Trigeminal neurinomas with extracranial extension: analysis of 28 surgically treated cases

Clinical article

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Object. The object of this paper was to review the authors’ experience with 28 cases of trigeminal neurinomas having an extracranial extension.

Methods. The authors analyzed 28 cases of trigeminal neurinoma in which there was an extracranial extension of the tumor. All patients were treated in their department between the years 1989 and 2009.

Results. There was tumor extension along the ophthalmic division of the nerve in 4 cases, along the maxillary division in 5, and along the mandibular division in 13. In 6 tumors there was diffuse extracranial extension and the exact extracranial division of nerve involvement could not be ascertained. In 10 cases, the tumor had a multicompartmental location—in the posterior fossa, the middle fossa, and the extracranial compartment. Tingling paraesthesiae, numbness, and diffuse pain in the distribution of the trigeminal nerve were common symptoms and were present in 90% of patients. The extracranial component had a well-defined perineural/meningeal membrane cover that was continuous with the middle fossa dura mater and isolated the tumor tissue from the adjoining critical structures. In 7 out of 10 cases, even the posterior fossa component of the tumor was entirely “interdural” (within the confines of the dura). The maximum dimension of the tumor was > 4 cm in 22 cases. A limited “transcranial” approach with (12 cases) or without (16 cases) zygomatic osteotomy was found suitable for resection of these tumors. In 4 cases a lateral orbitotomy was performed. Total tumor resection was performed in 20 cases and partial resection in 8. The duration of follow-up ranged from 6 months to 19 years. Two patients required additional surgery for symptomatic recurrence.

Conclusions. Extracranial extensions of trigeminal neurinomas have a well-defined meningeal covering. In most cases resection was performed via a minimally invasive cranial avenue (a “reverse skull base approach”). Radical resection was associated with an excellent long-term outcome. (DOI: 10.3171/2009.10.JNS091149)

Key Words • extracranial • infratemporal fossa • mandibular nerve • skull base • trigeminal neurinoma

Extracranial extension of trigeminal neurinomas along the division of the nerve is relatively uncommon but well described. Most of the available literature on the subject is in the form of isolated case reports.9,11–13,16,17,20 We have reviewed our experience with 28 cases in which there was extracranial extension of trigeminal neurinomas.

Methods

We have previously reported our experience with 73 cases of trigeminal neurinomas that were treated between 1989 and 2001.5 In that series, 7 cases were identified as having an extracranial extension. However, on review of cases in that series, we were able to identify an additional 5 cases of trigeminal neurinomas that had an extracranial extension. We retrospectively analyzed our current experience with 152 cases of trigeminal neurinomas treated between 1989 and 2009 and were able to identify 28 trigeminal neurinomas having an extracranial extension. Neurinomas located in the orbit and probably arising from branches of the CN V but having no middle fossa extension were excluded from the study.

Clinical Features

The clinical features of the cases are elaborated in Table 1. Tingling and/or numbness in the distribution of the trigeminal nerve was present in 25 cases. Vague facial pain was encountered in 13 cases. Patients had difficulty in mastication related to wasting of the temporalis and/or masseter muscles (24 cases) and related to a lump in the mouth (5 cases). In 7 cases, the main presenting symptom was progressive proptosis. Two patients had diffuse ear
pain. In 3 cases there were no symptoms related to the trigeminal nerve. In 11 cases 1 or more of the other CNs were affected (Table 1). One patient had associated neurofibromatosis. Two patients had contralateral hemiparesis at the time of tumor recurrence.

Radiological Features

The sizes of the tumors ranged from 27 to 78 mm in their maximum diameter (average 52 mm) (Figs. 1–6). Tumor extension into the orbit (Fig. 1) along the superior orbital fissure was seen in 4 cases, extension along the maxillary division of the nerve (Figs. 2 and 6) was seen in 5 cases, and extension along the mandibular division in 13 (Fig. 3). In 6 cases the tumor was identified in the infratemporal/pterygopalatine fossae, but the exact division of the nerve of origin could not be clearly identified (Figs. 4 and 5). All tumors had a middle fossa component. In 10 cases, the tumors were multicompartmental—that is, they extended into all 3 compartments (posterior fossa, middle fossa, and extracranial compartment). In 3 of these cases the extracranial component of the tumor was relatively small and appeared to be the middle fossa component bulging into the eroded foramina. One patient with neurofibromatosis had bilateral trigeminal neurinomas. The tumor on one side extended extracranially along the mandibular nerve, while on the contralateral side it was relatively small and was limited to the middle cranial fossa. In 4 cases, the tumors were distinctly firm and fibrous, and in 2 cases they were markedly vascular. In the rest of the cases, the tumors were characteristically soft and necrotic/cystic and only moderately vascular. In 14 cases, the tumors were necrotic and multicystic in nature (Figs. 4 and 5), and in 2 cases each of the multiple cysts had fluid levels (Fig. 5). Calcification within the confines of the tumor was encountered in 1 case. Despite the variations in the physical characteristics of the lesions, they were all well defined and demarcated by a perineural/ meningeal covering that separated them from the adjoining structures. Despite the very large sizes of the tumors in this series, none was found to have malignant features on histological examination.

Surgery

A basal temporal craniotomy was performed as previously described.3,5,6 Zygomatic osteotomy was performed in 12 cases, providing significant additional space and an inferiorly directed angle to the tumor. For tumors extending into the orbit, the zygomatic osteotomy was extended up to the frontal process of the zygoma and the temporal craniotomy was extended anteriorly. Lateral orbitotomy was performed in 4 cases, and the bone over the lateral aspect of superior orbital fissure was widely resected to expose both the orbital and middle fossa components of the tumor in the same surgical field. Depending on the nature and extensions of the tumor, the temporal craniotomy was extended further by drilling of the roots of the zygomatic arch, the roof of the mandibular condyle, and the roof of the external ear canal and by performing a partial mastoidectomy that extended up to the mastoid antrum.3,6 As the temporalis muscle and the other muscles of the infratemporal fossa were atrophied in most cases, their retraction was relatively straightforward. The temporalis muscle was retracted anteriorly, posteriorly, or inferiorly or split into 2 parts for exposure. Intraoperative lumbar drainage of CSF relaxed the brain, and its superior retraction provided approximately 1.5–2 cm additional space and 20° of additional angle for working on the tumor and manipulation of the instruments in the infratemporal/ pterygopalatine fossa. The middle fossa floor dura added safety for the temporal brain during retraction. The surgical strategy was to perform a small basal temporal craniotomy, resect the floor of the middle cranial fossa and expose both the middle cranial fossa and infratemporal fossa components of the tumor in the same field. The dura incised on the outer wall of the tumor, and the tumor was then debulked. Care was taken during debulking to avoid violating the meningeal/perineural layer, which formed a firm protective layer and prevented exposure of the carotid artery and CNs in the region. For multicompartmental tumors, after the resection of both the middle fossa and the extracranial components of the tumor, wide exposure of the part extending into the posterior cranial fossa was obtained. In 7 of 10 cases, the posterior fossa component of the tumor was found to be extradural and not subarachnoid. The presence of dura around the tumor anterior to the brainstem made the dissection in the region relatively safe. Intradural dissection and sectioning of the tentorium

### Table 1: Summary of demographic and clinical characteristics in 28 patients

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>No. of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>sex</td>
<td></td>
</tr>
<tr>
<td>M</td>
<td>16 (57)</td>
</tr>
<tr>
<td>F</td>
<td>12 (43)</td>
</tr>
<tr>
<td>age in yrs</td>
<td></td>
</tr>
<tr>
<td>1–10</td>
<td>0</td>
</tr>
<tr>
<td>11–20</td>
<td>2 (7)</td>
</tr>
<tr>
<td>21–30</td>
<td>10 (36)</td>
</tr>
<tr>
<td>31–40</td>
<td>8 (29)</td>
</tr>
<tr>
<td>41–50</td>
<td>4 (14)</td>
</tr>
<tr>
<td>51–60</td>
<td>4 (14)</td>
</tr>
<tr>
<td>Sx duration in mos</td>
<td></td>
</tr>
<tr>
<td>1–6</td>
<td>14 (50)</td>
</tr>
<tr>
<td>6–12</td>
<td>2 (7)</td>
</tr>
<tr>
<td>12–24</td>
<td>10 (36)</td>
</tr>
<tr>
<td>&gt;24</td>
<td>2 (7)</td>
</tr>
<tr>
<td>CN involvement</td>
<td></td>
</tr>
<tr>
<td>optic</td>
<td>4 (14)</td>
</tr>
<tr>
<td>trigeminal</td>
<td></td>
</tr>
<tr>
<td>sensory</td>
<td>25 (89)</td>
</tr>
<tr>
<td>motor</td>
<td>24 (86)</td>
</tr>
<tr>
<td>abducent</td>
<td>6 (21)</td>
</tr>
<tr>
<td>facial</td>
<td>4 (14)</td>
</tr>
<tr>
<td>hearing</td>
<td>4 (14)</td>
</tr>
<tr>
<td>lower CNs</td>
<td>2 (7)</td>
</tr>
</tbody>
</table>
Trigeminal neurinomas with extracranial extension

was performed in 5 cases either to resect or to confirm the resection of the posterior fossa component of the tumor. At least some fibers of the trigeminal nerve were saved in all cases.

Outcome

In 20 cases total tumor resection was performed and subtotal in 8. In 2 of these 8 cases, a part of the tumor was inadvertently left behind. In 2 others, the tumor was of a “plexiform” nature and radical excision was not considered necessary. There was no significant morbidity or any mortality following surgery. The duration of follow-up ranged from 6 months to 19 years (average 90 months). Four patients were lost to follow-up. In 4 cases the tumor recurred and 2 patients had to undergo additional surgery. In both cases, the resection was incomplete during the first operation and the tumors were larger than 6 cm in their maximum diameter. In the second operation in each of these cases, it was observed that the tumors freely breached the dural borders and entered into the subarachnoid space in the posterior cranial fossa. In both of the initial operations, the tumors were found to be clearly interdural in their entirety. In the subsequent operations, the tumors could only be partially resected. In the other 2 cases of recurrence, tumor regrowth after radical gross-total resection is being clinically and radiologically monitored. None of the patients received radiation treatment of any kind. All patients are leading active and productive lives. Although numbness and wasting persisted in all cases in which it was present preoperatively, in none of these cases did it actually worsen after surgery. One patient developed complete corneal opacity during the period of follow-up.

Discussion

Trigeminal neurinomas represent 0.2% of all intracranial tumors. Jefferson presented a classification scheme for trigeminal neurinomas that categorized these tumors according to their location. According to this classification, the 3 types are: middle fossa type (Type A); posterior fossa root type (Type B), in which the tumor is in front of the brainstem; and the dumbbell type, which has both middle and posterior fossa components (Type C). Yoshida and Kawase added a fourth type (Type D) to the classification to include tumors that extend into the extracranial compartment. Depending on the division of CN V involved, Type D tumors were further subclassified into orbital, infratemporal, and pterygopalatine fossa types. Extracranial trigeminal neurinomas are relatively rare and approximately 50 cases have been reported in the literature.

In terms of incidence, among neurinomas, trigeminal neurinomas are second only to acoustic neurinomas. Trigeminal neurinomas have characteristic clinical presenting features and radiological and anatomical characteristics. Even tumors with extracranial extension were seen to be unique in their clinical and physical characteristics.

Anatomical and Radiological Considerations

Trigeminal neurinomas with extracranial extension were in general significantly larger at the time of diagnosis than tumors limited to the cranium. In 22 cases, the tumors exceeded 40 mm in their largest dimension. Although such large tumors have been previously reported, they are distinctly uncommon. Due to the subtle nature of...
symptoms extending over long periods of time, and neglect of early symptoms probably due to illiteracy or ignorance in some of our cases, the majority of the tumors had become quite large before being diagnosed. Although not appropriately imaged in all our cases, presence of smooth erosion of the petrous apex and enlargement of the foramen ovale, foramen rotundum, or superior orbital fissure constituted important diagnostic features.

The middle fossa component of trigeminal neurinomas has been identified by us and by others to be “interdural” in location, meaning thereby that the entire tumor is located between the dural layers of the lateral wall of the cavernous sinus. The adjoining CNs, internal carotid artery, and the venous plexuses of the cavernous sinus are displaced by the tumor and its dural cover. It was observed that the extracranial component of the tumor is also confined within the membranous layer that was identified as meningeal/perineural and continuous with the dural sheaths of the middle cranial fossa. All tumors had a middle fossa component and extended at least up to the Meckel cave. The unique and hitherto unreported feature that was encountered in 7 of 10 such cases was that the posterior fossa component of the tumor was not subarachnoid in nature as has been generally recognized in “dumbbell”-shaped tumors, but had a well-defined dural cover. The dural cover of the posterior fossa component probably arose from the middle fossa dura that bulged posteriorly or from the dura of the petrous apex.

In contrast to the intracranial tumors, which are usually soft, necrotic, and only moderately vascular in character, tumors with extracranial extension had more varied physical characteristics. In 2 cases the tumors had multiple cysts, each cyst demonstrating fluid-fluid levels on MR images. Such a radiological feature has never been described in trigeminal neurinomas. Calcification was seen within the confines of the tumor in one case. We could not locate any report other than ours wherein a calcification was noted within a trigeminal neurinoma.

These tumors originate in a segment of the nerve, and the rest of the nerve is involved by displacement by the growing mass. On the basis of our experience we had earlier speculated that the site of origin of trigeminal neurinomas is somewhere in the region where the nerve enters the Meckel cave. Most trigeminal neurinomas irrespective of the site of spread have an association with this region of the nerve.

Clinical Features

The clinical presentation was commonly in the form of paresthesiae, usually in more than one division of the nerve. Severe or neuralgic pain was not encountered. The characteristic clinical features of numbness in the distribution of the nerve and wasting of the infratemporal fossa muscles distinguished these tumors from other tumors that occur in this location. Difficulty in chewing was the main symptom in 24 cases. Wasting of the temporalis and masseter muscles is also a characteristic feature and was present in 24 of the 28 cases. Proptosis was seen as a presenting symptom in 7 cases. In 5 cases, the patient’s main complaint was the feeling of a lump in the mouth. Such a symptom has not been reported earlier. One patient had associated neurofibromatosis.

**Fig. 3.** A: Coronal T1-weighted MR image obtained in a 42-year-old woman showing the trigeminal neurinoma extending into the infratemporal fossa along the mandibular division of the nerve. B: Sagittal T2-weighted MR image showing the tumor.

**Fig. 4.** A: Axial T1-weighted MR image obtained in a 30-year-old man showing a large trigeminal neurinoma extending up to the temporal convexity and into the orbit. Note the proptosis. B: Axial T2-weighted image showing the marked necrotic nature of the tumor.

**Fig. 5.** A: Axial T1-weighted MR image showing a large neurinoma extending into the infratemporal fossa and the orbit. B: Axial T2-weighted MR image showing the multiple fluid-fluid levels within the cysts of the tumor. C: Sagittal image showing the tumor and its extension into the infratemporal fossa and orbit.
Trigeminal neurinomas with extracranial extension

**Fig. 6.** A: Axial T1-weighted MR image obtained in a 54-year-old man showing a multicompartmental trigeminal neurinoma in the posterior fossa, middle fossa, and the extracranial compartment. B: Postoperative axial T2-weighted MR image showing the tumor resection cavity. C: Axial T1-weighted contrast-enhanced MR image obtained 6 years after surgery showing a massive recurrence.

**Surgery**

A transcranial approach was found to be suitable for resection of these tumors. The strategy of retraction of the brain to expose an extracranial tumor, although against the principles of skull base surgery, was seen to provide an easy, quick, and wide access to the entire tumor. The surgical strategy was to perform a small basal temporal craniotomy, resect the floor of the middle cranial fossa, and expose both the middle cranial fossa and infratemporal fossa in the same surgical field. Working within the confines of the dura avoided exposure of the carotid artery and the cavernous sinus. After the resection of both the middle fossa and the extracranial components of the tumor, wide access to the part extending into the posterior cranial fossa was possible through the exposure. Lateral and superior orientation of the dissection provided the possibility of exposure of the tumor, temporal brain, brainstem, and petrous carotid artery simultaneously in the surgical field allowing dissection of the tumor from these structures under direct vision and control. Moreover, these structures were closer in the surgical field with this approach than they would have been with most other available surgical approaches. In anterior transfacial and anterolateral approaches, such an elaborate and simultaneous exposure may not be possible. Trigeminal neurinomas do not involve all the fibers of the nerve. Some fibers are invariably spared, and these can usually be preserved. Working intratumorally, using blunt dissection with the help of suction or CUSA (cavitron ultrasonic suction aspirator), and avoiding coagulation as much as possible could prevent injury to these fibers.

**Recurrence**

Although there have been reports of malignant trigeminal neurinomas, none of the patients in our series had a malignant neurinoma. Loss of 4 patients (14%) to follow-up could bias the overall recurrence rate. However, the recurrence rate of 14% in the presented series is significantly higher than the overall recurrence rates of <2% that have been reported in the other published series of cases of trigeminal neurinomas. Although focused radiation therapy as a primary modality or as an adjuvant form of treatment has been suggested by some and found valuable particularly for initially small or residual lesions, such a treatment was not used in our series, even for cases in which there was recurrence or for residual tumor after a second surgery.

**Conclusions**

Trigeminal neurinomas with extracranial extensions have a well-defined perineural/meningeal cover that separates them from the adjoining nerves and vessels. A “reverse skull base approach” that involved a limited “transcranial” extradural avenue was used to resect the majority of tumors. Radical surgery is associated with an excellent clinical outcome and long-term tumor control.

**Disclaimer**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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