Lateral orbitotomy for a maxillary nerve schwannoma: case report

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Authors of this report describe a Fukushima Type D(b) or Kawase Type ME2 trigeminal schwannoma involving the right maxillary division in a 59-year-old woman who presented with intermittent right-sided facial numbness and pain. This tumor was successfully resected via a right lateral orbitotomy without the need for craniotomy. This novel approach to a lesion of this type has not yet been described in the scientific literature. The outcome in this case was good, and the patient’s intra- and postoperative courses proceeded without complication. The epidemiology of trigeminal schwannomas and some technical aspects of lateral orbitotomy, including potential advantages of this approach over traditional transcranial as well as fully endoscopic dissections in appropriately selected cases, are also briefly discussed.

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KEY WORDS trigeminal schwannoma; lateral orbitotomy; maxillary nerve; surgical technique

In the following article, in the context of a specific case report, we describe the techniques and nuances involved in the resection of a schwannoma arising from the maxillary division of the right trigeminal nerve using isolated lateral orbitotomy without separate craniotomy. The lesion discussed here involved the ventral margin of Meckel’s cave and the cavernous sinus on the right, with some extracranial extension including the deep aspect of the right orbit. This operative approach, while well established for many intraorbital lesions and a few intracranial ones including optic glioma and sphenoorbital meningioma, has not been described for trigeminal schwannomas of this or any other specific anatomical involvement. Many other operative approaches to intracranial schwannomas have been described, including many open techniques such as the pterional craniotomy, which probably would have been the best choice among the many transcranial options had one been applied in the presented case. Additionally, 5 fully endoscopic approaches to trigeminal schwannomas have been described, including the endoscopic endonasal transpterygoid and endoscopic endonasal transmaxillary transpterygoid approaches, which are also reasonable choices for a lesion with the anatomical characteristics described in this report. We aim to provide sufficient detail of the operative technique and specific anatomy encountered in this case so that this article effectively informs others’ consideration of the lateral orbitotomy approach to trigeminal schwannomas for future study or for application in patients with a similar presentation.

Case Report

History and Examination

A 59-year-old female presented with several months of subjective intermittent right-sided facial numbness, worst in the evenings, in the distribution of the right maxillary division of the trigeminal nerve. She had also experienced brief episodes of lancinating pain approximately 2–3 times each month in the same distribution. She denied headache, visual disturbances, vertigo, weakness, or paresthesias. Her medical history was notable for morbid obesity, hypertension, hyperlipidemia, diabetes mellitus, asthma, and seasonal allergies; her medications were appropriate for the management of these conditions. Her surgical, family, and social histories were noncontributory.
Her neurological exam was without deficit, including facial sensation throughout all branches of the trigeminal nerve bilaterally with preserved corneal reflexes. The remaining cranial nerves were intact. She was noted to have normal bulk as well as tone, full strength as well as sensation throughout, normal reflexes without pathological reflexes, and normal cerebellar function. Special testing was unremarkable. A general physical exam was notable for obesity and was otherwise within normal limits. No dermatological lesions, ophthalmological abnormalities, or hearing deficits were noted. Laboratory assessment showed elevated hemoglobin A1C and serum lipids but was otherwise unrevealing.

Radiological Findings

Magnetic resonance imaging with gadolinium demonstrated a right-sided 2.1 × 2.0 × 2.0–cm mass involving the ventral margin of Meckel’s cave and the cavernous sinus abutting the internal carotid artery. There was also noted extracranial extension involving the pterygopalatine fossa and the deepest aspect of the orbit, as well as resultant bony deformation of the nasal sinuses (Fig. 1). The mass demonstrated intermediate T1 signal, high T2 signal, and homogeneous postcontrast enhancement. These radiological findings were consistent with schwannoma and, in this anatomical position, with a Fukushima Type D(b) or Kawase Type ME2 trigeminal schwannoma specifically (Table 1). The differential diagnosis also included chordoma, meningioma, metastasis, epidermoid cyst, and primary lymphoma of Meckel’s cave; however, each of these lesions is generally associated with radiological and clinical findings not appreciated here and thus less likely.

Operative Approach and Resection

The patient was positioned supine in a Mayfield head holder with 1 pin over the left frontal area and 2 pins at the occiput. She was rotated approximately 15° to the left side, and a slight shoulder roll was placed on her right, elevating the right malar eminence to the highest point in the operative field. The operative site was then marked for both a right lateral orbitotomy and a possible right pterional craniotomy if additional exposure was needed for complete resection. The field was sterilized with ophthalmic Betadine solution and draped in the usual fashion. A brief surgical time-out was performed, and then a 3- to 4-cm lateral eyelid crease incision was made from the central eyelid to approximately 2 cm beyond the lateral canthal angle. Figure 2 upper demonstrates the lateral eyelid crease incision used for the lateral orbitotomy.

Dissection was carried down to the periosteum of the lateral orbital rim using monopolar cautery on a microdissection needle. Following this preseptal dissection, a periosteal incision was made over the lateral rim (Fig. 2).

TABLE 1. Classification of trigeminal schwannomas*

<table>
<thead>
<tr>
<th>Type</th>
<th>Involved Portions of Trigeminal Nerve</th>
<th>Cranial Fossae</th>
<th>Extracranial</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fukushima classification system</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>A</td>
<td>Ophthalmic, maxillary, or mandibular division</td>
<td>None</td>
<td>Exclusively</td>
</tr>
<tr>
<td>B</td>
<td>Gasserian ganglion &amp; intracavernous portion</td>
<td>Middle</td>
<td>None</td>
</tr>
<tr>
<td>C</td>
<td>Pre–gasserian ganglion root only</td>
<td>Posterior</td>
<td>None</td>
</tr>
<tr>
<td>D(a)</td>
<td>Pre–gasserian ganglion root, gasserian ganglion, &amp; intracavernous portion (dumbbell type)</td>
<td>Middle posterior</td>
<td>None</td>
</tr>
<tr>
<td>D(b)</td>
<td>Gasserian ganglion, intracavernous portion, &amp; ophthalmic, maxillary, or mandibular division</td>
<td>Middle</td>
<td>Partial</td>
</tr>
<tr>
<td>Kawase classification system</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>Gasserian ganglion, intracavernous portion, ophthalmic, maxillary, or mandibular division in intradural space</td>
<td>Middle</td>
<td>None</td>
</tr>
<tr>
<td>P</td>
<td>Pre–gasserian ganglion root in subdural space</td>
<td>Posterior</td>
<td>None</td>
</tr>
<tr>
<td>E1</td>
<td>Ophthalmic or maxillary division in orbit</td>
<td>None</td>
<td>Exclusively</td>
</tr>
<tr>
<td>E2</td>
<td>Maxillary or mandibular division in pterygopalatine or infratemporal fossa only</td>
<td>None</td>
<td>Exclusively</td>
</tr>
<tr>
<td>MP</td>
<td>Pre–gasserian ganglion root, gasserian ganglion, &amp; intracavernous portion (dumbbell type)</td>
<td>Middle posterior</td>
<td>None</td>
</tr>
<tr>
<td>ME1</td>
<td>Gasserian ganglion, intracavernous portion, &amp; ophthalmic or maxillary division in orbit</td>
<td>Middle</td>
<td>Partial</td>
</tr>
<tr>
<td>ME2</td>
<td>Gasserian ganglion, intracavernous portion, &amp; maxillary or mandibular division in pterygopalatine or infra-temporal fossa</td>
<td>Middle</td>
<td>Partial</td>
</tr>
<tr>
<td>MPE</td>
<td>Pre–gasserian ganglion root, gasserian ganglion, intracavernous portion, &amp; any or all peripheral divisions with extracranial extension</td>
<td>Middle posterior</td>
<td>Partial</td>
</tr>
</tbody>
</table>

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The periosteum was incised in this fashion along the lateral orbital rim, and a Freer elevator was used to elevate it posteriorly into the orbit away from the external surface of the bone. Next, a subperiosteal dissection was performed over the rim, lateral orbital wall, and temporalis fossa. The temporalis muscle was separated from the external lateral orbital wall with the Freer elevator.

A 2-cm section of the lateral orbital rim was marked with the superior edge just inferior to the lacrimal gland fossa and the inferior edge at the level of the zygomatic arch. The periorbita was protected with a malleable retractor, while a Sonopet ultrasonic bone aspirator (Stryker) was used to create osteotomies (Fig. 2 lower). The bone was fractured and removed (Fig. 3 upper). The ultrasonic bone aspirator was then fit with a Payner 360° tip and used to remove the greater wing of the sphenoid bone (Fig. 2 lower) to expose the anterior aspect of the middle cranial fossa. Ultrasonic bone removal was continued to the superior and inferior orbital fissures and terminated when dura mater was encountered and a thin shell of cortical bone remained over the temporal lobe. This remaining bone was carefully removed with rongeurs and Takahashi forceps to create a large bony window, providing excellent exposure of the tumor (Fig. 3 lower).

Figure 4 demonstrates, via 3D reconstruction from neuronavigation MRI of the lesion, this exposure as it was appreciated intraoperatively as well as the described dissection. Figure 4 also demonstrates intraoperative exposure of the mass from this same perspective, though with a closer view, before and after bone removal from the lateral and posterior orbit. This perspective is similar to, but narrower than, the more global anatomy demonstrated in Fig. 2 lower. This narrower field of view and trajectory closely approximates what the operating surgeon sees through the operative microscope. The dashed line in Fig. 4 indicates the space previously occupied by the lateral orbital rim, illustrating the wide exposure provided by removing this bone wedge.
Self-retaining retractors were placed into the wound with a brain spatula on a Fukushima retractor placed over the right temporal dura, and another Fukushima retractor with a brain blade was placed over the right orbital contents. The tumor capsule was readily apparent at the base of the exposure, and a small incision in the capsule was made to remove a tumor section that was sent for frozen pathology, which resulted in a diagnosis of schwannoma. The capsule incision was expanded using bipolar electrocautery and micro scissors. The bulk of the tumor was removed and sent for permanent pathology with bipolar electrocautery, suction, and Decker pituitary rongeur.

Debulking of the tumor was then continued with Broden dissecting instruments to separate the tumor from its capsule, pituitary ring curettes to remove residual tumor from the capsule walls, a pituitary rongeur for periodic removal of larger fragments, and an ultrasonic aspirator, under an operative microscope (Fig. 5). Magnetic resonance imaging–based neuronavigation was used to supplement the direct visualization provided by operative microscopy. In Fig. 6 the crosshairs represent the end of a probe in the deepest aspect of the tumor bed with regard to the axis of exposure, during the final stages of resection. Though the operative corridor was somewhat limited in this case, excellent visualization and working angles were achieved through the combined use of the operative microscope and a 30° endoscope. At the far edges of the tumor cavity, direct visualization with the microscope was unacceptably limited. For these portions of the case, a 30° endoscope was used, which allowed for excellent visualization and safe curettage of the few remaining fragments of neoplastic tissue in a manner similar to that possible in an endoscope-assisted transsphenoidal resection of a pituitary mass.

Following resection and hemostasis, the lateral orbital wall was reconstructed with a porous polyethylene-embedded titanium mesh implant (Medpor Titan MTB OFW, Stryker) to recreate the orbital surface of the deep lateral wall and secured to the posterior edge of the bone flap with titanium screws. The bone flap was then placed back into its anatomical position within the lateral orbital rim and fixed with titanium plates and screws (Fig. 7). The periosteum was closed over the lateral orbital rim using buried interrupted 5-0 Monocryl sutures, and the skin was closed with deep dermal 5-0 Monocryl sutures and a running 6-0 Prolene suture.

Intraoperative neurophysiological monitoring of motor evoked potentials and somatosensory evoked potentials and electromyography of cranial nerves (CNs) V and VII were used throughout the approach and resection of the tumor. Monitoring techniques have been well described for more common lesions, such as the resection of vestibular schwannoma and microvascular decompression for trigeminal neuralgia, and similar methods were applied in this case. No adverse events were recorded throughout the duration of neuromonitoring, and no complications were noted intraoperatively during the case. The patient was extubated in the operating room prior to transfer to the surgical intensive care unit for standard postoperative monitoring overnight.

Hospital Course and Follow-Up

Sensation in the distribution of the right maxillary nerve and throughout the face was mildly diminished but intact on examination on postoperative Day 1. The remainder of the patient’s exam was unchanged from her preoperative status, including examination of the right eye, which revealed no injury to the eye with normal extraocular movements, full visual fields, and preserved visual acuity. She experienced minimal pain and took acetaminophen only as needed throughout her hospital stay. She remained clini...
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ically stable with continued preserved but mildly diminished sensation in the right maxillary distribution prior to early discharge home on the 3rd postoperative day. Postoperative MRI revealed expected postsurgical changes, no discernable residual tumor tissue, and no hematoma or other unexpected finding (Fig. 8).

Approximately 14 months postresection, the patient continued to have slightly diminished sensation in the right maxillary nerve distribution, with an otherwise unchanged and normal neurological exam. The right eyelid crease incision had healed without complication, and less than 2 cm of scar was appreciable lateral to the natural eyelid crease with minimal adverse aesthetic consequence (Fig. 9).

Pathological Findings

Pathological assessment confirmed the diagnosis of schwannoma. Compact areas of tumor cells in fascicles

Fig. 6. Screen capture of intraoperative neuronavigation with insets of the 3 standard anatomical planes and the operative trajectory. Crosshairs represent the end of a probe in the deepest aspect of the tumor bed. Figure is available in color online only.

Fig. 7. Three-dimensional CT reconstruction of the patient’s skull after reconstruction of the right orbit. Figure is available in color online only.

Fig. 8. Postoperative coronal post-Gd enhanced T1-weighted MR image of the patient’s head.
with some focal areas of palisading, Antoni A areas, can be appreciated in Fig. 10A. Other areas showing a relative paucity of tumor cells on a vaguely microcystic background, Antoni B areas, can be appreciated in Fig. 10B. Finally, diffuse S100 positivity was noted, a marker associated with the neural crest embryologic origin of schwannomas (Fig. 10C).

Interestingly, this specimen also demonstrated areas of a relatively rare histological variant termed “ancient change” (Fig. 10D). Ancient change is an important finding in schwannoma, as the cytological atypia occurring in these cases is similar to that seen in malignant peripheral nerve sheath tumors. Careful inspection of the specimen is needed for accurate diagnosis, determination of prognosis, and guidance of postoperative management; malignant neural sheath tumor diagnosis is made on the basis of elevated mitotic index and necrosis, neither of which were present in our patient. Cellular atypia in the absence of increased mitoses and necrotic areas is acceptable in an otherwise benign-appearing lesion, and maximal resection is generally curative. Ancient change is rare in schwannomas in general, and only a single report of trigeminal ancient schwannoma could be found in the literature.

Discussion

Schwannomas of the trigeminal nerve, while quite rare overall, are the second most common site of origin among the cranial nerves, following the vestibulocochlear nerve. Trigeminal schwannomas account for 8% or less of all intracranial schwannomas. Most tumors arise from the gasserian ganglion proximal to the division of the fifth CN into its 3 peripheral divisions, and when the peripheral divisions are involved, the ophthalmic branch is most commonly affected. Our patient’s presentation is rare, as the lesion arose from the maxillary division distal to the gasserian ganglion with no root involvement.

Review of the literature yielded 14 relevant case series with a total of 594 patients documented between 1988 and 2014; in these series, more than a dozen open operative approaches are described for trigeminal schwannomas of all sites of intracranial origin and Fukushima or Kawase type. Nearly all involve craniotomy and transcra-nial dissection to some degree regardless of lesion location; examples of frequently used transcra-nial approaches include, but are not limited to, frontotemporal (pterional), subtemporal, and retrosigmoid craniotomy. Alternatively, the endonasal endoscopic approach to trigeminal schwannomas has also been described. In 2012 Komatsu et al. published a cadaveric study that describes 5 fully endoscopic options for an approach to trigeminal schwannomas of various anatomical locations. Among these options are 2—endoscopic endonasal transpterygoid and endoscopic endonasal transmaxillary transpterygoid approaches—that access Meckel’s cave from an anteromedial trajectory and would probably have been reasonable alternative approaches to our patient’s lesion.

The safety of the endonasal approach to Meckel’s cave has been documented in a case series of 40 patients with lesions in this region, including various head and neck carcinomas, chondrosarcomas, chordomas, endodermal and enterogenous cysts, juvenile angiofibromas, meningiomas, pituitary adenomas, and trigeminal schwannomas. Among these 40 patients, only 2 suffered permanent deficit as an operative complication (trigeminal branch hypesthesia in both cases). The complication in 1 of these patients was attributable to the deliberate sacrifice of the maxillary nerve due to invasion by a pituitary macroadenoma. Only 2 case series included patients who had undergone endonasal endoscopic resection of a trigeminal schwannoma, totaling 10 patients altogether, that is, 6 of the 40 patients from the case series mentioned above and 4 from an additional series published in 2014. Of these 10 patients, 9 had gross-total resection (1 had posterior fossa disease that could not be accessed by endoscopic means and thus required postoperative stereotactic radiosurgery). Two pa-
tients suffered permanent deficits as operative complications: 1 case of worsened ophthalmic division neuropathy with associated keratopathy and 1 case of mandibular division hypesthesia.

Importantly, many of the patients included in the various case series cited above actually suffered from neurofibromatosis Type 2. This inherited condition is known to predispose to multiple and bilateral intracranial schwannomas classically arising from CN VIII, but other lesions, including those of the trigeminal branches, are known to occur. It is possible that the inclusion of a large number of patients with neurofibromatosis in the relevant case series affected the reporting of best management practices for patients suffering only an isolated intracranial schwannoma, as in our patient.

The operative approach to our patient’s lesion enabled a small incision, minimal soft tissue dissection, and bone removal through a less invasive extradural approach while gaining adequate exposure for complete gross tumor resection. The lateral orbitotomy is a commonly used surgical approach for a variety of orbital procedures, including tumor removal, fracture repair, and orbital decompression for thyroid-related orbitopathy. However, there are few descriptions of its use for intracranial processes such as the management of optic glioma and laterally situated sphenoorbital meningioma.2,3 A single case series reports lateral orbitotomy in conjunction with a basal temporal craniotomy in 4 patients with trigeminal schwannomas, but no instances of lateral orbitotomy alone. A solo right-sided lateral orbitotomy for a maxillary division schwannoma has not been reported in the literature. Though this is a novel approach for this particular lesion, the approach to the cavernous sinus region in general via isolated lateral orbitotomy is well described; Altay et al. demonstrated with photo-documentation in a cadaveric dissection study that the cavernous sinus, with its contained and surrounding structures, can be effectively exposed through the lateral orbit, including the gasserian ganglion and all peripheral divisions of the trigeminal nerve.4

The general principles of lateral orbitotomy dissection, microsurgical technique, and instrument use have been described and illustrated in great detail by Maroon and Kennerdell, and subsequently by Cockerham et al.4,14 Lateral orbitotomy for decompression of Graves’ orbitopathy, classically described by Leone et al. in 1989,11 is most similar to the specific technique described for the lesion in the case featured here. The techniques differ only insofar as a rim of bone left overlying the temporal lobe in decompression for Graves’ orbitopathy, whereas the sphenoid wing was entirely removed to expose the underlying middle cranial fossa in our case. The description and order of the steps involved is discussed above in Operative Approach and Resection for the benefit of our readers who may be considering this approach for one of their patients. However, an exhaustive discussion of the lateral orbitotomy with intent to fully educate the reader in its safe and successful performance is beyond the scope of this paper and is better done through the reference of one of the aforementioned sources.

The potential advantages of the lateral orbital approach to appropriately situated trigeminal schwannomas over the previously reported transcranial methods lie in its relative noninvasiveness while still providing excellent visualization throughout the surgical field. Lateral orbitotomy requires an incision of only a few centimeters, which can be largely hidden within the natural eyelid crease, producing excellent aesthetic results. No bone removal beyond the lateral orbital wall is required, sparing the patient any potential morbidity associated with the transcranial approach. Of course, the endonasal endoscopic approaches to trigeminal schwannomas share this advantage of minimal invasiveness, requiring no skin incisions at all, though perhaps at the cost of compromised visualization of the operative field and the theoretically higher risk of infection after having traversed the nonsterile nose. For the tumor featured in this report, we believe that the lateral orbital approach provided better exposure than an endoscopic approach would have, though the transpterygoid and transmaxillary transpterygoid approaches discussed above would have been reasonable alternatives. Our case also highlights the value of multidisciplinary collaboration, which allowed the combined expertise of neurosurgery and oculoplastic/orbital surgery to effect an excellent outcome for the patient.

This combination of microscopic, endoscopic, and minimally invasive lateral orbitotomy for a trigeminal lesion has not, to our knowledge, been previously described in the scientific literature. The favorable postoperative outcome, with generally preserved sensation in the distribution of the right maxillary nerve and no intra- or postoperative complications, suggests that this technique is safe and offers an alternative to the more extensive dissections that have been previously reported in cases of trigeminal schwannomas. Of course, only a minority of trigeminal schwannomas will lie in anatomical spaces that can be approached in the described fashion, and the technique’s feasibility must be considered based on individual anatomy. It is our hope that this case encourages further study to elucidate the advantages of this approach in conjunction with the tools available to the modern neurosurgeon.

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References


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Conception and design: Wallace, Cirivello. Acquisition of data: Meyer. Analysis and interpretation of data: Meyer. Drafting the article: Meyer, Wallace. Critically revising the article: Wallace, Cirivello, Cho. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Meyer. Administrative/technical/material support: Meyer. Study supervision: Cirivello, Cho.

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