

Introduction

- Uveal melanomas are the most common primary intraocular malignancy in adults.¹
- Uveal melanoma with perineural spread along the optic nerve to the optic chiasm is exceedingly rare, with only a handful of cases documented in the literature.
- Here, we report a case of a patient presenting with monocular vision loss, found to have a retinal detachment and ultimately a locally advanced uveal melanoma that extended along the optic nerve into the optic chiasm to cause compression on the contralateral optic nerve, that was treated successfully with surgery and postoperative immunotherapy.

Case presentation

- 65-year-old female presented with progressive right-sided monocular vision loss over the past year.
- On examination, she was found to have a right-sided retinal detachment with no serviceable vision in that eye.
- She then developed left temporal visual field loss, which led to MRI of the brain showing an avidly enhancing, 3.3 x 2.9 x 2.2 cm right intraconal mass extending along the optic nerve to the orbital apex and optic chiasm, growing superiorly with an approximately 2.8 x 1.9 x 1.5 cm suprasellar component (Figure 1A, B).
- She was referred to our emergency department for multidisciplinary care.
- The patient was admitted to the hospital and underwent a right orbitotomy for biopsy by ophthalmology, which was unrevealing.
- The decision was made for a right modified orbitozygomatic craniotomy for resection of the suprasellar disease to preserve her left-sided vision and provide a tissue diagnosis for potential targeted therapeutics.

Procedure

- Modified OZ approach was performed to gain access to the bilateral optic nerves
- Intrafascial fat pad dissection was performed to avoid injury to the temporal branch of the facial nerve
- The dura was opened and the distal sylvian fissure dissected to allow the frontal and temporal lobes to relax
- Melanotic tumor was seen exiting from the ipsilateral optic nerve, filling the suprasellar space, and compressing the contralateral optic nerve
- Tumor was debulked and an intradural anterior clinoidectomy was performed
- Once the tumor was debulked and the contralateral optic nerve was fully decompressed, the orbit was opened and intraorbital disease was removed.
- Surgery was tolerated well and a subtotal resection was achieved due to tumor cells investing into the optic chiasm.
- The patient is being treated with temozolomide and bevacizumab, which she tolerates well, and is alive with stable disease at 1 year postoperative.

Discussion

- The mechanisms of optic nerve invasion of uveal melanomas is unclear, with the primary hypothesis being that elevated intraocular pressure and proximity provide a mechanical, rather than a molecular stimulus for invasion.²
- Two key next generation sequencing findings—GNAQ p.Q209P and SF3B1 p.H662R—both carry favorable prognostic implications compared to more aggressive molecular subtypes.³
- Recent advances in immunotherapies such as Tebentafusp show a survival benefit for patients with metastatic uveal melanoma, however, the patient in the present case was ineligible due to being HLA-A*02:01-negative.⁴
- A one-piece orbitozygomatic approach was chosen to access the suprasellar cistern while minimizing frontal lobe retraction.

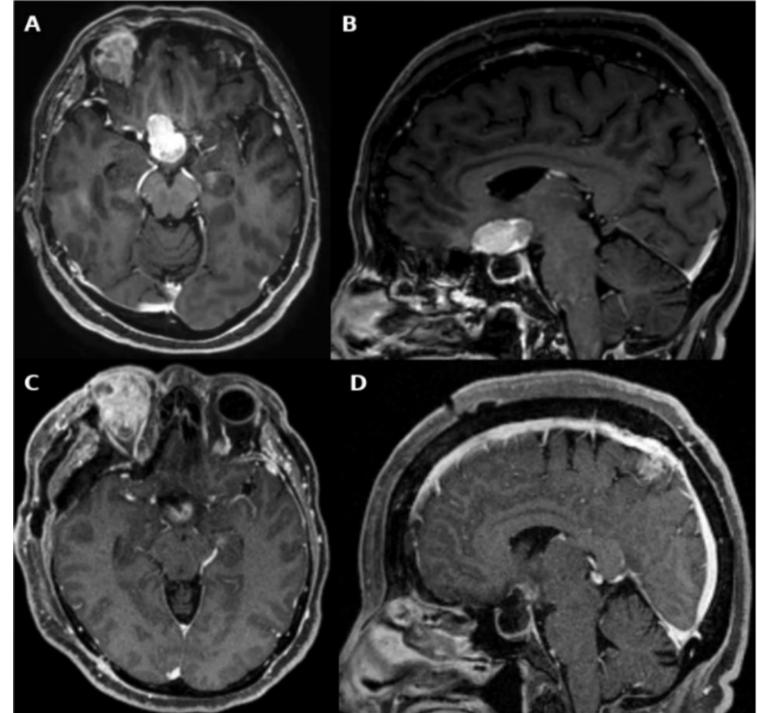


Figure 1. Preoperative axial (A) and sagittal (B) T1-weighted post-contrast magnetic resonance imaging demonstrating an enhancing right orbital intraconal mass with extension through the orbital apex along the right optic nerve to the optic chiasm extending superiorly to the suprasellar region. Postoperative axial (C) and sagittal (D) post-contrast magnetic resonance imaging shows interval resection of the enhancing suprasellar mass.

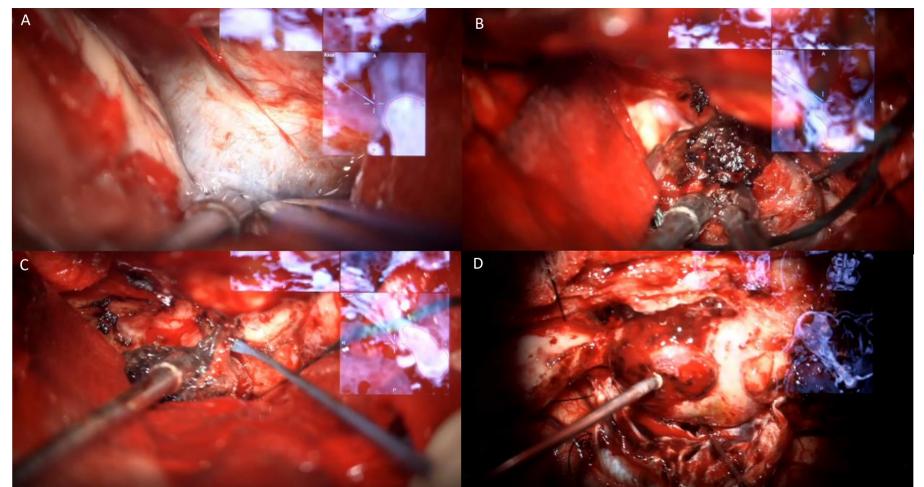


Figure 2. Melanotic tumor in the suprasellar space (A, B) and invasion of the right optic nerve (C). Intraconal disease burden (D) was revealed upon reflection of the dura flap and opening of the periorbita.

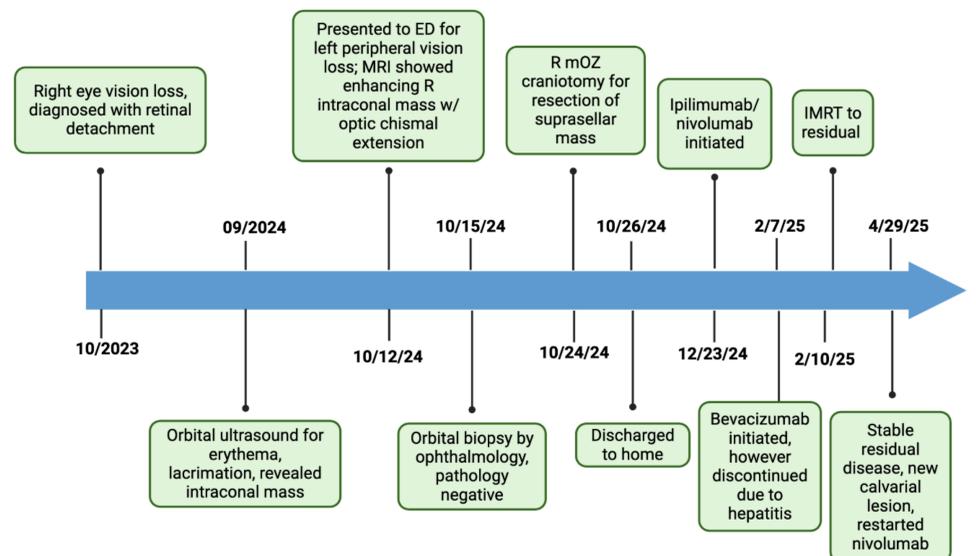


Figure 3. Patient-care timeline

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