Recurrent sphenoid-orbital meningioma

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A series of 15 patients who underwent neurosurgical procedures for recurrent sphenoid-orbital meningioma is reported. There were 11 women and four men, with a mean age of 46 years. The mean duration between the first and second operations was 46 months. Progressive proptosis without neurological deficit was the most common symptom. All tumors were large at the time of reoperation and involved the greater and lesser wings of the sphenoid bone and the orbit. Aggressive resection in all patients resulted in no deaths and only slight morbidity, with the exception of one patient who developed blindness 24 hours after surgery due to central retinal artery occlusion. Fourteen patients were improved cosmetically and one patient, treated early in the series, had persistent proptosis due to inadequate bone removal. No attempt was made to remove tumor within the cavernous sinus in patients who were neurologically normal. Although postoperative imaging demonstrated complete gross excision of tumor in nine patients, 10 underwent conventional radiation therapy for residual tumor visualized at the time of surgery in the dura of the superior orbital fissure, the cavernous sinus, or the basal optic canal.

Although this study is inconclusive and requires further long-term documentation, no recurrences have been seen to date in the follow-up period, ranging from 16 to 95 months. The following important points are discussed: 1) the failure by experienced surgeons to radically excise bone, tumor, and involved dura at the first operation; 2) the importance of early aggressive therapy, depending upon the patient's age and medical condition; 3) the almost invariable intracranial dural involvement, which at times was seen only by gadolinium-enhanced magnetic resonance imaging and not visualized on computerized tomography; 4) an illustrated stepwise surgical technique for complete resection through a small craniotomy without the need for complicated reconstruction of the orbit or temporal fossa; 5) the role of radiation therapy when removal is incomplete or deemed hazardous because of cavernous sinus involvement; and 6) the excellent cosmetic results possible with minimal morbidity and no mortality.

KEY WORDS • sphenoid wing • orbit • meningioma • surgical treatment • tumor recurrence

The vagaries of diagnosis and treatment of sphenoid-orbital meningiomas have taxed the ingenuity and surgical expertise of the most skilled neurosurgeons including Cushing,15 Olivecrona,40 MacCarty,32 Guiot, Derome, and colleagues,16,17,21,24 Basso, et al.,1 Castellano, et al.,12 Stern,46 Dolenc,8,19 Jane and colleagues,27,28 Al-Mefty and colleagues,2,7 and others. Their observations and contributions, however, are sometimes overlooked by neurosurgeons and ophthalmologists confronted by these unusual tumors, which are located in the complex anatomical boundary zone between the intracranial and orbital compartments and usually present with proptosis only and minimal, if any, neurological symptoms.

Complete surgical excision is the benchmark of successful surgery for meningioma. Despite attempts at this, however, there is still an estimated 35% to 50% recurrence rate associated with sphenoid wing meningiomas with orbital extension.4,30 Our purpose is to summarize our experience with 15 patients who had recurrent or initially inadequately resected sphenoid-orbital meningioma. The following points will be emphasized: 1) the frequent failure to diagnose this lesion correctly; 2) the utility of advanced neuroimaging modalities; 3) the importance of early aggressive surgical therapy; 4) the failure (by ourselves and others) to radically excise bone, tumor, dura, and periorbita at the first operation; 5) the microsurgical technique for removal through a small craniotomy without complicated reconstruction of the orbit or temporal fossa; and 6) the
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TABLE 1

Presenting symptoms in 15 patients with recurrent sphenoid-orbital meningioma

<table>
<thead>
<tr>
<th>Symptom</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>proptosis</td>
<td>13</td>
</tr>
<tr>
<td>blindness</td>
<td>1</td>
</tr>
<tr>
<td>decreased visual acuity or blurring</td>
<td>6</td>
</tr>
<tr>
<td>abnormal visual fields</td>
<td>8</td>
</tr>
<tr>
<td>palsy of 3rd, 4th, or 6th cranial nerve</td>
<td>2</td>
</tr>
<tr>
<td>decreased sensation in 5th cranial nerve</td>
<td>3</td>
</tr>
</tbody>
</table>

role of radiation treatment of residual tumor in patients who would experience serious morbidity from attempted total removal.

Clinical Material and Methods

Patient Population

From 1975 to 1992, we managed 130 patients with sphenoid wing meningioma having various degrees of orbital involvement. A detailed review of these patients is currently underway and will be reported separately. Three patients with sphenoid-orbital involvement returned after undergoing attempts at surgical removal with the development of recurrent tumor, necessitating a second operation. An additional 12 patients were referred to us from other institutions under the same conditions. In all 15 patients there had been inadequate primary resection of bone, dura, or periorbita during the initial operation, which contributed to the recurrence. None of these patients underwent radiation therapy prior to the second operation.

There were 11 women and four men in this series. The mean age at the time of initial diagnosis was 46 years (range 28 to 62 years). The mean time between operations was 46 months (range 12 to 118 months). Ten of the 15 patients received radiation therapy following the second operation (approximately 5000 rad locally to the tumor).

The most common presenting complaint of these patients with sphenoid-orbital meningioma was progressive proptosis (Table 1), usually occurring over several years. Four patients had a history of proptosis that was initially misdiagnosed as sinusitis. The most common cranial nerve deficit was optic neuropathy, which was more common after the tumor had recurred in the region of the optic canal or orbital apex. Two patterns of visual field deficit were apparent: an inferior arcuate field loss was seen with posterior optic nerve encasement, and a constricted field with enlarged scotoma was seen in those tumors primarily involving the optic foramen. In three patients diminished sensation was found in the ophthalmic and maxillary divisions of the fifth cranial nerve. All tumors were large at the time of reoperation and involved the orbit and the greater and lesser wings of the sphenoid bone.

Neuroimaging Techniques

Although plain roentgenograms routinely demonstrate hyperostosis, computerized tomography (CT) and magnetic resonance (MR) imaging were diagnostic of orbital and intracranial neoplasm and were most useful in mapping out the area of planned resection. In addition to hyperostotic thickening, bone remodeling of the orbital roof and anterior sphenoid wing were indicative of tumor infiltration. In all cases, gadolinium-enhanced MR imaging proved invaluable in diagnosing recurrent intradural tumor (Fig. 1). In three cases with prominent sphenoid wing hyperostosis, the enhanced CT scan failed to show dural or intracranial tumor. The MR image with gadolinium enhancement, however, demonstrated intradural tumor undetected by CT. In our experience, virtually all patients with hyperostotic sphenoid wing meningioma have infiltration of the anterior temporal dura that requires contrast-enhanced MR imaging for preoperative visualization.

It was common for marked hyperostosis to be misdiagnosed as fibrous dysplasia; these cases were more difficult surgically but radical resection was still possible (Fig. 2). Carotid angiography was not routinely performed unless there was marked hyperostosis, in which case preoperative embolization was considered.
Embolization was performed concurrently if angiography demonstrated a specific accessible feeding vessel supplying the tumor (Fig. 3).

Operative Technique

Residual or recurrent sphenoid-orbital tumors may be quite extensive (Fig. 4A). The goal of surgery is to resect the entire greater and lesser wings of the sphenoid bone up to the superior orbital fissure and then the anterior clinoid process and the orbital roof, and to decompress the foramen rotundum, foramen ovale, and the optic canal (Fig. 4B). A relatively small temporal incision centered on the pterion is used (Fig. 4C). Larger incisions and more extensive bone flaps do not give increased access to these neoplasms. A burr hole is made just behind the superior line of the hyperostotic bone and the bone cut is extended linearly (Fig. 4D). A high-speed drill* is used for expeditious removal of the thickened bone after dissecting the dura from the greater and lesser wings of the sphenoid bone (Fig. 4E and F).

The dissection remains entirely extradural during removal of the greater and lesser sphenoid wings and of the anterior clinoid process. The optic canal may or may not be unroofed, contingent upon tumor involvement. If there is marked hyperostosis, it is prudent to open the dura in order to identify the optic nerve intradurally prior to unroofing the optic canal.

After radical resection of the sphenoid bone, the roof of the orbit, and the bone of the anterior temporal fossa, the basal temporal dura is opened to remove the tumor, which has invariably invaded the basal dura. We do not advocate opening the dura over the cavernous sinus for tumor removal in patients without cranial neuropathy and in those with proptosis only. Nerves passing through the cavernous sinus and the superior orbital fissure are relatively resistant to neoplastic compression, but are clearly subject to palsy from surgical attempts to remove the tumor-involved dura; however, we appreciate that there are differences of opinion in this regard.

The orbital component of these sphenoid-orbital tumors usually invaginates rather than invades the orbital content. The periorbital fascia is therefore excised with its intraorbital tumor, akin to removing a convexity meningioma from the brain. If the ethmoid or sphenoid sinus is entered, meticulous repair with a pericranial flap or temporal muscle is required to prevent the formation of a cerebrospinal fluid fistula. Thorough waxing of the often aerated anterior clinoid process is performed. Pericranium or lyophilized dura is used to reconstruct the basal temporal dura. We have not found it necessary to reconstruct the roof of the orbit or to inlay any bone or other materials in the capacious space remaining after resection of tumor-involved bone. Pulsating exophthalmos has not occurred in over 200 cases treated with orbital decompression. A radiotranslucent cranioplasty is used to cover the lateral skull defect and provide support for the temporal muscle. A subgaleal drain is placed for 2 days to reduce swelling and peri-orbital ecchymosis.

Results

A marked reduction in proptosis of up to 14 mm was noted in 14 of the 15 patients. Two of six patients presenting with decreased visual acuity or slight visual blurring returned to normal visual acuity, and three of eight patients with abnormal visual fields showed improvement on perimetric testing. Cosmetically, all patients were satisfied with the operative result.

There have been no deaths and only minimal morbidity, with the exception of one patient who developed blindness 24 hours after surgery due to central retinal artery occlusion, possibly related to the use of Gelfoam and/or fibrin glue. One patient, treated early in the series, suffered persistent proptosis due to inadequate lateral orbital bone removal, one patient developed a fourth cranial nerve palsy, and one had a temporary third cranial nerve palsy. An additional complication in one patient was hypopituitarism and diabetes insipidus after aggressive removal of a tumor with a large medial component and suprasellar extension.

Pulsating exophthalmos occasionally occurred for the first several days following surgery and then was no longer noticeable. Ptosis and double vision for several days or weeks were common, due to the manipulation and dissection around the superior rectus and levator muscles. Follow-up imaging demonstrated complete gross excision of tumor in nine of these cases (Fig. 5). In three cases the tumor extended into the cavernous sinus: in one case hyperostosis remained in the sphenoid sinus and in the other two residual tumor was left within the dura of the basal optic canal. Five patients had residual tumor remaining in the dura of the superior orbital fissure or the cavernous sinus, noted at surgery but not always visualized on postoperative scans. Ten patients underwent radiation therapy, con-

* For example, the high-speed drill manufactured by Midas Rex, Inc., Fort Worth, Texas.
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Fig. 4. A: Drawing showing the usual extent of a sphenoid-orbital meningioma (superior view). B: Drawing depicting the goal of surgical resection. C: Skin incision for removal of a recurrent sphenoid-orbital meningioma. D: Location of the burl hole placed prior to drilling and removal of hyperostotic bone. E: Removal of the lesser sphenoid wing and the lateral orbital roof with a high-speed drill. F: Appearance after extradural excision of the greater and lesser wings of sphenoid bone and removal of the orbital roof and the lateral wall of the orbit and the tumor-involved periorbita.

sisting of approximately 5000 rad administered so as to avoid the retina and lacrimal gland. Long-term follow-up review is required, but we have not seen progressive growth to date in those patients who received radiation therapy, with a follow-up period ranging from 16 to 95 months.

Discussion

Sphenoid wing meningiomas constitute approximately 18% of all intracranial meningiomas. Many involve the peri orbital tissues, since the sphenoid bone forms the posterior and lateral walls of the orbit. Despite attempts at total excision, in some series there is an approximate 5-year probability of recurrence of 34% and a 10-year probability of about 54%, with or without orbital involvement.

The reasons for recurrence include the following: 1) failure to diagnose the tumor early due to confusion with other entities such as fibrous dysplasia, Graves' disease, and chronic sinusitis; 2) inadequate surgical resection due to the complicated anatomical boundaries of the orbit, the cavernous sinus, and the sensitive cranial nerves and vascular structures in the medial aspect of the sphenoid wing; 3) failure to appreciate that hyperostotic bone is indeed part of the neoplastic process and almost always has dural and en plaque extension, which is often not seen on CT scans; 4) the biological behavioral tendency of these tumors to insinuate themselves into the foramina, fissures, crevices, and interstices of the orbit, basal skull, and dura; and 5) the apprehension of surgeons that they will produce iatrogenic morbidity or even mortality associated with a too-radical tumor resection in this difficult neuroanatomical area.

In most patients, the initial symptom is unilateral exophthalmos, a painless cosmetic deformity with no neurological deficits. There is an understandable tendency to delay surgical intervention for such an ap-
The role of radiation therapy versus direct surgical intervention in the cavernous sinus and supraorbital fissure remains controversial. Recently, many authors have suggested that the cavernous sinus should not be a barrier to total excision of nonmalignant tumors. \(^2\), \(^7\), \(^43\) Although we recognize the tremendous surgical advances in this area, morbidity and mortality are significantly increased with such dissection and Ojemann’s recent admonition \(^2\) to first do no harm in skull-base and cavernous sinus surgery is well advised and strongly supported by us. Furthermore, virtually all of our patients were asymptomatic except for cosmetic or visual symptoms preoperatively. We have, therefore, been aggressive in recommending postoperative radiation when tumors are subtotally removed because of dural or cavernous sinus invasion or if there is recurrence demonstrated by neuroimaging scans.

Recent reports tend to substantiate our approach. Taylor, et al., \(^47\) in commenting on “the meningioma controversy” demonstrated that postoperative radiation therapy improved long-term local control of recurrent or subtotally removed meningiomas. They reported a 10-year survival rate of 81% for patients who received postoperative radiation therapy compared with 49% for those who did not. In the recurrent category, they demonstrated an 89% survival rate for patients who received radiation therapy compared with 43% for those who underwent another operation and did not receive additional therapy.

Miralbell, et al., \(^37\) reported on 36 patients with meningioma treated for primary or recurrent disease by subtotal resection and external beam radiation. They compared this group to a control group of 79 patients managed by subtotal resection alone from 1962 to 1980. They concluded that progression-free survival was significantly better in those patients receiving irradiation following initial surgery. In that study, 88% of patients were failure-free at 8 years compared with 48% of patients failure-free at 8 years if they underwent surgery alone. When treatment was given at first recurrence, the irradiated group again did better, with a 78% 8-year failure-free survival rate compared with only 11% in those treated by surgery alone. More recently, Kumar, et al., \(^36\) used stereotactic brachytherapy for tumors extending into the cavernous sinus and clivus and concluded that both recurrent and primary meningiomas of the skull base can be treated effectively with brachytherapy. Although not all authors are supportive of postoperative radiation therapy for meningiomas, many recent publications have supported its application for unresectable recurrent meningioma. \(^6\), \(^11\), \(^20\), \(^22\), \(^29\), \(^36\), \(^37\), \(^44\), \(^46\)

**Conclusions**

We believe sphenoid-orbital tumors should be removed early and as radically as possible to obviate future recurrence, patient age and medical condition permitting. Although the word “radical” may have unpleasant connotations, these operations are primarily performed extradurally with minimal cerebral retraction and morbidity. We do not recommend aggressive
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resection of tumor-invaded structures in the cavernous sinus or superior orbital fissure because of the attendant serious morbidity if cosmetic defects or minimal symptoms are the only complaints. Rather, radiation therapy should be considered if any residual tumor remains postoperatively or as soon as follow-up neuroimaging demonstrates recurrent tumor after an initial radical bone and tumor removal. Orbital reconstruction is not required and a temporal craneoplasty is sufficient for excellent cosmetic results.

References

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Manuscript received September 16, 1992. Accepted in final form March 12, 1993.
This work was supported by Allegheny Singer Research Institute and a grant from the Cameron Coca Cola Corporation.
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