Orbital tumors treated using transcranial approaches: surgical technique and neuroophthalmological results in 41 patients

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Object. Orbital tumors can be divided schematically into primary lesions, originating from the orbit itself, and secondary lesions, extending to the orbit from neighboring structures. These tumors are variable in their biological nature and in their location. The authors evaluate 41 cases of benign and malignant tumors involving the orbit and discuss the surgical challenge, which involves tumor removal, preserving visual function and cosmetic reconstruction.

Methods. The authors performed a retrospective analysis of a series of all cases involving patients who underwent surgery for treatment of orbital tumors in their hospital between December 2003 and December 2006. Data were collected from the patients’ files in the hospital’s outpatient clinic, operative notes, and pre- and postoperative imaging studies.

Results. The authors identified 41 patients who met the inclusion criteria (age range 14–82 years, mean 42.2 years, standard deviation 22.4 years). The most common presenting symptoms were proptosis and/or partial ptosis (11 cases [27%]) and headache (7 cases [17%]). In 5 (12%) cases, the tumors were primary intraorbital lesions, and in 34 cases (83%) they were secondary. Two patients had metastases to the orbit. The most common lesion types were meningioma (10 cases [24%]) and osteoma (7 cases [17%]). In 24 cases a midline approach through a frontal craniotomy or a subcranial approach was used; in 13 a lateral approach was used; and in 4 a biopsy procedure was performed. Maxillectomy through a Weber–Ferguson approach or a facial degloving approach was added in 5 cases to complete tumor removal. Duration of follow-up was 1–38 months (mean 20 months). None of the patients died as a result of the procedure, and there were relatively few complications. Excluding the patients who underwent orbital exenteration, none of the patients had visual deterioration following surgery, and most had no change in their visual condition. Two patients had temporary diplopia, 1 had a cerebrospinal fluid leak, and 1 had enophthalmos as a result of the procedure, and there were relatively few complications. Excluding the patients who underwent orbital exenteration, none of the patients had visual deterioration following surgery, and most had no change in their visual condition. Two patients had temporary diplopia, 1 had a cerebrospinal fluid leak, and 1 had enophthalmos as a result of the procedure.

Conclusions. Orbital tumors can be treated safely using transcranial approaches in many cases. Preoperative imaging can accurately define the compartments involved and the surgical approach needed for tumor removal. A multidisciplinary team of surgeons facilitates optimal tumor removal and skull base sealing as well as good cosmetic results. (DOI: 10.3171/FOC-07/11/E11)

Key Words • orbital decompression • orbital tumor • reconstructive surgery • skull base tumor • surgical approach

Orbital tumors may be primary, meaning they originated from orbital structures, or secondary, meaning they extend into the orbit from structures outside the orbit. A third group of tumors, involving the orbit are metastatic. These tumors are variable in their biological nature and in their location. Orbital tumors are also divided anatomically, into intraconal, extraconal, and infraconalicular. This distinction is made on the basis of the tumor’s relationship with the muscle cone, with the infraconalicular group of tumors being those lesions that are least partially within the optic canal. Patients with primary or secondary orbital tumors usually present with exophthalmos, pain, diplopia, swelling, tearing, and blurred vision. Although exophthalmos is a common sign in orbital tumors, this sign can be seen in other conditions, such as thyroid disease, orbital varices, collagen vascular disease, prolapse of intracranial contents into the orbit, and congenital defect of the orbital wall.

There are 2 major types of surgical approaches to orbital tumors: the transorbital approaches, which are usually performed by an ophthalmologist, and the extraorbital approaches, which are best performed by a team that includes a neurosurgeon or a head and neck surgeon as well as an ophthalmologist. In most instances, anterior lesions are
treated via transorbital approaches, whereas lesions of the posterior third are best managed via extrorbital approaches. There may be some instances, however, where this rule, cannot be applied. In some cases, posterior lesions can be approached via extended or combined transorbital approaches, and lesions of the middle third of the orbit are easily accessible via extrorbital approaches. The drawback to this approach is the need for extensive dissection. The transcranial approach with the orbital rim preserved was initially developed by Dandy and has since been modified several times.

In addition to location, other factors, such as the size of the lesions, the goal of the surgery (biopsy, debulking, or gross–total excision), and the characteristics of the tumor must be considered when selecting an approach. In this paper we focus on extrorbital approaches and describe our series of patients treated using these approaches. The orbital approaches are described in detail elsewhere.

The transcranial approach in which the orbital rim is preserved was initially developed by Dandy and has been subsequently modified. The transcranial approach with orbital osteotomy was initially described by Frazier in 1913, and has since been modified several times. This approach is useful in cases of tumors involving the medial orbit and the anterior skull base. The subcranial approach can also be useful in midline lesions involving the orbit, the anterior skull base, and the paranasal sinuses.

Imaging studies used in the diagnosis and management of orbital tumors have changed dramatically in the last few decades. Ultrasonographic examination of the orbit is still helpful in the evaluation of cystic lesions and also for vascular lesions. Magnetic resonance imaging gives high-resolution images of the normal components of the orbit and of nonosseous lesions in three dimensions. In cases of osseous lesions, CT is the modality of choice, either alone or in combination with an MR imaging. The drawback to using CT in examining the eye is that it uses ionizing radiation and may produce cataract.

Angiography is still the imaging modality of choice for vascular lesions, such as arteriovenous malformations and low-flow arteriovenous fistulas. It is also useful in cases of the more vascular orbital tumors (such as meningiomas) for defining the extent of vascularity prior to surgery. Cerebral angiography should be undertaken in any patient with pulsatile exophthalmia.

**Clinical Material and Methods**

We performed a retrospective review of all the cases (41 cases) in which patients underwent surgery for the treatment of orbital tumors at our institution between December 2003 and December 2006. In most of these cases (66%), surgery was performed by a cooperative team of neurosurgeons and head and neck surgeons, with or without plastic surgeons. Data was collected from the hospital records, the outpatient clinic, and operative notes. All patients underwent surgery for the treatment of orbital tumors in our institution. The patients’ ages ranged from 14 to 82 years (mean 42.2 years, standard deviation 22.4). There were 25 female patients (61.9%) and 16 male patients (38.1%). The most common clinical presentations were proptosis and/or partial ptosis (11 patients, 26.8%) and headache (7 patients, 17.1%). In 5 patients (12.2%) the lesions were primary intraorbital tumors; in 34 (82.9%) they were classified as secondary. Two patients had metastases to the orbit. The most common lesions were meningiomas (10 cases [24.4%]) and osteoma (7 cases [17.1%]). Other benign lesions encountered in our case series were angiofibroma, esthesioneuroblastoma, histiocytosis, and ossifying fibroma. Malignant neoplasms were found in 12 (29.3%) of the patients. Malignant orbital tumors included squamous cell carcinoma, hemangiopericytoma, malignant peripheral nerve sheath tumor, high-grade osteosarcoma, adenoid cystic carcinoma, metastatic Ewing sarcoma, and metastatic breast carcinoma (Table 1). In 4 patients with malignant tumors involving the orbit, we performed an orbital exenteration. Four of the patients underwent a biopsy procedure.

**Operations and Outcomes**

Surgery was performed by a multidisciplinary team in 65.8% of the cases; 27 operations were performed by neurosurgeons and head and neck surgeons, and in 8 of these 27 cases a plastic surgeon joined the team. Follow-up time

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**TABLE 1**

*CA = carcinoma; PNST = peripheral nerve sheath tumor.*

<table>
<thead>
<tr>
<th>Pathological Condition</th>
<th>No. of Cases (%)</th>
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<tbody>
<tr>
<td>meningioma</td>
<td>10 (24.4)</td>
</tr>
<tr>
<td>osteoma</td>
<td>7 (17.1)</td>
</tr>
<tr>
<td>squamous cell CA</td>
<td>4 (9.8)</td>
</tr>
<tr>
<td>angiofibroma</td>
<td>4 (9.8)</td>
</tr>
<tr>
<td>esthesioneuroblastoma</td>
<td>3 (7.3)</td>
</tr>
<tr>
<td>metastasis</td>
<td>3 (7.3)</td>
</tr>
<tr>
<td>histiocytosis</td>
<td>2 (4.9)</td>
</tr>
<tr>
<td>ossifying fibroma</td>
<td>2 (4.9)</td>
</tr>
<tr>
<td>osteosarcoma</td>
<td>2 (4.9)</td>
</tr>
<tr>
<td>malignant PNST</td>
<td>1 (2.4)</td>
</tr>
<tr>
<td>hemangiopericytoma</td>
<td>1 (2.4)</td>
</tr>
<tr>
<td>osteoid osteoma</td>
<td>1 (2.4)</td>
</tr>
<tr>
<td>adenoid cystic CA</td>
<td>1 (2.4)</td>
</tr>
</tbody>
</table>

We divided the 41 patients into 4 groups depending on the extent of tumor removal: complete (gross–total) tumor removal, subtotal removal (1–2% of the lesion left in place); partial removal (substantial tumor left in place); biopsy procedure only.

Postoperative follow-up was performed in the outpatient clinic and included a general clinical and neuroophthalmological examination. An MR imaging study was performed 3 months following surgery. The extent of tumor resection was determined by the surgeons’ impression during surgery and confirmed by means of the 3-month postoperative MR images.

**Results**

**Patient and Lesion Characteristics**

Between the years of 2003 and 2006, 41 patients underwent surgery for the treatment of orbital tumors in our institution. The patients’ ages ranged from 14 to 82 years (mean 42.2 years, standard deviation 22.4). There were 25 female patients (61.9%) and 16 male patients (38.1%). The most common clinical presentations were proptosis and/or partial ptosis (11 patients, 26.8%) and headache (7 patients, 17.1%). In 5 patients (12.2%) the lesions were primary intraorbital tumors; in 34 (82.9%) they were classified as secondary. Two patients had metastases to the orbit. The most common lesions were meningiomas (10 cases [24.4%]) and osteoma (7 cases [17.1%]). Other benign lesions encountered in our case series were angiofibroma, esthesioneuroblastoma, histiocytosis, and ossifying fibroma. Malignant neoplasms were found in 12 (29.3%) of the patients. Malignant orbital tumors included squamous cell carcinoma, hemangiopericytoma, malignant peripheral nerve sheath tumor, high-grade osteosarcoma, adenoid cystic carcinoma, metastatic Ewing sarcoma, and metastatic breast carcinoma (Table 1). In 4 patients with malignant tumors involving the orbit, we performed an orbital exenteration. Four of the patients underwent a biopsy procedure.
Orbital tumors and transcranial approaches

TABLE 2

Distribution of surgeries according to the extent of resection

<table>
<thead>
<tr>
<th>Extent of Resection</th>
<th>No. of Patients (%)</th>
</tr>
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<tbody>
<tr>
<td>complete (gross-total resection)</td>
<td>30 (73.2)</td>
</tr>
<tr>
<td>subtotal (up to 2% of lesion left)</td>
<td>5 (12.2)</td>
</tr>
<tr>
<td>partial</td>
<td>2 (4.9)</td>
</tr>
<tr>
<td>biopsy only</td>
<td>4 (9.8)</td>
</tr>
</tbody>
</table>

ranged between 1 and 38 months (mean 20 months). None of the patients died as a result of the surgical procedure and there were relatively few complications. Two patients died as a result of progression of their disease. Two patients had temporary diplopia, 1 patient had a CSF leak, and 1 patient had enophthalmos following removal of an orbital osteosarcoma. In most of the cases (85%) complete or subtotal removal of the tumor was achieved (Table 2).

Visual function in most patients was excellent. Twenty-three patients had normal vision prior to surgery and no change in their vision following surgery. Four patients underwent orbital exenteration. Two patients died as a result of tumor progression and we were not able to follow their visual status following surgery. Five patients were unavailable for follow-up examination, and 3 were blind prior to surgery. One patient had rapid visual deterioration prior to surgery, in both eyes, and following surgery on the right eye she had a relative afferent pupillary defect, a nasal defect, and disc pallor; the left side was normal. One patient was severely disabled, and hence was unable to undergo a thorough ophthalmological evaluation. Two patients had diplopia prior to surgery, which resolved following surgery.

Surgical Technique

Different tumor locations and lesion types mandated different surgical techniques for removal of the tumors. We divided the surgical techniques into 2 categories: 1) a lateral approach and 2) anterior approaches including a frontal craniotomy or a subcranial approach. In the lateral approach category we focused on our technique for sphenoorbital meningiomas. These lesions were the most common in our case series (10 of 41 cases). We used the anterior approaches for tumors involving the anterior skull base or the medial part of the orbit with or without extension to the paranasal sinuses.

The Lateral Approach. The patient is placed supine, in a 3-point fixation system, with the head slightly extended and turned contralateral to the tumor. The skin is incised starting a few millimeters anterior to the tragus, continuing to the midline. We use interfascial dissection to preserve the frontal branch of the facial nerve. The temporalis muscle is elevated. If there is tumor involvement, as there was in some of our cases, the tumor is removed and the temporalis vasculature is preserved. In most of these sphenoorbital meningiomas the soft component of the tumor as well as hyperostosis can be seen at this point during the surgery. After removal of the soft component, the hypertrophic bone is removed by drilling. In cases in which the bone is very thickened by the tumor, a craniectomy is performed using a high-speed drill. Bone is removed until normal dura mater is reached posteriorly and the periorbit anteriorly. A frontotemporal craniotomy is performed around the cranial approach depending on the compartments involved by the tumor. The subcranial approach has been described in detail elsewhere. Briefly, after incising the skin above the hairline, we create a bicoronal flap in a supraperiosteal plane. A flap is elevated anteriorly beyond the supraorbital ridges and laterally superficial to the fascia of the temporalis. The pericranial flap is elevated up to the periorbita, and the supraorbital nerves and vessels are carefully separated from the supraorbital notch. The lateral and medial walls of the involved orbit are exposed, and the anterior ethmoidal arteries are coagulated and cut. The pericranium is elevated above the nasal bones, and the flap is rotated forward and held over the face throughout the rest of the procedure. Titanium miniplates are applied to the frontal bones and removed before the osteotomies to facilitate the exact repositioning of the bone segments at the end of the procedure. At this stage the tumor is removed from the intracranial compartment, and the orbital part is removed as described previously for the lateral approach. One or both sides of the cribiform plate and olfactory filaments are preserved whenever possible.

Figure 2 (left) shows a bone window CT scan obtained in a patient with malignant osteosarcoma involving the ethmoidal sinus and the left orbit. This patient underwent surgery via a subcranial approach. Figure 2 (right), is a T2-weighted MR image obtained in the same patient and shows the severe compression of the optic nerve and orbital structures caused by the lesion.

Reconstruction in cases of anterior skull base lesions must prevent a CSF leak through the paranasal sinuses and must also achieve a good cosmetic result. We use fascia lata...
sutured in a watertight manner for reconstruction of the dura mater. As described for the lateral orbital wall, in most cases there was no need for reconstruction of the osseous orbit. However, reconstruction of the medial orbital wall was performed in cases in which total removal of this segment was necessary or a large piece of the periorbit was excised. In these cases we used split calvarial bone grafts or 3D titanium mesh coated with pericranium. If the eye globe was exenterated we used a temporalis muscle flap and split-thickness skin graft to cover the socket. A dacryocystorhinostomy is performed in patients undergoing orbital wall resection or medial maxillectomy following reconstruction. A centripetal compression of both globes is performed in order to reduce telecanthus. We perform cranialization of the frontal sinus and the nasofrontal duct is plugged with a piece of muscle and biological glue.

Discussion

There are 2 major types of surgical approaches for removal of orbital tumors: the transorbital approaches and the extraorbital approaches. This article describes cases in which extraorbital approaches were used. In most cases, anterior lesions are treated via transorbital approaches, whereas lesions of the posterior third of the orbit are best managed via extraorbital approaches. In some cases, however, posterior lesions can be approached via extended or combined transorbital approaches, and lesions of the middle third of the orbit are easily accessible via extraorbital approaches. In addition to location, other factors, such as the size of the lesions, the goal of the surgery (biopsy, debulking, or gross–total excision), and the characteristics of the tumor must be considered in selecting an approach.

Using transcranial approaches facilitates visualization of intraorbital lesions through the lateral wall of the orbit and its roof. These approaches are best used for lesions in the orbital apex and the superior orbital fissure. Optic nerve gliomas and meningiomas are examples of apical lesions, and hemangiomas are typical superior orbital fissure lesions. The approach used for sphenoid wing meningiomas is similar to that used to reach lateral lesions, but because the former can extend medially into the optic canal, they can be treated by means of a combined approach. Most tumors in our case series were secondary tumors, meaning they originated from structures outside the orbit. We used different variations of the 2 approaches described in the surgical technique section depending on the tumor location, size, and other variables. None of the patients in our case series died as a result of the surgical procedure, and there were relatively few complications. Two patients had temporary diplopia, 1 patient had a CSF leak, and 1 had enophthalmos following removal of a high-grade osteosarcoma. In most of our cases, the surgical procedure was performed by a multidisciplinary team. Our team consists of a head and neck surgeon, a neurosurgeon, and a plastic surgeon. We have found this combination of skills helps us to achieve complete tumor removal, optimal sealing of the skull base and CSF space, and good cosmetic results.

Our experience shows that in most cases using the tran

FIG. 1. Axial (left) and coronal (right) Gd-enhanced T1-weighted MR images showing a sphenoorbital meningioma. The lesion was treated via a frontotemporal approach.
Conclusions

Orbital tumors can be treated safely using a transcranial approach in many cases. Preoperative imaging can accurately define the compartments involved and the surgical approach needed for tumor removal. A multidisciplinary team approach facilitates optimal removal of the tumor and skull base sealing as well as good cosmetic results. Good visual outcomes were achieved in most cases in our series.

References

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Fig. 2. Left: Bone window CT showing malignant osteosarcoma involving the ethmoidal sinus and the left orbit. The lesion was treated via a frontal approach. Right: A T2-weighted MR image obtained in the same patient. Note the severe compression of the optic nerve and orbital structures.