

Orbitozygomatic Resection of Meningiomas of the Orbit

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Objective: To review our experience with the surgical treatment of orbital meningiomas. **Methods:** A retrospective review of all patients presenting to the author for treatment of orbital meningiomas over a 5 year period with a minimum of 1 year follow-up were included in this review. Demographic data, surgical approaches, and recurrence rates were documented. **Results:** A total of seven patients were treated for this condition. Complete surgical resection was achieved with the outlined technique in 71% of the patients, and gross tumor removal was achieved in the remaining 29% of patients. This latter group received post-operative gamma-knife treatment, and only a single patient has evidence of persistent disease that is non-progressive at 3-year follow-up. **Conclusions:** Meningioma of the orbit, whether primary (ectopic) or secondary, should be treated as at other sites with complete surgical excision if possible. Gamma knife or intensity modulated radiotherapy may be useful for residual microscopic disease. Surgical clearance is facilitated with the broad field exposure afforded by the orbitozygomatic approach outlined in the article, with particular attention directed at positive identification of key landmarks particularly in retro-orbital and cavernous sinus extension. **Key Words:** Skull base surgery, orbital reconstruction, orbital tumors, meningiomas.

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INTRODUCTION

Meningiomas are generally benign tumors that may arise from a variety of cell types, the most common of which are the meningotheelial cells found within the arachnoid layer of the meninges. They comprise 15% of all intracranial neoplasms, affecting females at a rate two to three times that of males. They are often slow growing, have expansile lesions, and commonly involve both dura and bone when they arise at the level of the cranial base. Four distinct histologic patterns are noted: angioblastic,

transitional, syncytial, and fibroblastic. This histologic diversity does not have any prognostic significance, but it may give rise to significant difficulty in differentiating meningiomas from other intracranial neoplasms such as carcinomas, hemangiopericytomas, schwannomas, and paragangliomas. Immunohistochemical reactivity to epithelial membrane antigen, vimentin, and occasionally keratin and S-100 protein and lack of reactivity to desmin, neurofilament, synaptophysin, chromogranin, and glial fibrillary acidic protein assist one in reaching a definitive diagnosis in the majority of circumstances.^{1–3} Meningiomas always require an intradural approach to gain complete access to the tumor and facilitate removal. However, dural and osseous involvement does not signify malignancy. Only the presence of distant metastatic disease is sufficient evidence of malignant meningioma.⁴

Extracranial meningiomas, comprising 2% of all meningiomas, most often represent extension of an intracranial neoplasm directly outside of the confines of the calvarium or spinal canal.⁵ These secondary meningiomas maintain a connection to the central nervous system (CNS). They may also arise from arachnoid cells of the perineural sheath of cranial nerves as they exit their foramina.⁶ In such primary meningiomas, there may not be an identifiable connection to the CNS. Extracranial orbital involvement by meningioma may occur at any site within the orbit, but it most commonly involves the orbital walls.^{7,8} Orbital meningiomas account for only 5% of all tumors of the orbit.^{9,10} Ectopic rests of arachnoid tissue within the retrobulbar fat may serve as the origin of free-lying intraorbital meningioma.¹¹ Most often, however, direct extension of intracranial meningioma into the orbit is the primary source of origin of these tumors. Often, one will note underlying reactive hyperostosis where the tumor contacts the walls of the orbit. In this article, we will review our experience with this unusual neoplasm of the orbit, outlining our approach to surgical resection and reconstruction in this patient population.

METHODS AND RESULTS

All patients with a diagnosis of orbital meningioma treated by the author from 1997 to 2002 were included in this retrospective review. A total of seven such patients were identified (Table I). All of our patients were female, with an average age of 39.8 years. Complete tumor clearance was achieved in five of the seven

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TABLE I. Patient Demographics.										
Age (years)	Sex	Prior Treatment	Presenting Signs and Symptoms	Extent of Disease at Presentation	Preoperative (postoperative) Visual Acuity	Preoperative (postoperative) Extraocular Movement	Preoperative (postoperative) Upper-Lid Function	Complete Excision (residual disease)	Postoperative Radiation/Gamma Knife	Recurrence (follow-up)
34	Female	None	Unilateral proptosis	Sphenoid wing tumor	20/20 (20/20)	Restricted laterally (normal)	Normal (normal)	Yes (none)	None	None (3 years)
49	Female	None	Unilateral proptosis, orbital pain	Sphenoid wing tumor	20/40 (20/40)	Restricted laterally (normal)	Normal (normal)	Yes (none)	None	None (1.5 years)
46	Female	Prior intracranial meningioma resection	Enlarging firm upper eyelid mass, ptosis secondary to mass effect	Tumor confined to upper eyelid with minor attachment to superior orbital rim	20/200 (20/200)	Restricted in all dimensions (no change in restriction)	Ptosis (ptosis unchanged)	Yes (none)	None	None (1.5 years)
50	Female	Prior craniotomy for meningioma on two occasions	Decrease in visual acuity, diplopia, headaches	Extensive disease of bilateral cavernous sinus, orbital apex, and subfrontal area	20/100 (20/60)	Restriction in superior gaze (no change in preoperative restriction)	Normal lid function (mild ptosis)	No (microscopic disease left at level of cavernous sinus)	Yes (gamma knife)	None (1 year)
45	Female	Prior craniotomy on three occasions and radiation (external beam)	Blindness left eye, proptosis, extracranial tumor extension through temporal fossa	Extensive tumor of left orbit, temporal fossa, subfrontal area, and cavernous sinus	Blind (blind)	Restriction in all directions (no change in restriction)	Moderate ptosis (no change in ptosis)	No (microscopic disease along optic chiasm and internal carotid artery within cavernous sinus)	Yes (gamma knife)	Yes (nonprogressive residual disease at level of cavernous sinus and optic chiasm at 3-year follow-up)
36	Female	Prior craniotomy on three occasions	Bilateral blindness, unilateral proptosis, extensive hyperostosis of orbitocranial skull base	Extensive subfrontal tumor with involvement of orbital walls, bilateral cavernous sinus and orbital chiasm, ethmoid extension	Blind (blind)	Restriction in superior and lateral movement (exentiation)	Normal (transient ptosis)	Yes	None	None (1.5 years)
55	Female	Prior infratemporal fossa approach for meningioma excision and external beam radiotherapy	Orbital pain	Radiographic evidence of tumor involving orbital apex with inferior extension into nasopharynx	20/80 (20/60)	Restriction in inferior gaze (no change in inferior gaze)	Normal (normal)	Yes	None	None (3.5 years)

patients, with microscopic residual disease remaining in two patients, one of which responded completely to gamma-knife treatment postoperatively. The one patient with persistent microscopic disease at the level of the optic chiasm presented a clinical challenge, because, although the disease process was resectable, the patient had expressed a desire preoperatively to minimize dissection that would endanger her only seeing eye. The residual microscopic disease at the level of the chiasm and the cavernous sinus in this patient was also not readily amenable to complete gamma-knife clearance either, leaving the patient with stable residual disease at this level after gamma-knife treatment.

Technique Highlights

All patients underwent an orbitozygomatic approach to their neoplasm in conjunction with a craniotomy. Typically, two distinct osseous flaps are created: frontotemporal craniotomy and superolateral orbit with malar eminence (Figs. 1 and 2). Titanium miniplates are preadapted across the frontal bar and orbitozygomatic segments to allow for restitution of the craniomaxillofacial skeleton in its proper position at the completion of the procedure. Once the craniotomy bone flap is removed, the intraorbital cuts are completed with dural protection from the superior approach. The inferomedial limit of dissection within the orbit is the inferior orbital fissure. Once the orbitozygomatic segment is removed, the temporalis muscle is reflected inferiorly, providing for broad access to the infratemporal fossa and orbital apex. In the case of meningiomas of the greater wing of the sphenoid, this bone is now readily accessible for osteotomy (Figs. 3–7). The tumor is removed from the triangular space defined by the foramen spinosum posteriorly, the squamosa of the temporal bone anterior to the temporomandibular joint laterally, and the foramen rotundum medially. The underlying dura is opened and removed to allow for clearance of the typical dural tail seen in this subset of patients.

If further exposure of the optic chiasm or cavernous sinus is required, the temporal bone inferior to the craniotomy site is drilled away. Next, the floor of the middle cranial fossa is drilled with a medium round burr to the level of the middle meningeal artery (foramen spinosum) and the foramen ovale. The internal

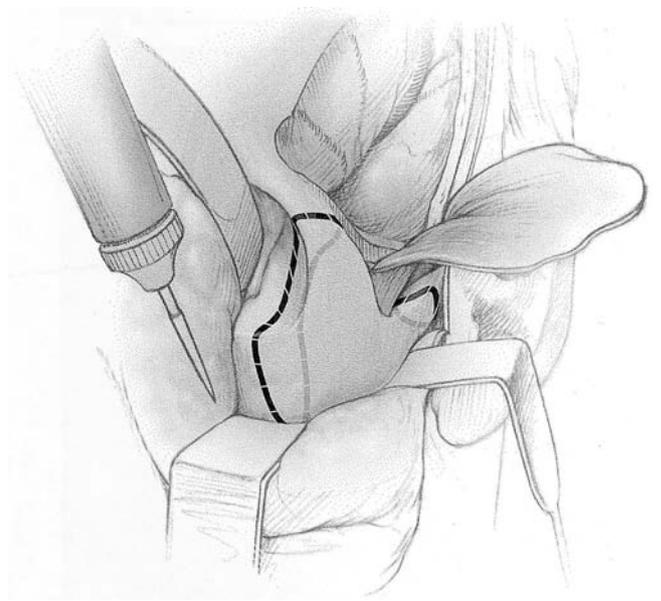


Fig. 2. Craniotomy flap has been removed, and osteotomy is being performed to allow removal of the orbitozygomatic segment.

carotid artery (ICA) is next identified in its extracranial course immediately posterior to this level (Fig. 8). This facilitates safe exposure of the cavernous sinus and orbital apex by broadening the safe exposure to this area. The optic canal may next be directly opened with a small diamond burr. In addition, the anterior clinoid is usually drilled down and removed with a rongeur. This widely opens the space defined by the oculomotor nerve inferolaterally, the clinoidal ICA inferiorly, and the optic nerve medially. This space is often involved with meningioma in the case of cavernous sinus involvement and is generally easily approached for clearance with this technique (Figs. 9 and 10). The

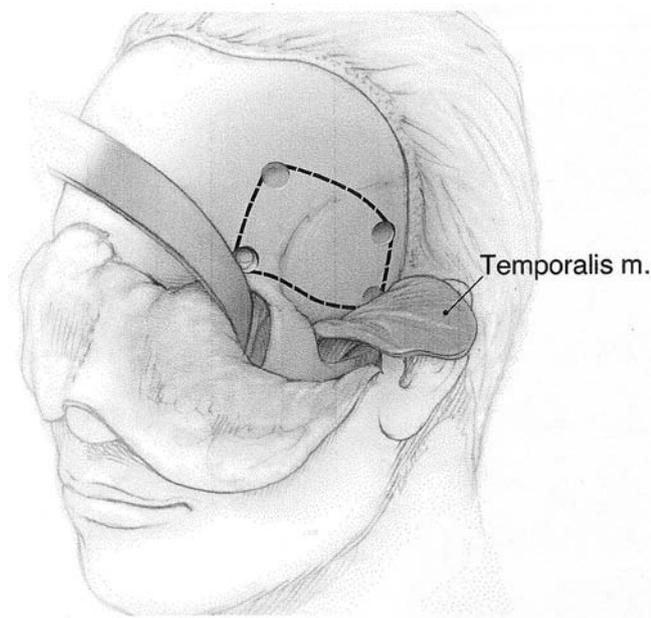


Fig. 1. Proposed frontotemporal craniotomy site and temporalis muscle reflected inferiorly.

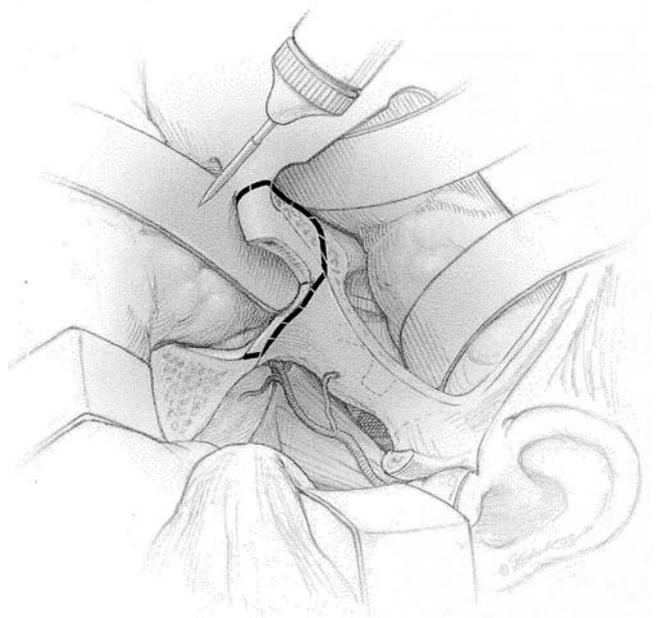


Fig. 3. Orbitozygomatic segment has been removed. Osteotomy of the greater wing of the sphenoid for meningioma involvement is being demonstrated.



Fig. 4. Basal view demonstrating unilateral left sided proptosis in patient with sphenoid wing meningioma (Figs. 4–7).

oculomotor nerve is almost invariably splayed by meningiomas of the cavernous sinus. Decompression of these tumors from a lateral approach may facilitate identification of this nerve and ultimately improve preservation. If the nerve cannot be identified from the lateral approach even after tumor decompression, then it should be traced from its subarachnoid segment into the cavernous sinus. This was necessary in two of our patients. In addition, we prefer to trace the ICA from its infratemporal course intracranially into the cavernous sinus. The tumor is readily peeled off of the ICA. The Sylvian fissure is opened widely from lateral to medial with division of temporal tip bridging veins. This will minimize frontal and temporal-lobe traction and allow for safer access to the supraclinoid ICA from the middle cerebral artery level. This is useful if the intracavernous portion of the carotid is involved by tumor.

Once the resection has been completed, the dural defects are patched with xenograft and fibrin sealant to achieve a watertight closure. Temporalis or pericranial flaps are used to fill the dead space created by tumor extirpation and augment dural patching, which is always required. The osseous segments are replaced, and any missing bone is reconstructed with autografts (calvarial bone) or alloplasts (titanium mesh-hydroxyapatite cement)^{8,9}



Fig. 5. Axial computed tomography scan demonstrating meningioma of the greater wing of the sphenoid bone. Note underlying intracranial dural tail typical of meningiomas in this area (arrow).

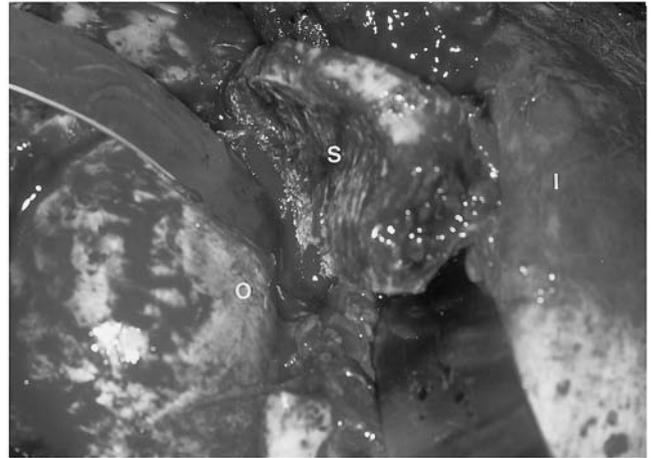


Fig. 6. Meningioma has been completely exposed. Malleable retractor is reflecting orbital contents, exposing the area of the orbital apex to assure tumor clearance. O = orbital contents; S = meningioma of greater wing of sphenoid; I = intracranial cavity.

(Figs. 11–15). Perioperative tracheotomy is used in all patients to decrease intracranial contamination and pneumocephalus in the early postoperative period.

No mortality, meningitis, prolonged pneumocephalus, per-

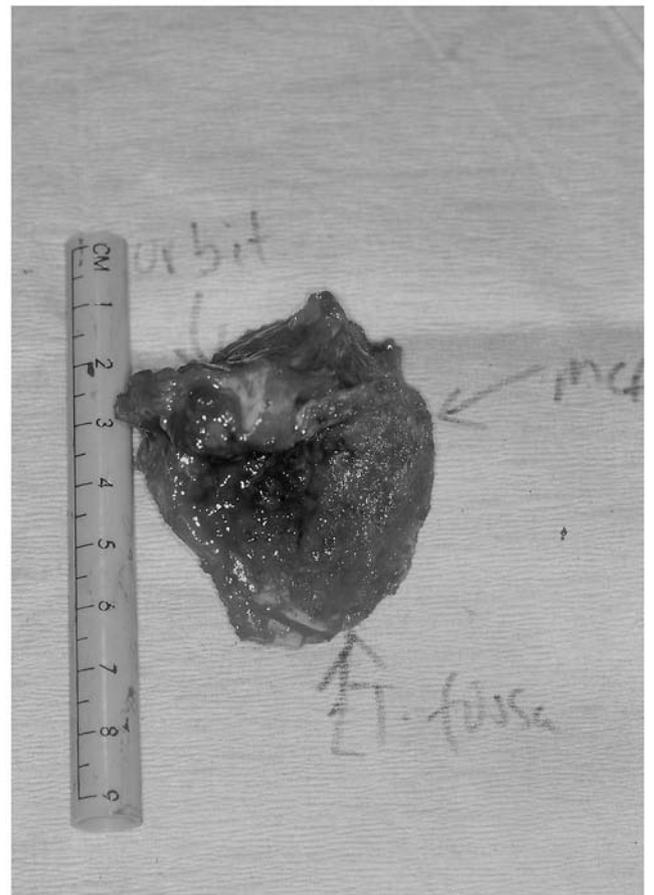


Fig. 7. Resected specimen. Orbit = intraorbital component; T. fossa = extension toward temporal fossa; mcf = extension of dural tail to middle cranial fossa.

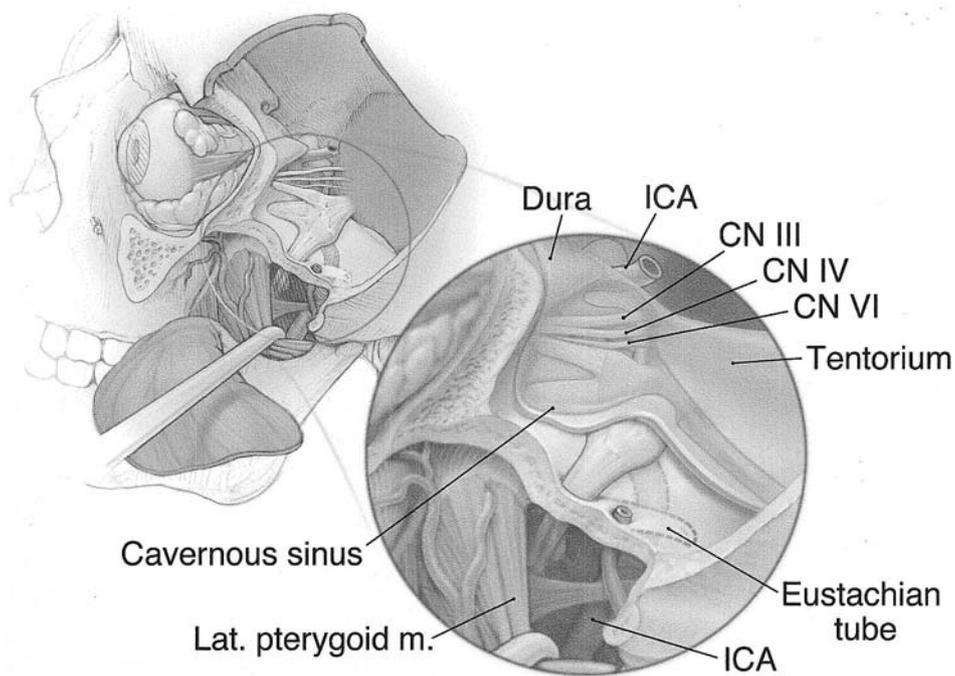


Fig. 8. Exposure achieved after removal of temporal bone inferior to craniotomy site and removal of bone along anterior floor of middle cranial fossa until level of foramen ovale and spinosum is identified. Note the middle meningeal artery crossing anterior to the internal carotid artery in its extracranial course. This affords good exposure of the intracranial and extracranial internal carotid artery, trigeminal and cavernous sinus. ICA = internal carotid artery; lat. pterygoid m. = lateral pterygoid muscle; m = middle meningeal artery; t = trigeminal ganglion; CN, cranial nerve.

sistent loss of mental functioning, or worsening of vision was noted in this subset of patients. A single patient with preoperative blindness did undergo orbital exenteration because of overt involvement of orbital contents by meningioma.

DISCUSSION

Mass effect within the orbit from a neoplasm can have clear esthetic and functional consequences secondary to herniation of orbital contents and compression at the level of the orbital apex. Furthermore, retro-orbital involvement of the cavernous sinus can have devastating neurologic sequelae if left unattended. Although meningiomas represent a benign entity, they tend to have a relentless recurring course at the level of the orbitocranial skull base if incompletely resected. Gross tumor clearance followed by localized gamma-knife treatment if there is a



Fig. 9. Circumferential involvement of intracavernous carotid arteries (arrows) by orbital apex meningioma.

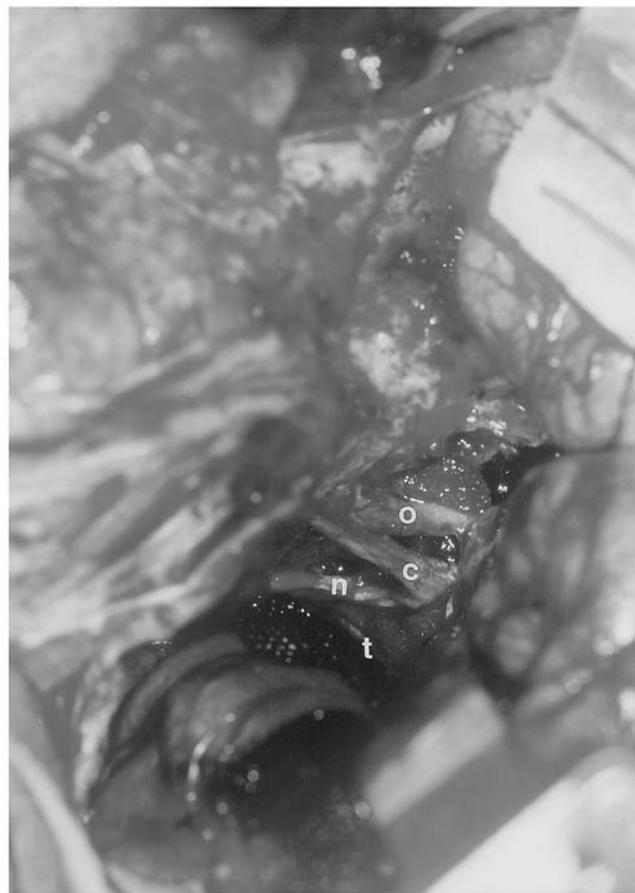


Fig. 10. Clearance of tumor from cavernous sinus. O = optic nerve; c = carotid artery; n = oculomotor nerve; t = trochlear nerve.



Fig. 11. Coronal computed tomography scan demonstrating massive recurrent meningeoma of the left orbit with extension into the temporal and infratemporal fossa (Figs. 11–15).

suspicion of residual microscopic disease may be associated with long-term disease remission or clearance.

Jane et al.¹⁰ and Al-Mefty et al.^{11–13} have made tremendous contributions with their innovative initial development of the zygomaticofrontal approach to the skull base. However, there are few reports in the literature regarding orbital meningiomas.^{14–21} Thus, optimal treatment has not been defined in any large series of patients. It seems, however, that surgical resection through broad field exposure followed by precise three-dimensional orbital wall reconstruction is most widely accepted in the treatment of this entity. Gamma knife or intensity modulated radiotherapy may be beneficial, but further study in a larger series of patients is necessary to determine its true efficacy.

The outlined technique allows for reproducible safe exposure of this neoplasm to facilitate resection. Defining the “spaces” that meningiomas tend to favor at this level of the skull base intraoperatively through positive sequential identification of the outlined key neurologic structures

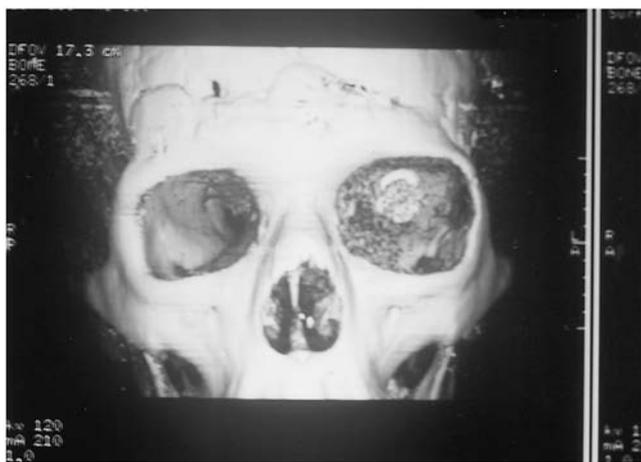


Fig. 12. Three-dimensional computed tomography scan demonstrating severity of left orbital wall destruction.



Fig. 13. Resected meningeoma. Note small cuff of intraorbital fat taken with the specimen (arrows) to assure completeness of resection in this patient with multiple recurrences.

will allow for systematic clearing of the tumor load and minimize neurologic sequelae. Meningiomas are readily dissected from nerves and the ICA as well as the cavernous sinus as long as a clean plane of dissection is identified. Occasionally, the oculomotor nerve and more often the trochlear nerve are attenuated to such a degree that preservation and complete tumor clearance may not be compatible. Nerve sacrifice or leaving of microscopic residual disease around the attenuated nerve may be required in these circumstances.

It is important to recognize that intradural exposure is required for all of these tumors unless they are truly completely ectopic within the orbit or eyelid. Dural resection in the area of involved tumor is often performed to assure clearance of disease at this level. Persistent dural disease is often associated with early multifocal recurrence.

Orbital wall reconstruction is required to maintain adequate globe position in an anteroposterior dimension and allows for extraocular muscle functioning. Alloplastic



Fig. 14. Orbital and temporal fossa defect reconstruction with titanium mesh.



Fig. 15. Postoperative three-dimensional computed tomography scan of patient in Figures 11 to 14, demonstrating adequate restoration of orbital form with alloplastic reconstruction.

and autograft reconstruction is used by the author with success. Preoperative fabrication of a computed tomography-based, three-dimensional model is invaluable in aiding the reconstructive effort because it provides the surgeon with the precise anatomy of the preoperative orbit during the surgical restoration of this structure at the completion of the procedure.

CONCLUSION

Meningiomas of the orbit represent a rare location for a rather common and generally benign intracranial neoplasm. As with meningiomas at other sites, surgery is the mainstay of treatment. More complete surgical resection is possible through the broad field orbitozygomatic skull base approach using the techniques presented in this article. Residual microscopic disease may be amenable to treatment with radiotherapy. Surgical restoration of the orbital wall anatomy is essential in maintaining postoperative form and function.

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