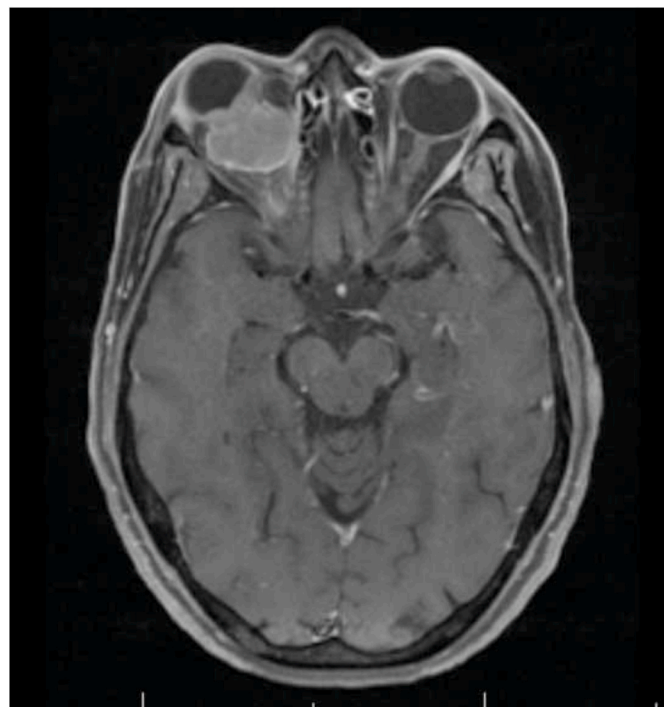
**Fig. 3.** External photograph of the patient 1 year later with extreme orbital progression.



**Fig. 1B.** External photograph in ant's eye view to demonstrate right eye proptosis.



**Fig. 4.** Epithelioid cells (200x, hematoxylin and eosin) with abundant cytoplasm and numerous mitotic figures.



**Fig. 2.** T1-weighted axial fat-saturated post-gadolinium magnetic resonance image of a homogenous, enhancing mass involving the right globe and orbit.

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**Conflicts of interest**

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### Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

### Research ethics

We further confirm that any aspect of the work covered in this manuscript that has involved human patients has been conducted with the ethical approval of all relevant bodies and that such approvals are acknowledged within the manuscript.

IRB approval was obtained (required for studies and series of 3 or more cases)N/A.

Written consent to publish potentially identifying information, such as details or the case and photographs, was obtained from the patient(s) or their legal guardian(s).

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1. Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; AND
2. Drafting the work or revising it critically for important intellectual content; AND
3. Final approval of the version to be published; AND

4. Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

### Contact with the editorial office

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