Sphenoorbital meningiomas: surgical limitations and lessons learned in their long-term management

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Object. Sphenoorbital meningiomas (SOMs) are complex tumors involving the sphenoid wing, orbit, and cavernous sinus, which makes their complete resection difficult or impossible. Sphenoidal hyperostosis that results in incomplete resection makes these tumors prone to high rates of recurrence with postoperative morbidity resulting in a nonfunctional globe. High-dose radiation therapy has often been described as the only treatment capable of achieving tumor control, although often at the expense of the patient’s progressive visual deterioration.

Methods. This series consisted of 25 patients who were retrospectively analyzed over a 12-year period. Visual function was evaluated pre- and postoperatively in all patients. A standardized surgical approach to a frontoetemporal craniotomy and orbitozygomatic osteotomy with intra- and extradural drilling of the optic canal and all the hyperostotic bone was performed. Orbital and cranial reconstruction was performed in all patients. The follow-up period was 6 months to 12 years (average 5 years).

The patients presented with the classic triad of SOM: proptosis (86%), visual impairment (78%), and ocular paralysis (20%). A gross-total resection was achieved in 70% of patients with surgery limited by the superior orbital fissure and the cavernous sinus. Proptosis improved in 96% of patients with 87% improvement in visual function. Ocular paralysis improved in 68%, although 20% of patients experienced a temporary ocular paresis postoperatively. There were no perioperative deaths or morbidity related to the surgical approach or reconstruction. Ninety-five percent of patients reported an improved functional orbit. There was tumor recurrence in 8% of patients; in one case recurrence was delayed for longer than 11 years.

Conclusions. Sphenoorbital meningiomas are a distinct category of tumors complicated by potentially extensive hyperostosis of the skull base. Successful resection requires extensive intra- and extradural surgery, necessitating drilling of the optic canal and an orbital osteotomy within anatomical limitations. The bone resection requires reconstruction with autograft, allografts, or alloplast for improved orbital function. All aspects of the clinical triad improved. A radical resection can be achieved with low morbidity, providing a significantly improved clinical outcome in the long-term period.

Key Words • meningioma • orbital tumor • sphenoid • orbital reconstruction

Abbreviations used in this paper: CT = computedized tomography; MR = magnetic resonance; SOF = superior orbital fissure; SOM = sphenoorbital meningioma.
We present our surgical techniques for resection and reconstruction, functional and aesthetic outcome, complications, and follow up in a consecutive series of 25 patients with SOMs.

**Clinical Material and Methods**

**Patient Population**

We analyzed our meningioma database from the period of 1991 to 2003 and found 25 consecutive patients with SOMs in whom long-term follow up was available. Our report consists of a retrospective case series review. Patients with nonhyperostotic sphenoid wing meningiomas, cavernous sinus meningiomas with secondary orbital involvement, primary optic nerve sheath meningiomas, and clinoidal meningiomas were excluded from this study. None of these patients had undergone surgery or radiation therapy before their first presentation. There were 22 women and three men in our patient population. The age range was 22 to 76 years, with an average age of 50.6 years. The intraoperative course was also noted in each patient, including the extent of surgery, duration of hospitalization, and postoperative morbidities.

**Clinical Neurological Examination**

All patients underwent comprehensive pre- and postoperative neurological examinations with detailed cranial nerve testing. Presenting symptoms and clinical signs were recorded. All patients underwent visual function assessment, including visual acuity and visual field tests.

**Radiological Evaluation**

All patients underwent both CT and MR imaging as part of the diagnostic workup; CT imaging with bone-window algorithm was useful in demonstrating the area of sphenoid hyperostosis (Fig. 1). Gadolinium-enhanced and fat suppression MR sequences were obtained primarily to delineate the intradural and intraorbital tumor portion (Fig. 2).

None of the patients in this series underwent preoperative angiography or embolization. Standardized attempts were made to determine tumor size. Measurements of maximum tumor diameter were performed by selecting the Gd-enhanced axial T1-weighted MR image, in which evidence of hyperostosis was seen and calculating the entire tumor area involved, including dura and periorbita. A conformal area was identified and a maximum diameter was measured with the same computer-based software.

**Surgical Technique and Reconstruction**

**Positioning.** The patient is positioned supine with the head and neck rotated to the opposite side by approximately 45°. Bilateral tarsorrhaphy sutures are applied and both eyes are draped into the field so as to permit comparison with the normal side during the reconstruction.

**Incision and Bone Opening.** Either a bicoronal or a large frontotemporal incision is made. Subfascial dissection is performed to elevate the scalp flap, protecting the frontal branch of the facial nerve, and exposing the superior and lateral rims of the orbit and the zygomatic arch. While elevating the temporal muscle, tumor involvement in the muscle must be carefully noted and excision must be performed when involvement is seen. Typically the area of hyperostosis is encountered early at this stage of dissection or even earlier as there can be tumor infiltration of the temporal muscle as well (Fig. 3). The entire area of thickened hyperostosis is then removed using a series of cutting burrs, which results in a craniectomy exposing the frontal and temporal dura and the periorbita. Usually a zygomatic osteo-
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During this bone drilling, the SOF should be carefully identified because it is often distorted and severely compressed. Further drilling around the SOF is performed under high magnification of the surgical microscope after completing the craniotomy, which provides access to the convexity dura that may be involved over a wide area.

Excision of the Dura and Dissection Around the Optic Nerve and SOF. The dura over the frontal and temporal lobes involved by the tumor is excised and the remainder of the deep dissection is performed with the aid of the microscope.

The foramen ovale and rotundum are identified extradurally and bone drilling is continued up to the inferior margin of the SOF. The anterior clinoid process, which is often hyperostotic, is removed. The optic canal is fully unroofed in its entire course. The optic foramen is unroofed and the optic sheath is opened to allow for a wide decompression of the optic nerve. All of the hyperostotic bone is systematically resected. Openings that have been created into the sphenoid and ethmoid sinuses during the unroofing of the optic canal must be recognized and sealed with a piece of muscle. The dura of the lateral wall of the cavernous sinus is dissected lateral to the trigeminal branches and this dura is resected if involved. No further dissection in the cavernous sinus or SOF is performed, even if there is tumor extension.

Removal of the Orbital Part of the Tumor. Involvement of the orbit is directly through the periorbita. Starting anteriorly, the periorbita is incised carefully, protecting the lacrimal gland and the lacrimal nerve, and resected in a piecemeal fashion, with careful dissection of the extraocular muscles and the anulus of Zinn. Tumor is resected up to the SOF and the anulus of Zinn. If there is tumor within these areas it is not aggressively pursued.

Reconstruction. The dura is repaired with a free graft by using an acellular dermis patch. The periorbita is not reconstructed. The orbitozygomatic bone segment is reattached with titanium miniplates and screws. The lateral wall of the orbit is reconstructed using a split cranial bone graft, which is cut to size, attached to the orbital rim, and secured with titanium miniplates. The orbital skeletal reconstruction was commonly performed through the use of an alloplast (Medpor; Porex Surgical, Inc., Newnan, GA), which allows for a precise orbital skeletal contouring without interfering with postoperative imaging. This is secured into place with titanium miniplates. The final position of the plates is adjusted by observing the symmetry of both orbits.

The convexity bone defect is covered with a titanium mesh cranioplasty. If there is a large cavity between the orbital and dural reconstruction and the cranioplasty, autologous fat graft harvested from the abdomen is used to fill this up and prevent a postoperative pseudomeningocele.

The temporal muscle is resuspended through small drill holes in the calvaria to the titanium mesh and then reapproximated; the wound is closed in anatomical layers.

Postoperative Imaging and Follow Up

Postoperative orbital swelling and chemosis is common. Ice packs and lubricating drops are routinely used. All patients underwent initial head CT scanning on postoperative Day 1 to assess any postsurgical hematoma or pneumo-
The clinical triad of SOM was observed in our patients. Visual Function and the SOM Clinical Triad

Results

Visual Function and the SOM Clinical Triad

The most common presenting clinical symptoms in patients were generalized headaches and focal retroorbital pain. Patients with SOMs typically present with a clinical triad composed of proptosis, visual field defects, and ocular paresis. The clinical triad of SOM was observed in our patient series: proptosis in 22 patients, visual impairment (acuity change) in 20 patients, and ocular paresis in five. The ocular paresis is more a restriction than a true nerve palsy. Features associated with sphenoorbital hyperostosis such as optic neuropathy (five patients), inferior arcuate field loss (three patients), and a constricted field with an enlarged scotoma (two patients) were found in the diagnostic workup. These tests were not performed in all patients preoperatively.

The presenting clinical triad of symptoms associated with SOM were improved in all of our patients after surgery. Proptosis was corrected in 24 patients. Visual acuity remained the same in 18 patients and improved in eight patients. In no patient was there an observed worsening of visual acuity. Among the five patients presenting with ocular paresis, there was improvement in three patients immediately postoperatively. This degree of ocular paresis continued to improve during the follow-up period. The other two patients had an improved ocular paresis by 9 months. There was also a significant improvement in visual field defects. Among the nine patients in whom formal visual field testing was performed preoperatively, eight experienced an improvement in their postoperative visual field deficit; in one patient there was no change.

Tumor Resection and Pathological Composition

Preoperative CT and MR imaging were used to aid in diagnosing the tumor and planning the resection. The primary feature in all cases was the presence of extensive hyperostosis, as demonstrated on both modalities of radiographic imaging. On CT scanning, characteristic patterns that were found that helped in the diagnosis of SOM included a periosteal pattern of hyperostosis, inward bulging of the vault lesion, surface irregularity of the hyperostotic bone, and subtle intracranial changes. In addition to hyperostotic thickening, bone remodeling of the orbital roof and anterior sphenoid wing were indicative of tumor infiltration. In all patients in our series, Gd-enhanced MR sequences demonstrated SOM infiltration of the dura. Infiltration varied from a thin line of enhancement on the dura to an actual tumor mass toward the brain and the orbit.

Although gross-total resection of all tumor components was achieved in 18 patients, complete resection was found to be limited by areas of anatomical involvement (Table 1). Among the seven patients with residual tumor, there was tumor remaining in the cavernous sinus and the SOF (Fig. 4). Tumor involvement of the infratemporal fossa, sphenoid sinus, or the ethmoid sinus did not limit a complete resection. In all 25 patients the areas of bone hyperostosis were completely removed. Postoperative Gd-enhanced MR images and CT scans were used to verify these results. All tumors in our series, based on the World Health Organization classification, were low-grade (Grade I) meningiomas, mostly meningotheial. There were no atypical (Grade II) or malignant meningiomas in our patient population. Hyperostotic specimens submitted consisted of true pathological tumor invasion in the bone in all cases.

Orbital Reconstruction

The method of orbital skeletal reconstruction used evolved during the time period of this study. Initially, the split calvaria technique was used for the cranioplasty and the orbital component. This was performed in five patients. This method proved to be time consuming and tedious. Over time titanium mesh was used, but this material proved to interfere with postoperative imaging. Titanium mesh for the orbit was used in 11 patients, and almost all the cranioplasties for frontotemporal sphenoid bone defects were reconstructed with this material. More recently a nonmetallic orbital prosthesis consisting of porous high-density polyethylene has been used. This material was placed in nine patients. Proptosis was corrected in 24 of 25 patients. One patient had continued although decreased proptosis due to residual tumor. Postoperatively, there were no cases of pulsatile enophthalmos or hypoglobus (vertical asymmetry of the orbits) recorded.

Morbidity and Mortality

None of our patients died, and there was no morbidity directly related to the surgical approach used or to the orbito- and/or zygomatic osteotomies (such as temporomandibular joint dysfunction and trismus). Temporary morbidity was mostly related to cranial nerve impairment in involved anatomical areas. There was new third nerve paresis in 17 patients postoperatively; this was due to drilling of the hyperostotic lesser sphenoid wing and anterior clinoid process. A hyperpathic trigeminal sensation was noted in four patients, mostly in the V1 and V2 distribution. A subgaleal cerebrospinal fluid collection was observed in three patients, although this number was reduced completely through the routine use of meticulous duraplasty and fat graft. Permanent morbidities in our series consisted of temporal hollowing of moderate grade in two patients, a permanent third
nerve injury in one patient, and a permanent fourth nerve injury in one patient.

Tumor Recurrence and Neurological Outcome in Long-Term Follow Up

During our long-term follow-up period, which averaged 5 years (range 6 months–12 years), there were two patients who experienced tumor recurrence. One patient had tumor recurrence 11 years postoperatively with recurrence from the SOF. This patient underwent repeated operation and new reconstruction; postoperative fractionated radiotherapy to a total of 5.4 Gy was administered. In the other patient tumor recurred after 1 year from direct extension from the posterior orbit and the SOF. This patient underwent a second operation and new reconstruction and is scheduled to undergo radiotherapy. Despite repeated operations, these patients harbor residual tumor in the posterior orbit that is stable. Radical resection was not performed so as to preserve ocular function. Among all other patients, whether they underwent gross-total resection or a limited resection, there was no observed tumor regrowth at their last 6-month follow-up examination. In the four patients with residual tumor in the cavernous sinus and in the four patients with residual tumor in the SOF there has been no growth of tumor during an average 5-year follow-up period.

Patients with improved visual function remained stable in the long term. Visual acuity remained the same in 18 patients (72%) and improved in seven (28%). In no patient was there an observed worsening of visual acuity. Visual acuity and visual field defects continued to improve in the long term; an additional eight patients (32%) had improved results on visual examination, recorded on their latest follow-up visit. Proptosis, once corrected postoperatively, also remained stable in the long term. Based on a subjective rating scale conducted at the 6th postoperative month visit, 23 patients reported an overall improvement in their cosmesis.

In the patients who presented with ocular paresis preoperatively, there continued to be improvement in function in the long-term period, up to an overall improvement in 68%. Among the observed cranial nerve paresis that occurred postoperatively, there was complete resolution in 30% of patients by 3 months, 60% of patients by 6 months, and in 90% of patients by 9 months. Patients with the hyperpathic trigeminal pain syndrome were controlled postoperatively with adjusted doses of either gabapentin or tegretol. In almost all of these patients high initial doses were able to be tapered over a 6- to 9-month period after surgery. There were no cases of permanent complete trigeminal impairment.

Discussion

Sphenoorbital Meningioma as a Separate Category of Tumor

Sphenoorbital meningiomas have historically been difficult to cure due to their extensive bone, orbital, and neural involvement. Because of the en plaque nature of the tumor’s morphology, they tend to grow in a carpetlike fashion with limited thickness, and form a thin layer that closely follows the contours of the inner table of the skull base, eventually encompassing a large surface area overall. The hyperostosis associated with SOM is now known to be direct tumor invasion and may explain the disproportionate amount of hyperostosis relative to the dural size of the tumor. Sphenoorbital meningiomas are not routine sphenoid wing meningiomas and clinically do not have the same presentation or produce the same set of patient symptomatology. Sphenoorbital meningiomas are tumors that have a distinct orbital component caused by direct tumor extension from the intracranial component. In the initial workup of the patients in our study, up to 25% of radiological reports were read as normal, and it was not until subsequent imaging was performed that an accurate diagnosis was achieved. Attempts at resection have resulted in a high rate of recurrence. Surgery of these tumors usually resulted in a postoperative nonfunctional globe with associated pulsating...
tile enophthalmos coupled with a significant cosmetic defect.\textsuperscript{29,31} High-dose radiation therapy was often viewed as the only treatment capable of achieving tumor control.\textsuperscript{2,24}

We report herein long-term outcome in a series of SOMs treated by primary resection and reconstruction. An effective resection first requires the recognition of the tumor entity and all of its involved anatomical boundaries.

Sphenoorbital Meningioma and the Necessity for Orbital Reconstruction

Although many authors believe there is no need for orbital reconstruction after surgery,\textsuperscript{8,12,23,24} some such as Gallard, et al.,\textsuperscript{3} have proposed reconstruction only when more than one orbital wall has been removed. Proper surgical reconstruction shares equal importance with resection in the achievement of a successful outcome. The addition of the reconstructive material further establishes known landmarks with which tumor recurrence on postoperative imaging can be detected. Attempts at surgery without proper orbital resection have resulted in meningocceles,\textsuperscript{5} diplopia, bulbar dysopia, pulsating enophthalmos, and ocu-lomotor muscle fibrosis, which then result in an ophthalmoplegia and a nonfunctional globe.

We performed skeletal orbital reconstruction in all patients in this series. The orbital reconstruction did not produce any associated morbidity. The globe has to be positioned to avoid orbital dystopia and hypertelorism.\textsuperscript{12} The resection of hyperostotic bone from the sphenoid around the tumor disrupts the structural integrity that the sphenoid wing provides to the posterior orbit and frontal and middle cranial fossae. There is disruption of the bone wall as well as the periorbita, which is why we believe orbital reconstruction is needed. The reconstruction begins with meticulous duroplasty and bone craniectomy correction. Proper contouring with auto- or allograft materials allows for a proper buttress with which to continue the cranioplasty.

Many different materials\textsuperscript{29} can be safely used for orbital reconstruction (all materials are inert and there has never been any documented reactivity between these reconstructive materials); however, all must be designed individually to recess the orbit in proper symmetry with the face and to recreate the necessary orbital volume that may have been lost due to involved periorbita and fat.

Anatomical Limitations to a Complete Resection

Based on our previous surgical experience with cavernous sinus tumors, we have purposefully limited our resection up to the cavernous sinus and the SOF. There was no cavernous sinus exploration for tumor in this series. It has been shown that meningiomas in the cavernous sinus can directly infiltrate cranial nerves and spread via direct extension along connective tissue planes\textsuperscript{23} in between these nerves. Manipulation of cranial nerves in the cavernous sinus can increase the possibility of permanent deficits, as is evident from other reports. This aspect of cavernous sinus lesions continues to make their complete tumor resection lead to permanent cranial nerve morbidities.\textsuperscript{7,9,20} Despite these areas of residual meningioma, the regrowth rate has been quite small (two patients). Other reported series, including our own experience, have shown the low regrowth rate of cavernous sinus residual meningiomas.

Improvement of the Clinical Triad

The visual function recorded preoperatively is believed to be directly related to the degree of bone hyperostosis because we did not find meningioma directly against the optic nerve or in the optic canal. The exuberant hyperostosis seen in SOM has been demonstrated by Al-Mefty and colleagues\textsuperscript{40} to be direct tumor invasion of the bone. Complete removal of all involved hyperostotic bone and decompression of the optic nerve is crucial in ensuring postoperative recovery of function. Although postoperatively visual acuity improved in only 28% of patients, 72% remained the same. Why some patients experienced improvement in their visual acuity while others did not is most probably related to the degree and period of optic nerve compression.\textsuperscript{7,12} The combined intra- and extradural drilling of the optic nerve foramen and all of its bone compression is just as important as opening the optic nerve sheath and providing for release of the optic nerve\textsuperscript{22} in its entire course intracranially to the orbit. As has been previously reported, the best prognostic indicators for optic nerve function postoperatively are the degree of deterioration preoperatively and the total extent of decompression.\textsuperscript{22}

Radiation Therapy and Tumor Recurrence

Except in the two patients with documented tumor recurrence, no patient in our series received any postoperative radiation therapy. Because the recurrence rate that we encountered was very low at 8% (two patients), we do not advocate postoperative radiation therapy for all patients with residual tumor in the cavernous sinus or the SOF. Rather than administration of postoperative radiation therapy in all patients, the best outcome is achieved with accurate early diagnosis and aggressive surgery at the outset before the tumor has entered the cavernous sinus, and a more complete resection without risk of postoperative recurrence is possible. For recurrent tumors we perform repeated surgery to resect as much tumor as possible and then begin postoperative radiation therapy. Even in patients with recurrent tumors, functional ocular outcome is possible along with control of the tumor. The type of radiation therapy and dosage depends on the size and location of the residual tumor, especially in relation to the optic apparatus. As a delayed recurrence did occur in one patient in our series (after 11 years), postoperative imaging is warranted yearly to monitor carefully the known residual tumor. The delineation of the final end points of tumor follow up is still unknown and remain a controversial issue for all benign tumors.

Conclusions

Patients with SOMs present with exuberant hyperostosis of the skull base, visual impairment, and a clinically progressive deformity. Proper treatment of these tumors involves detailed MR and CT imaging and visual evaluations. Detailed visual examinations both pre- and postoperatively are essential in determining the outcome of treatment. An effective resection requires removal of all hyperostosis, involved dura, and periorbita through a large frontotemporal craniectomy/craniotomy with an additional orbitozygomatic osteotomy. Decompression of the optic nerve is necessary along its entire length if tumor or hyperostosis extends to...
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the nerve. We do not enter the cavernous sinus or the SOF to avoid encuring permanent cranial nerve morbidities. A thorough orbital and cranial skeletal reconstruction is necessary to ensure a functional and cosmetically acceptable outcome. The long-term follow up of patients with these tumors has demonstrated that although delayed tumor recurrence can occur, the incidence is low in this population. Therefore, postoperative radiation therapy for all patients with residual tumor is not required. For tumor recurrences we recommend secondary surgery with postoperative radiation therapy.

References


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