

Recurrent Skull Base Adenoid Cystic Carcinoma Resection

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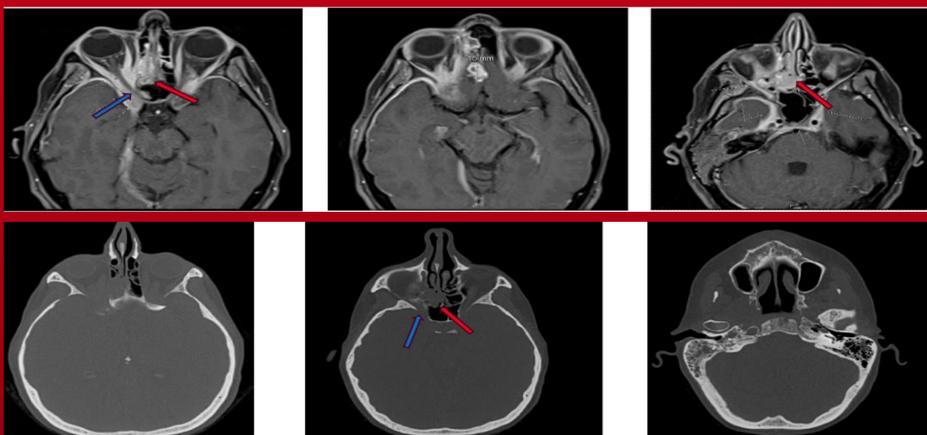
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Patient Presentation and Neurologic Exam

- 41-year-old woman with recurrent adenoid cystic carcinoma (ACC)(Grade I/III) s/p biopsy
- Had received prior RT (5940 cGy in 33 fractions, 2018)
- Patient presents with acute right-sided vision loss and diplopia
- Ophthalmic findings: R pupil nonreactive, R eye light perception only, EOM grossly intact
- Cranial nerves: V1-V3 decreased sensation on R, hearing preserved, facial symmetry, tongue midline, accessory nerve intact
- Motor/sensory exam: Normal strength, reflexes, coordination

Preoperative Neuro-imaging Findings

- MRI: T1 post-contrast axial MRI slices are shown, depicting a 3 cm diameter focus of infiltrative mass invading the posterior right ethmoid sinus, orbital apex involving extraocular muscles, and annulus of zinn with encasement of the optic nerve. Additional extensive enhancement is seen along the cavernous sinus, jugular foramen, with perineural spread along the right trigeminal nerve, and dura overlying the temporal lobe. Enhancement extends inferiorly into the suboccipital musculature.
- CT: Axial CT demonstrates the expansile/lytic nature of the lesion focused in the right posterior ethmoid & orbital apex with optic canal and superior orbital fissure and erosion, lytic destruction of the right clinoid process and clivus.



Patient Positioning

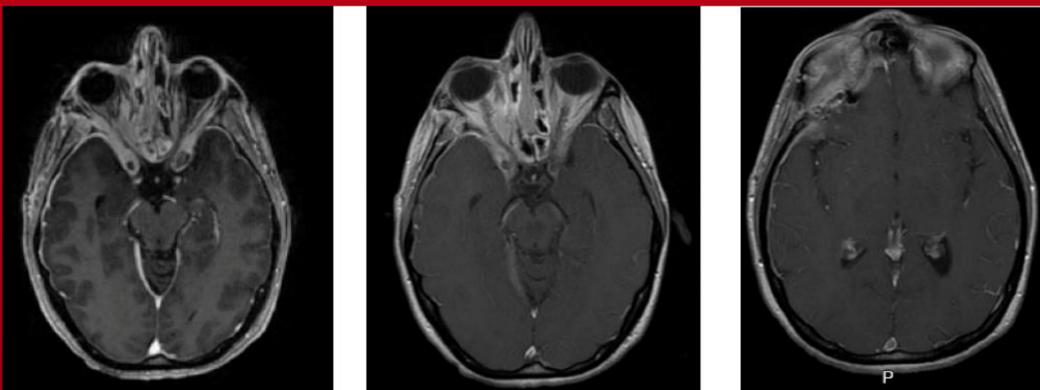


Key Surgical Steps

1. Endoscopic sinus surgery (maxillary, ethmoid, sphenoid, frontal)
2. Medial orbital wall decompression
3. Right optic nerve decompression
4. Extradural approach to medial optic canal & orbital apex
5. Bone removal with drill/curette
6. Transorbital exposure via upper eyelid crease incision
7. Lateral orbitotomy performed to access orbital apex
8. Lamina papyracea removed for medial decompression
9. Annulus of Zinn opened, extraocular muscles mobilized
10. Optic nerve sheath fenestrated for decompression
11. Hemostasis achieved, orbit reconstructed with titanium mesh
12. Orbitozygomatic craniotomy
13. Subfrontal dissection to orbital apex & anterior skull base
14. Optic nerve decompression: falciform ligament released, nerve freed
15. Tumor removed piecemeal, hemostasis with Flo-Seal/Surgicel
16. Reconstruction with pericranial flap + Duramatrix

Post-operative Neuro-imaging Findings

- Decompression achieved at orbital apex and surrounding optic nerve, cytoreduction in the regions of surgery in the ethmoid sinuses and clinoidal region with complete removal of cribriform region tumor
- Stable residual disease
- No acute complications



Patient Outcome

- Discharged on POD#5, no CSF leak observed
- Endorses visual improvement but with continued right ptosis
- No new cranial neuropathies
- KPS 80
- Has been started on avelumab/axitinib, tolerating systemic therapy, no dose-limiting toxicities
- Restaging MRI planned

Acknowledgments

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