Cranio-Orbital Approach for Single-Stage En Bloc Resection of Optic Nerve Glioma: Technical Note

Vance L. Fredrickson, MD
Guilherme J. Agnoletto, MD
Todd C. Hollon, MD
Bornali Kundu, MD, PhD
Vance R. Mortimer, AS
William T. Couldwell, MD, PhD

Department of Neurosurgery, Clinical Neurosciences Center, University of Utah, Salt Lake City, Utah, USA

Correspondence:
William T. Couldwell, MD, PhD,
Department of Neurosurgery,
Clinical Neurosciences Center,
University of Utah School of Medicine,
175 N Medical Dr East,
Salt Lake City, UT 84132, USA.
Email: neuropub@hsc.utah.edu

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BACKGROUND AND IMPORTANCE: There is no consensus on the optimal surgical approach for managing optic nerve gliomas. For solely intraorbital tumors, a single-stage lateral orbitotomy approach for resection may be performed, but when the nerve within the optic canal is affected, two-stage cranial and orbital approaches are often used. The authors describe their technique to safely achieve aggressive nerve resection to minimize the probability of recurrence that might affect the optic tracts, optic chiasm, and contralateral optic nerve.

CLINICAL PRESENTATION: A 28-yr-old woman presented with painless progressive vision loss, resulting in blindness. The second of 2 transorbital biopsies was diagnostic and consistent with low-grade glioma. The lesion continued to grow on serial imaging. The patient was offered a globe-sparing operative approach, with aggressive resection of the lesion to minimize the probability of tumor recurrence, which could possibly affect vision in her contralateral eye. The patient did well postoperatively, with clean tumor margins on pathological analysis and no evidence of residual on imaging. On postoperative examination, she had a mild ptosis, which was nearly resolved at her 6-wk outpatient follow-up.

CONCLUSION: This aggressive single-stage en bloc resection of an optic nerve glioma can achieve excellent tumor margins and preservation of extraocular muscle function.

KEY WORDS: Cranio-orbital, En bloc, Glioma, Optic nerve, Orbitotomy, Single-stage surgery

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Familiarity with cranio-orbital approaches is important for skull base neurosurgeons because many lesions involve both the intraorbital and intracranial compartments. Optic nerve gliomas involve the anterior visual pathway[1,2] and comprise ~1% of intracranial tumors.[1,2] Indications for resection include visual loss, disfiguring proptosis with exposure keratopathy, growth toward the chiasm, optic canal enlargement, and an intracranial portion of the tumor.[3] We illustrate the technical details of a single-stage cranio-orbital approach for en bloc optic nerve resection, which we use because tumor cells frequently extend beyond the boundaries of the lesion seen on MRI.[4] This aggressive nerve resection minimizes the probability of recurrence.

CLINICAL PRESENTATION

A 28-yr-old woman presented with painless right vision loss that had progressed to blindness over 1.5 yr. Transorbital optic nerve biopsy was consistent with low-grade glioma. On neurological examination, the patient was unable to see light with that eye. She had an afferent pupillary defect and mild ophthalmoparesis after biopsy. Funduscopic examination revealed 3+ optic disc pallor and 1+ optic disc edema. Her left eye function and remaining neurological examination were intact. She had no stigmata of neurofibromatosis. MRI revealed a contrast-enhancing right optic nerve lesion, with growth on serial imaging over 6 mo (Figure 1). She requested tumor resection with preservation of the globe. We used frontotemporal craniotomy with cranio-orbital approach for en bloc tumor removal with tumor-free margins (Figure 2). Pathological analysis was consistent with low-grade glioma. Postoperative MRI demonstrated no evidence of residual disease (Figure 3). Mild ptosis postoperatively was nearly resolved by her 6-wk follow-up.

Institutional review board approval was not required; the patient consented for treatment and to the publication of her images.

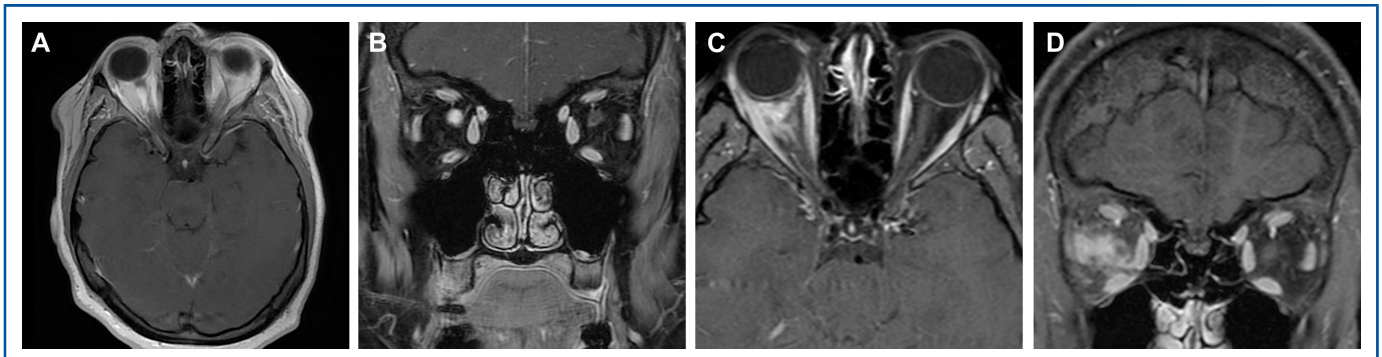

FIGURE 1. *Preoperative T1-weighted axial* **A** *and coronal* **B** *MRI of the brain with contrast enhancement demonstrating a right optic nerve lesion suggestive of an optic nerve glioma. T1-weighted axial* **C** *and coronal* **D** *MRI of the brain with contrast enhancement demonstrating growth of the right optic nerve lesion.*

Operative Technique

The operative technique is shown in the Video. The patient is positioned supine with a gel roll under the shoulder (Figure 4A). The head is elevated to optimize venous outflow from the cranial vault, pinned using the Mayfield frame (Integra NeuroSciences), and rotated to the contralateral side, so the orbital apex is perpendicular to the floor for a traditional frontotemporal craniotomy (Figure 4B and 4C).

The one-piece myocutaneous scalp flap is dissected from the calvarium and reflected anteriorly with fish hook retractors. A frontotemporal craniotomy is extended anteriorly to the orbital roof and medially to ~2 cm from the midline. The dura is bluntly dissected from the lesser sphenoid wing and drilled flat using a round burr to the meningo-orbital band at the lateral edge of the superior orbital fissure. The meningo-orbital artery is cauterized using bipolar cautery. The meningo-orbital band is then sharply transected to expose the proximal anterior clinoid. The superior portion of the orbit is removed to the level of the globe using the diamond burr and Kerrison rongeurs. The optic canal is unroofed using the diamond burr and the optic nerve identified. The lateral orbit is removed, and the anterior clinoid process and optic strut are disconnected using a diamond burr. The remaining anterior clinoid is removed using a pituitary rongeur. The periorbita is kept intact during the orbitotomy.

The dura is opened in a "T"-shaped fashion following the Sylvian fissure. The frontal lobe is sharply released from its arachnoid adhesions to the optic nerve. The ipsilateral optic nerve is sharply divided ~2 mm anterior to the optic chiasm to maintain the Wilbrand knee (Figure 5A). The falciform ligament is sharply

FIGURE 2. *Gross pathological specimen demonstrating en bloc resection of the right optic nerve. Chiasmatic end of the nerve on the right, globe side on the left.*

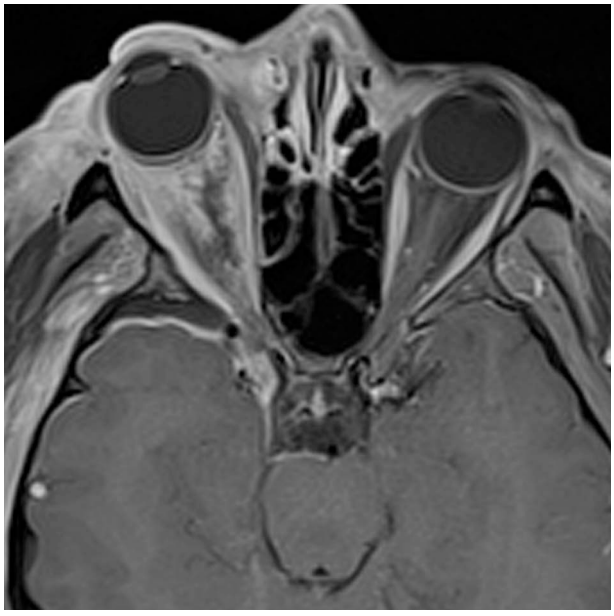

FIGURE 3. *Postoperative axial T1-weighted MRI of the brain with gadolinium enhancement demonstrating complete resection of the optic nerve lesion. Only extraocular muscles remain as shown.*

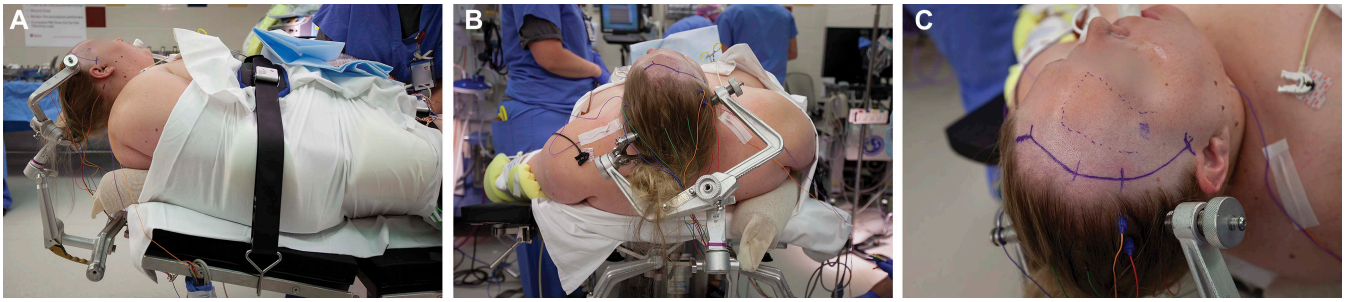

FIGURE 4. *Photographs showing patient positioning and operative planning.* **A**, *Lateral positioning with a small shoulder roll in place.* **B**, *Lateral positioning with head turned to the contralateral side so that the orbital apex is perpendicular to the floor.* **C**, *The solid line drawn on the scalp is the incision line for a frontotemporal craniotomy. The dotted line represents the estimated bony removal for the frontotemporal craniotomy with the majority of bony removal frontally for adequate access to the orbit.*

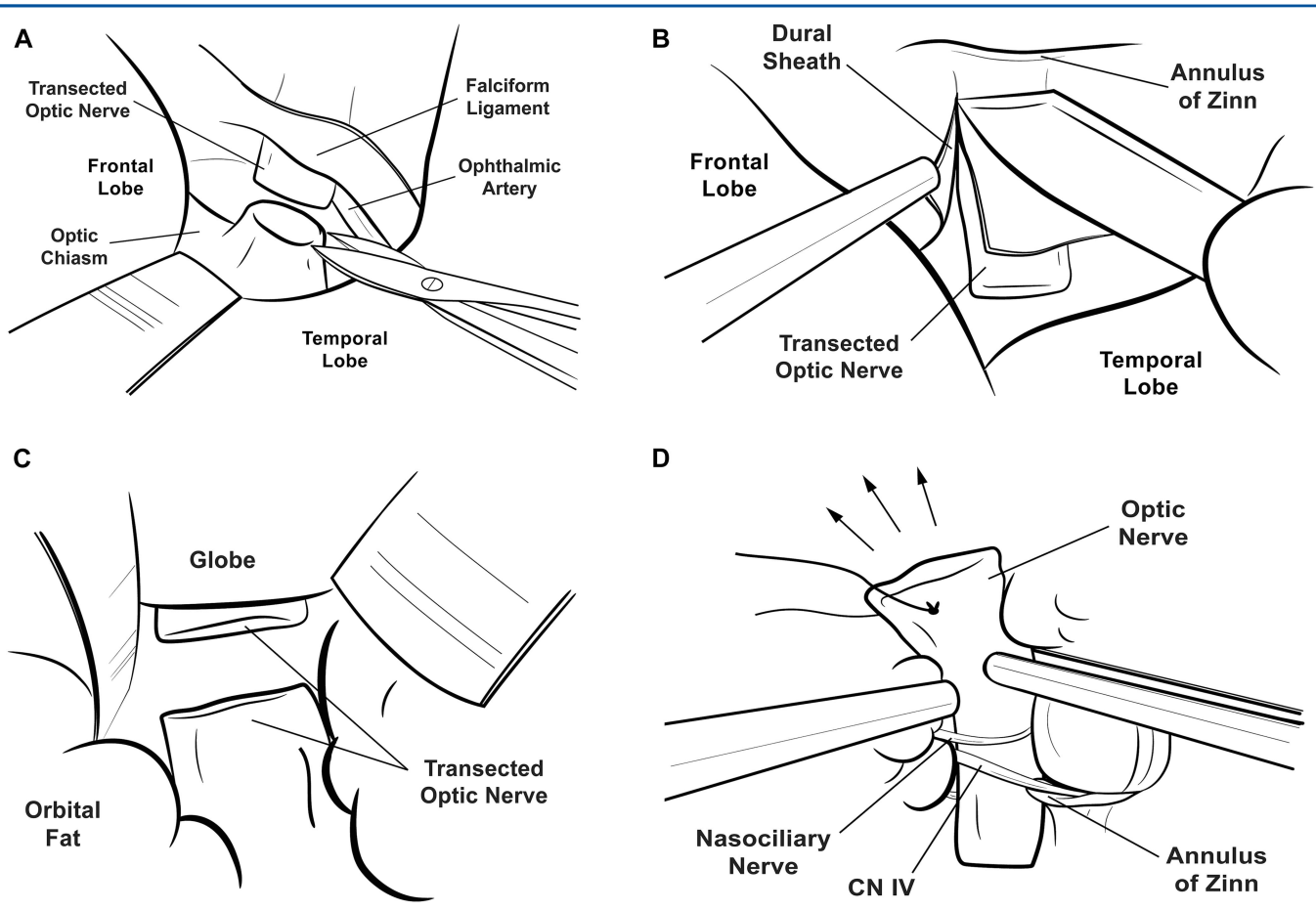

FIGURE 5. *Illustration of the technique.* **A**, *The diseased optic nerve is sharply divided approximately 2 mm anterior to the optic chiasm to avoid injury to the Wilbrand knee.* **B**, *The falciform ligament, dural sheath, and annulus of Zinn are opened medial to the optic nerve.* **C**, *The optic nerve is sharply transected at the globe after its exposure from the chiasm to the globe.* **D**, *Placement of a stitch in the optic nerve on the end excised from the globe and gentle removal of the nerve by pulling it anteriorly and superiorly after it has been freed from its investments in the orbit and annulus of Zinn.* © *Department of Neurosurgery, University of Utah.*

transected, using an 11-blade scalpel, on the medial side of the optic nerve, opening the optic dural sheath and the annulus of Zinn (Figure 5B). The periorbita is sharply opened medial to the levator palpebrae muscle and lateral to the superior oblique muscle. The frontal branch of V1 is visualized as it courses over the levator palpebrae muscle. Blunt dissection is performed medial to the levator palpebrae and superior rectus muscles and over the proximal portion of the optic nerve, progressing anteriorly. Self-retaining retractors are placed to retract the levator palpebrae and superior rectus muscles and periorbital fat. The nasociliary nerve and ophthalmic artery, which cross the optic nerve, are identified. The optic nerve is sharply transected at the globe (Figure 5C). The inferior portion of the nerve is dissected, moving toward the annulus of Zinn, where it is sharply dissected from its investments, without placing excessive traction on the nasociliary or trochlear nerves, so it can be removed en bloc (Figure 5D). The annulus of the Zinn, periorbita, and dura are reapproximated using sutures. The orbit is reconstructed using a preformed implant. A standard closure is performed.

DISCUSSION

The rarity of optic nerve gliomas and their ambiguous natural history contribute to the lack of a gold-standard treatment.[5] The solely orbital or cranial approaches used currently for optic nerve transection have notable drawbacks of limiting exposure and hindering visualization of portions of the nerve and other structures.[3,6-8] Some surgeons argue that subtotal resection or tumor debulking through an orbital approach provides good long-term results with rare progression,[9-11] but recurrence leading to chiasmal invasion, contralateral vision impairment, or even death has been reported.[7,12-16] We believe that in patients with severe vision loss or progressive tumor growth, the affected nerve should be resected entirely, so a cranio-orbital approach is the most appropriate. Although the existence of the Wilbrand knee is debated as an artifact because of optic atrophy, we transect the optic nerve ~2 mm anterior to the optic chiasm to maintain it.[17,18] In this case, the patient had optic atrophy secondary to her long-standing optic nerve glioma.

This approach differs from the frontozygomatic approach and also the combined transcranial-orbital approach described by Shriver et al.[19] Although all 3 allow access to both intracranial/intracanalicular and intraorbital aspects of the optic nerve, the combined approach is less complex and more efficient, with excellent cosmetic results, avoiding the anterior orbital exposure. A standard frontotemporal craniotomy and drilling of the orbital roof and lateral wall are performed, avoiding the need for the zygomatic portion of the craniotomy or even en bloc craniotomy of the orbital rim, roof, and lateral wall, which fundamentally increase the complexity. The exposure of the intracranial, intracanalicular, and intraorbital portions of the optic nerve and adjacent structures are extremely clear in our combined approach, allowing for preservation of the trochlear nerve and superior division of the oculomotor and other orbital nerves, all while achieving an en bloc complete resection of the tumor with a margin-free transection of the optic nerve.

An alternative to our approach is the endoscopic endonasal approach combined with a transconjunctival-medial orbitotomy.[20] This two-stage approach also allows for resection of the intraorbital optic nerve in its entirety. Although the approach requires a multidisciplinary team of surgeons, it also provides an en bloc resection of the nerve with reported reduced risk of injury to cranial nerves III, IV, VI, and V1, when compared with more traditional approaches.

CONCLUSION

We present the technical details for a single-stage cranio-orbital approach to en bloc resection of an optic nerve glioma, obviating the need for a separate simultaneous approach.

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Disclosures

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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Acknowledgments

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VIDEO. This video demonstrates the technique used to safely achieve aggressive nerve resection to minimize the probability of recurrence with potential to affect the optic tracts, optic chiasm, and contralateral optic nerve. It includes details of the case, imaging, and step-by-step technical details of the procedure. © Department of Neurosurgery, University of Utah.