Neurilemomas of the trigeminal nerve

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Sixteen patients with trigeminal neurilemoma have been treated at the University Health Center of Pittsburgh during the last 15 years. Two patients had middle fossa tumors arising from the trigeminal ganglion, four had posterior fossa tumors arising from the trigeminal roots, six had “hourglass” lesions extending above and below the tentorium and involving the trigeminal ganglion and its roots, and four had tumors arising from the trigeminal branches and extending through the superior orbital fissure, foramen rotundum, or foramen ovale. In seven patients, tumor had also invaded the cavernous sinus. The clinical and radiographic features of these tumors, the operative approaches employed, and the postoperative outcome are discussed. Complete tumor excision was achieved in 12 patients; all 12 remain free of recurrence 3 to 157 months after surgery. In contrast, all four patients who underwent subtotal tumor excision showed progressive neurological deterioration from regrowth of residual tumor within 3 years of the initial surgery. Two of these four subsequently had total tumor excision and both are disease-free 23 and 34 months after the second procedure. Major morbidity developed in only one of the 16 patients. There were no operative deaths. Nine patients had preserved or improved trigeminal function after treatment.

Key Words • neurilemoma • brain neoplasm • trigeminal nerve • cavernous sinus

Neurilemomas of the trigeminal nerve account for 0.07% to 0.28% of intracranial tumors and 0.8% to 8% of intracranial neurilemomas. Because these tumors may originate in any section of the nerve between the root and the distal extracranial branches, they can produce a variety of symptoms and signs depending on the direction and extent of tumor growth. Neurilemomas arising from the trigeminal ganglion can remain localized in the middle fossa and present solely with symptoms of trigeminal nerve dysfunction; however, the majority of these lesions grow along the course of the nerve into the posterior fossa proximally or into the cavernous sinus and extracranial structures distally. Neurilemomas arising from the trigeminal root in the posterior fossa often remain strictly infratentorial and present as a cerebellopontine angle mass. Less commonly, these tumors originate from the intracranial branches of the trigeminal nerve and extend extracranially via the superior orbital fissure, foramen rotundum, or foramen ovale, manifesting as an orbital, paranasal sinus, or nasopharyngeal mass.

Between 1973 and 1988, 16 patients with trigeminal neurilemoma were treated at our institution. The clinical presentation, operative approaches employed, and outcome after treatment in these patients are the focus of this report.

Clinical Material and Methods

Composition of the Series

All patients with nerve-sheath tumors arising from the intracranial and/or intraorbital portions of the trigeminal nerve, who were treated during the last 15 years at the University Health Center of Pittsburgh, were included in the study. Two of these cases have been reported previously. The histological diagnosis in all cases was based on a review of the surgical specimens by a neuropathologist. Three of the 16 patients had evidence of neurofibromatosis at the time of diagnosis.

Clinical Features

The clinical characteristics of the 16 patients are delineated in Table 1. The most frequent initial symptoms were pain (four patients), diplopia (three patients), and hearing loss (three patients). By the time of diagnosis, seven patients had pain in a trigeminal distribution and 11 had trigeminal sensory loss. Weakness of the masticatory muscles was noted in only three patients. None of the affected patients had complete im-
### TABLE 1
Symptoms and signs in 16 patients with trigeminal neurilemoma*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Initial Symptom</th>
<th>Duration Before Diagnosis</th>
<th>Trigeminal Symptoms</th>
<th>Other Affected Cranial Nerves</th>
<th>Exophthalmos</th>
<th>Cerbellar/Pyramidal Tract Signs</th>
<th>Other Clinical Findings</th>
<th>Tumor Size &amp; Location†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1‡</td>
<td>31, F</td>
<td>lt hemifacial spasm</td>
<td>6 mos +</td>
<td>V&lt;sub&gt;1-3&lt;/sub&gt; -</td>
<td>VI-VIII -</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>3, P</td>
</tr>
<tr>
<td>2</td>
<td>42, F</td>
<td>diffuse head pain</td>
<td>5 yrs +</td>
<td>- -</td>
<td>-</td>
<td>VIII -</td>
<td>-</td>
<td>-</td>
<td>3, P</td>
</tr>
<tr>
<td>3</td>
<td>53, M</td>
<td>rt V&lt;sub&gt;3&lt;/sub&gt; pain</td>
<td>10 yrs +</td>
<td>V&lt;sub&gt;3&lt;/sub&gt; -</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>0.6, P</td>
</tr>
<tr>
<td>4</td>
<td>55, F</td>
<td>lt hearing loss</td>
<td>5 mos -</td>
<td>- -</td>
<td>VII, VIII -</td>
<td>cerebellar -</td>
<td>-</td>
<td>-</td>
<td>2, P</td>
</tr>
<tr>
<td>5</td>
<td>29, M</td>
<td>rt V&lt;sub&gt;1&lt;/sub&gt; pain</td>
<td>3 mos +</td>
<td>V&lt;sub&gt;1-3&lt;/sub&gt; +</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>2, M</td>
</tr>
<tr>
<td>6</td>
<td>41, M</td>
<td>lt V&lt;sub&gt;1&lt;/sub&gt; pain</td>
<td>2 mos +</td>
<td>V&lt;sub&gt;1-2&lt;/sub&gt; -</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>1, M</td>
</tr>
<tr>
<td>7</td>
<td>17, F</td>
<td>lt lateral diplopia</td>
<td>2 mos -</td>
<td>- -</td>
<td>VI +</td>
<td>pyramidal -</td>
<td>-</td>
<td>-</td>
<td>5, D</td>
</tr>
<tr>
<td>8‡</td>
<td>17, F</td>
<td>rt lateral diplopia</td>
<td>3 yrs +</td>
<td>V&lt;sub&gt;1-3&lt;/sub&gt; +</td>
<td>VI-VIII -</td>
<td>-</td>
<td>rt conductive hearing loss</td>
<td>-</td>
<td>5, D</td>
</tr>
<tr>
<td>9</td>
<td>31, M</td>
<td>rt V&lt;sub&gt;2,3&lt;/sub&gt; numbness</td>
<td>4 mos -</td>
<td>V&lt;sub&gt;2,3&lt;/sub&gt; -</td>
<td>VII-X -</td>
<td>both</td>
<td>papilledema, rt hand numbness</td>
<td>-</td>
<td>8, D</td>
</tr>
<tr>
<td>10</td>
<td>33, F</td>
<td>rt lat diplopia</td>
<td>2 wks -</td>
<td>- -</td>
<td>VI -</td>
<td>-</td>
<td>middle ear effusion</td>
<td>-</td>
<td>6, D</td>
</tr>
<tr>
<td>11</td>
<td>47, F</td>
<td>ataxia</td>
<td>2 yrs -</td>
<td>- -</td>
<td>VIII, IX -</td>
<td>cerebellar</td>
<td>dementia</td>
<td>-</td>
<td>7, D</td>
</tr>
<tr>
<td>12</td>
<td>66, F</td>
<td>rt hearing loss</td>
<td>3 mos +</td>
<td>V&lt;sub&gt;1-3&lt;/sub&gt; +</td>
<td>VII, VIII -</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>6, D</td>
</tr>
<tr>
<td>13‡</td>
<td>1, M</td>
<td>proptosis</td>
<td>11 mos -</td>
<td>V&lt;sub&gt;1&lt;/sub&gt; -</td>
<td>II-IV, VI +</td>
<td></td>
<td>papilledema</td>
<td>-</td>
<td>7, V&lt;sub&gt;1&lt;/sub&gt;</td>
</tr>
<tr>
<td>14</td>
<td>41, F</td>
<td>nasal obstruction</td>
<td>3 mos -</td>
<td>V&lt;sub&gt;2&lt;/sub&gt; -</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>8, V&lt;sub&gt;2&lt;/sub&gt;</td>
</tr>
<tr>
<td>15</td>
<td>58, F</td>
<td>rt hearing loss</td>
<td>1 yr -</td>
<td>V&lt;sub&gt;1&lt;/sub&gt; -</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>3, V&lt;sub&gt;3&lt;/sub&gt;</td>
</tr>
<tr>
<td>16</td>
<td>57, M</td>
<td>proptosis</td>
<td>2 yrs -</td>
<td>V&lt;sub&gt;1&lt;/sub&gt; -</td>
<td>II +</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>2, O</td>
</tr>
</tbody>
</table>

* Cranial nerves are given in roman numerals. + = feature present; - = feature absent.
† Maximum diameter is given in cm. Abbreviations: P = posterior fossa primarily; M = middle fossa primarily; D = dumbbell growth pattern above and below the tentorium; V<sub>1</sub> = V<sub>1</sub> tumor involving the cavernous sinus, middle fossa, and orbit; V<sub>2</sub> = V<sub>2</sub> neurilemoma extending from the cavernous sinus through the foramen rotundum into the pterygopalatine and infratemporal fossae, sphenoid sinus, and maxillary sinus; V<sub>3</sub> = V<sub>3</sub> neurilemoma extending from the middle fossa through the foramen ovale to the infratemporal fossa; O = extracranial V<sub>1</sub> tumor extending through the orbital apex.
‡ Patients with associated neurofibromatosis.

Correlation Between Symptoms and Tumor Location

Four patients (Cases 1 to 4) had comparatively small tumors (≤ 3 cm in diameter) that were entirely or nearly entirely localized to the posterior fossa. Two of these patients had both trigeminal pain and sensory loss at the time of diagnosis. Three of the four also had involvement of adjacent cranial nerves. Two patients (Cases 5 and 6) had small middle fossa tumors (≤ 2 cm in diameter) involving the area around Meckel’s cave. In both of these patients, trigeminal pain and sensory loss were the sole presenting symptoms. Six patients (Cases 7 to 12) had large tumors (≥ 5 cm diameter) with significant extension into both the posterior and middle fossae. Although three of these patients had no objective evidence of trigeminal dysfunction, all had impairment of adjacent cranial nerves. Three patients also had cerebellar and/or pyramidal tract findings.

Three patients (Cases 13 to 15) had neurilemomas arising from the intracranial trigeminal branches and extending extracranially. Another patient (Case 16) had a neurilemoma of the first division of the fifth nerve (V<sub>1</sub>) originating in the orbital apex and growing intracranially. Although all four patients had decreased sensation in the distribution of the trigeminal branch from which the tumor arose, the presenting complaint in each of these patients related to extracranial mass effect (in the orbit or nasopharynx). Both patients with V<sub>1</sub> neurilemomas presented with proptosis and visual loss, the patient with a second division (V<sub>2</sub>) lesion complained of nasal obstruction, and the patient with a third division (V<sub>3</sub>) neurilemoma presented with chronic serous otitis media and decreased hearing.

Imaging Features

The radiographic characteristics of these lesions are detailed in Table 2. The most common finding on skull roentgenography was erosion of the petrous apex, which was seen in seven patients. Three patients also had erosion of the posterior clinoid and/or dorsum sellae. A child with a large V<sub>1</sub> tumor had enlargement of the superior orbital fissure and optic foramen and erosion of the sphenoid wing. The patients with V<sub>2</sub> or V<sub>3</sub> neurilemoma both had extensive erosion of the middle fossa floor. Skull films were completely normal in five patients, and in another patient showed nonspecific enlargement of the sella turcica; these six patients all...
had relatively small lesions (≤ 3 cm in maximum diameter).

With the exception of a single patient (Case 16), who was treated before the advent of computerized tomography (CT), all patients underwent CT as a part of the initial evaluation. Typically, the lesions were hyperdense on unenhanced scans, with homogeneous enhancement after the administration of intravenous contrast material. Less commonly, the neurilemomas were isodense or of mixed density on unenhanced images, or enhanced in an irregular or ring-like fashion after the administration of intravenous contrast medium. In two patients treated in 1977 (Cases 1 and 3), small lesions were not visualized with a low-resolution CT scanner; however, pneumoencephalography correctly identified the size and location of the tumor.

Angiography was performed in 14 patients and typically demonstrated an avascular mass with displacement of surrounding vessels. In three patients, however, small feeding vessels were identified leading from the external and/or internal carotid arteries (ICA's) to the region of the tumor. Magnetic resonance (MR) imaging was performed in six patients and provided valuable delineation of the location and size of the tumor and the relationship of surrounding structures. Figure 1 illustrates the typical MR appearance of these lesions. In all six patients, the tumors were well circumscribed and, compared with surrounding brain, showed uniformly decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images.

During the last 3 years, a balloon occlusion test of the ipsilateral ICA has routinely been used in patients

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Temporal fossa, combined intracranial and extracranial tumor resection was performed by a surgical team extending into the orbit, pterygopalatine fossa, or infra-temporal fossa. Removal of the superior rim and lateral wall of the orbit with electrode placement and monitoring techniques have been described elsewhere. In another patient with recurrent tumor (Case 11) undergoing a preoperative balloon occlusion test; none of the five had evidence of compromised cerebral perfusion during the study. 

**Treatment**

Table 3 summarizes the operative management in this series. During all operations performed in the last 10 years, brain-stem auditory evoked potentials and somatosensory evoked potentials were monitored. When clinically indicated, third, fourth, sixth, and seventh cranial nerve function was monitored by electromyography of the extraocular and facial muscles. Electrode placement and monitoring techniques have been described elsewhere. In the five patients with tumors involving the cavernous sinus who had preoperative balloon occlusion testing, and in another patient with recurrent tumor (Case 13) who had not undergone a balloon occlusion study, proximal and distal control of the ICA was achieved before tumor excision from the sinus. Exposure of the cavernous sinus region was facilitated by removal of the petrous apex and extending into the posterior fossa and middle fossa. Removal of the zygomatic arch, in addition to a standard frontotemporal craniotomy with division of the tentorium. No attempt was made to remove additional tumor at a second procedure. Tumor excision was also incomplete in a young child with a large V2 neurilemoma involving the cavernous sinus, excision was also incomplete in a young child with a large V2 neurilemoma involving the cavernous sinus (Case 11); the bulk of the tumor was removed via a combined intra- and extradural frontotemporal approach. A 2-cm orbital apex V1 neurilemoma was completely removed by a subfrontal and lateral orbital approach (Case 16), and a 3-cm V3 neurilemoma was totally excised through a combined frontotemporal and infratemporal fossa approach (Case 15).

**Operative Results**

**Tumor Size and Extent of Initial Resection**

**Small Tumors.** Total excision of the tumor was achieved in all eight patients with small lesions (≤ 3 cm in diameter). All four posterior fossa neurilemomas were excised completely through a retromastoid paracerebellar approach. Both patients with small neurilemomas restricted to the middle fossa had total tumor excision via a combined intra- and extradural frontotemporal approach. A 2-cm orbital apex V1 neurilemoma was completely removed by a subfrontal and lateral orbital approach (Case 16), and a 3-cm V3 neurilemoma was totally excised through a combined frontotemporal and infratemporal fossa approach (Case 15).

**Large Tumors.** Total excision was achieved initially in only four of the eight patients with large neurilemomas. Among the six patients with large hourglass-shaped lesions (≥ 5 cm in diameter) extending into both the middle fossa and posterior fossa, only three had complete tumor resections initially: two of these patients underwent staged supra- and infratentorial procedures separated by a 3- to 6-week recovery period, and a third patient had total tumor excision via a frontotemporal craniotomy with division of the tentorium. A total removal of a large V2 neurilemoma also was achieved in two stages (Case 14); the bulk of the tumor was removed via a combined extradural frontotemporal and infratemporal fossa approach, and the remaining cavernous sinus component was removed through an intradural approach. In three patients with hourglass neurilemomas, 10% to 20% of the tumor was left behind after an attempted single-stage excision via either a frontotemporal (two patients) or retromastoid (one patient) approach with division of the tentorium. No attempt was made to remove additional tumor at a second procedure. Tumor excision was also incomplete in a young child with a large V1 neurilemoma involving the cavernous sinus, middle fossa, and posterior orbit (Case 13).

**Outcome**

Outcome after surgery correlated closely with the extent of tumor excision. All 12 patients who had initial complete tumor resection remained free of clinical or CT evidence of recurrent disease at 3 to 157 months after surgery (median 73 months). In contrast, within 3 years of initial surgery all four patients with subtotal tumor removal suffered symptomatic regrowth from residual tumors within the cavernous sinus. In all four of these patients, additional surgery was required because of progressive neurological deterioration. In two of them (Cases 11 and 13), residual tumor was
### TABLE 3
Treatment and outcome in 16 patients with trigeminal neurilemoma

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Origin</th>
<th>Tumor Location &amp; Growth Pattern</th>
<th>Size (cm)</th>
<th>Surgical Approach(es)</th>
<th>% Re- section</th>
<th>Complications</th>
<th>Outcome$</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>root</td>
<td>posterior &gt; middle fossa extension</td>
<td>3 x 2 x 1</td>
<td>retromastoid craniectomy</td>
<td>100</td>
<td>—</td>
<td>NED, 130 mos</td>
<td>—</td>
</tr>
<tr>
<td>2</td>
<td>root</td>
<td>posterior &gt; middle fossa extension</td>
<td>3 x 2 x 2</td>
<td>retromastoid craniectomy*</td>
<td>100</td>
<td>partial VI, VII nerve dysfunction</td>
<td>NED, 125 mos</td>
<td>—</td>
</tr>
<tr>
<td>3</td>
<td>root</td>
<td>posterior fossa</td>
<td>0.6 x 0.4 x 0.3</td>
<td>retromastoid craniectomy</td>
<td>100</td>
<td>—</td>
<td>died of MI with NED, 84 mos</td>
<td>—</td>
</tr>
<tr>
<td>4</td>
<td>root</td>
<td>posterior fossa</td>
<td>2 x 2 x 2</td>
<td>retromastoid craniectomy</td>
<td>100</td>
<td>—</td>
<td>NED, 111 mos</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>ganglion</td>
<td>middle fossa, cavernous sinus</td>
<td>2 x 2 x 2</td>
<td>frontotemporal craniotomy</td>
<td>100</td>
<td>—</td>
<td>NED, 8 mos</td>
<td>—</td>
</tr>
<tr>
<td>6</td>
<td>ganglion</td>
<td>expanding Meckel's cave</td>
<td>1 x 1 x 1</td>
<td>frontotemporal craniotomy</td>
<td>100</td>
<td>—</td>
<td>NED, 10 mos</td>
<td>—</td>
</tr>
<tr>
<td>7</td>
<td>ganglion</td>
<td>middle &amp; posterior fossa extension</td>
<td>5 x 4 x 3</td>
<td>1: frontotemporal craniotomy, transsylvian approach 2: retromastoid craniectomy† 3: frontotemporal craniotomy, subtemporal approach</td>
<td>100</td>
<td>—</td>
<td>NED, 150 mos</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>ganglion</td>
<td>middle &amp; posterior fossa extension</td>
<td>5 x 5 x 3</td>
<td>frontotemporal craniotomy</td>
<td>90</td>
<td>—</td>
<td>growth of posterior fossa tumor, retromastoid craniectomy 14 mos postop with subtotal excision, LTF 24 mos later</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>ganglion</td>
<td>posterior &gt; middle fossa extension, cavernous sinus</td>
<td>8 x 6 x 3</td>
<td>1: retromastoid craniectomy* 2: frontotemporal craniotomy, tentorium incised</td>
<td>100</td>
<td>—</td>
<td>NED, 24 mos</td>
<td>—</td>
</tr>
<tr>
<td>10</td>
<td>ganglion</td>
<td>middle &gt; posterior fossa extension</td>
<td>6 x 5 x 3</td>
<td>frontotemporal craniotomy, tentorium incised</td>
<td>100</td>
<td>—</td>
<td>NED, 65 mos</td>
<td>—</td>
</tr>
<tr>
<td>11</td>
<td>ganglion</td>
<td>middle &amp; posterior fossa extension with growth into cavernous sinus</td>
<td>7 x 6 x 3</td>
<td>retromastoid craniectomy</td>
<td>80</td>
<td>pneumonia, temporal contusion‡</td>
<td>regrowth 16 mos postop, 100% resection via fronto-temporal craniotomy; NED, 50 mos</td>
<td>—</td>
</tr>
<tr>
<td>12</td>
<td>ganglion</td>
<td>middle &amp; posterior fossa extension</td>
<td>6 x 6 x 3</td>
<td>frontotemporal craniotomy, tentorium incised</td>
<td>80</td>
<td>—</td>
<td>regrowth of tumor, repeat subtotal excision at 3, 5, &amp; 6 yrs; alive with disease, 7 yrs</td>
<td>—</td>
</tr>
<tr>
<td>13</td>
<td>V1 branch, intracranial cavernous sinus, middle &amp; anterior fossa extension</td>
<td>7 x 5 x 3</td>
<td>frontotemporal craniotomy, orbital decompression</td>
<td>80</td>
<td>cerebrospinal fluid leak‡</td>
<td>regrowth 16 mos postop, orbital exenteration with fronto-temporal craniotomy; NED, 39 mos</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>V1 branch, intracranial cavernous sinus, middle &amp; infratemporal fossae, nasopharynx</td>
<td>8 x 5 x 3</td>
<td>1: frontotemporal &amp; infratemporal fossa approach 2: frontotemporal &amp; infratemporal intradural approach</td>
<td>100</td>
<td>—</td>
<td>NED, 3 mos</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>V1 branch, intracranial middle &amp; infratemporal fossa cavernous sinus orbital apex</td>
<td>3 x 3 x 2</td>
<td>frontotemporal &amp; infratemporal fossa approach</td>
<td>100</td>
<td>—</td>
<td>NED, 3 mos</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>V1 branch, extracranial</td>
<td></td>
<td>2 x 1.5 x 1</td>
<td>lateral orbitotomy with frontal craniotomy</td>
<td>100</td>
<td>—</td>
<td>NED, 157 mos</td>
<td>—</td>
</tr>
</tbody>
</table>

* Ventriculoperitoneal shunt placed before initial surgical procedure.
† Preoperative embolization of external carotid feeders.
‡ Complications occurred after procedures for recurrent tumor.
§ Abbreviations: NED = no evidence of disease; MI = myocardial infarction; LTF = lost to follow-up. Postoperative duration is given.

totally removed at aggressive second procedures performed 16 months after the first operation. Both patients are now free of disease, 23 and 34 months after their second procedures. A third patient (Case 12), who has undergone four subtotal tumor resections, is alive with disease 7 years after initial diagnosis. The fourth patient (Case 8) was lost to follow-up evaluation 2 years after her second subtotal tumor excision. None of the 16 patients in this series received radiation therapy at any point during their course.
Postoperative Neurological Function

Fourteen of the 16 patients had decreased fifth nerve function immediately after surgery, seven of whom regained or surpassed their preoperative level of motor or sensory function within 6 months after surgery. In the remaining seven, one or more fascicles of the nerve were divided during tumor removal, and diminished facial sensation was permanent. Four patients had transient worsening of sixth (three patients), seventh (two patients), and eighth (two patients) cranial nerve function in the immediate postoperative period, which resolved within 6 months after surgery. In one additional patient (Case 2), partial impairment of sixth and seventh nerve function was permanent.

Complications

Two patients had complications after radical procedures for the removal of recurrent tumor. One patient with a large hourglass tumor (Case 11) had a temporal contusion and overlying subdural hematoma, which was evacuated 3 days postoperatively. She subsequently developed pneumonia, which resolved with treatment. In the patient with recurrent tumor in the cavernous sinus, anterior and middle fossae, and orbit (Case 13), a cerebrospinal fluid leak developed, but resolved with serial lumbar punctures and head elevation. There were no operative deaths. One patient died 7 years postoperatively after a massive myocardial infarction.

Discussion

The first description of an intracranial trigeminal neurilemoma was provided by Smith in 1849 in his monograph on neurilemomas.46 Since that time, approximately 250 cases have been reported. Patients in their fourth and fifth decades of life are most frequently affected; however, trigeminal neurilemomas have been reported in patients as old as 77 years and as young as 5 years.

Clinical Features

Because the fifth nerve has an extensive intracranial course and traverses several distinct anatomical regions, a number of varied modes of clinical presentation have been described, depending on the site at which the tumor arises and the direction and extent of growth.

Middle Fossa Neurilemomas. Fifty percent of all intracranial trigeminal neurilemomas arise from the trigeminal ganglion and remain predominantly localized in the middle fossa. Patients typically complain of pain and/or paresthesias in a trigeminal nerve distribution, which may spread from one to all three divisions of the nerve, often followed by progressive sensory loss and, less commonly, by atrophy of the masticatory muscles. Although loss of a corneal reflex is not uncommon, total trigeminal anesthesia and severe masticatory muscle wasting are rare and suggest a malignant process rather than a benign neurilemoma. Trigeminal neuralgia has been a fairly common presenting symptom. Lower cranial-nerve palsy and signs of increased intracranial pressure are also found in about 30% to 50% of patients. Pyramidal tract signs are seen in 30% to 40% of patients and frequently are ipsilateral, presumably resulting from compression of the cerebral peduncle against the contralateral tentorial edge by slowly growing tumor. Rarely, patients with infratentorial tumors have presented with headache of acute onset resulting from hemorrhage into the tumor or into the subarachnoid space.

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"Hourglass" Neurilemomas. Hourglass tumors arising from the trigeminal ganglion or distal roots account for 25% of all intracranial trigeminal neurilemomas and produce an often confusing combination of clinical findings, reflecting involvement of supratentorial and infratentorial structures. Long-tract signs are particularly common, owing to the presence of brain-stem compression at the tentorial hiatus. Six patients with hourglass neurilemomas were included in the present series. All six had involvement of one or more adjacent cranial nerves and three had associated cerebellar or pyramidal tract signs.

Neurilemomas Arising from Trigeminal Branches. In a small subgroup of patients, neurilemomas arise from the distal intracranial branches of the fifth nerve and extend extracranially, often exhibiting signs of extracranial mass effect. These lesions most commonly originate from the ophthalmic division of the nerve and present with proptosis and oculomotor palsies with or without visual loss. One such patient in the present series, a 1-year-old boy with neurofibromatosis and a large nerve-sheath tumor arising from V1, also had extensive involvement of the cavernous sinus and middle and anterior fossae, with extension of tumor into the orbit via the superior orbital fissure (Fig. 2). The paucity of reports of trigeminal neurilemomas arising from the maxillary and mandibular divisions is probably due to the shorter intracranial courses of these nerves; tumors of these branches are difficult to distinguish from those originating from the ganglion and growing distally along the nerve or those starting extracranially and penetrating the skull base. A single well-documented case of a mandibular-division neurilemoma growing down into the infratemporal fossa and up into the middle fossa has been described by Nager. In several other patients in whom the site of tumor origin was less definitively established, extracranial extension of an intracranial neurilemoma into the pterygopalatine fossa, sphenoid sinus, or nasopharynx has been reported. In our series, we encountered one case each of maxillary- and mandibular-division tumors that arose from the intracranial portion of the nerve and extended extracranially through a widely expanded foramen rotundum (Case 14, Fig. 3) or foramen ovale (Case 15). The former patient presented with nasal obstruction, and the latter with chronic serous otitis media and hearing loss from eustachian tube obstruction. Trigeminal sensory loss was mild in both patients, involving the V2 and V3 distributions, respectively.

Extracranial Neurilemomas with Intracranial Extension. Another small subgroup of patients have tumors that arise from the extracranial branches of the fifth nerve, progressively erode the skull base, and subsequently extend intracranially. This picture has been seen most commonly with orbital neurilemomas, which present with a combination of oculomotor paralysis, exophthalmos, and visual loss. One such case (Case 16) was included in the present series. Intracranial extension from neurilemomas arising in the infratemporal fossa has also been described.

Imaging Features

On plain skull radiography, neurilemomas of the middle fossa and hourglass lesions with significant middle fossa extension typically produce a sharply margined bone defect in the anteromedial portion of the petrous apex, which also may involve the floor of the middle fossa. Not uncommonly, the foramen ovale, foramen rotundum, and superior orbital fissure are smoothly enlarged to several times their normal size. Erosion of the sphenoid wing and anterior clinoid process, enlargement of the optic foramen, and erosion of the dorsum sellae, posterior clinoid process, and clivus are frequently seen with larger lesions. Infratentorial trigeminal neu-
nulelomomas may cause erosion of the inferomedial surface of the petrous bone; however, the internal acoustic meatus is rarely enlarged, which distinguishes these tumors from acoustic neurilemomas.17

The angiographic features of trigeminal neurilemomas have been well described elsewhere.3,8,14,30,37,38,49 These tumors generally appear as avascular mass lesions, although abnormal vascularity, represented by enlarged cavernous ICA or external carotid artery branches, is identified in about 25% of cases.8,14,30,38,49 A true angiographic blush is less common and was noted in only one patient in our series.

In recent years, high-resolution CT has been the diagnostic procedure of choice for demonstrating the location and extent of tumor growth. Typically, trigeminal neurilemomas are isodense or slightly hyperdense in comparison to surrounding brain and enhance homogeneously after administration of intravenous contrast medium.13,34,39 Although this CT appearance resembles that of a meningioma, bone erosion rather than hyperostosis is generally seen with a trigeminal neurilemoma. In addition, the absence of a pronounced tumor blush on angiography is more characteristic of a nerve-sheath tumor than a meningioma. A hypodense lesion with irregular or ring-shaped enhancement is less common, but was seen in two patients in the present series and has also been noted by other authors.34,47

Although experience with MR imaging of these tumors is limited,39 improved resolution in the region of the petrous bone and the ease of multiplanar reconstruction make this technique a helpful adjunctive study in evaluating and following patients. In all six of our patients who underwent MR imaging, neurilemomas had decreased signal intensity on T1-weighted images and increased signal intensity on T2-weighted images in comparison to surrounding brain.

**Treatment**

The first attempted excision of a gasserian tumor was performed by Krogius in 1895.22 Frazier10 reported the first successful complete excision of a gasserian ganglion tumor in 1918. Jefferson,18 who described the first sizable series of trigeminal neurilemomas, reported attempted excision in six of seven patients; two patients died perioperatively and one had recurrence of tumor. In reviewing the literature through 1957, Schisano and Oliverona41 found 39 operative cases, of which 16 (40%) died within 1 year of surgery. Their own results, however, were far superior: among their 19 treated patients, tumor removal was complete in eight, nearly complete in four, and partial in seven; they reported only one perioperative death and two recurrences. They recommended a supratentorial approach, even for patients with posterior fossa symptoms, and removed tumor via this route in 13 patients. They used a suboccipital approach in four patients and a combined supra- and infratentorial approach in an additional patient.41 In recent years, improvements in imaging techniques have led to earlier detection of trigeminal neurilemomas, while they are still relatively small and before they have caused significant neurological impairment. Favorable postoperative results have been reported by several authors.1,2,5,26,33

At present, the major impediments to complete tumor removal have been involvement of the cavernous sinus,4,6,33 adherence to crucial vascular structures, and inadequate exposure.41 In view of the high rate of symptomatic regrowth of tumor in patients with incompletely resected trigeminal neurilemomas, we have favored a more aggressive stance toward management of these tumors during the last several years. Even though the trigeminal nerve lies in the lateral wall of the cavernous sinus, a large trigeminal neurilemoma may actually extend into the cavernous sinus and must be dissected medially from the intracavernous ICA and the abducens nerve in order to achieve a total excision. With careful microsurgical technique, the tumor can be dissected from the wall of, or from within, the cavernous sinus with minimal morbidity. In six patients in the present series (including two with recurrent tumors), tumor invading the cavernous sinus was excised completely.

In patients with large hourglass neurilemomas extending above and below the tentorium, we believe that both supra- and infratentorial procedures, performed in either one or two stages, should be employed to achieve total tumor resection. With meticulous microsurgical dissection, trigeminal sensation and motor function often can be partially preserved. Trigeminal branch neurilemomas extending into the orbit, pterygopalatine fossa, and infratemporal fossa frequently require a combined intra- and extracranial approach performed by a surgical team consisting of a neurosurgeon working in conjunction with an otolaryngologist, an ophthalmologist, and/or a plastic surgeon.

In elderly patients and those with severe medical problems that would make a lengthy intracranial procedure unduly hazardous (for example, Case 12 in this series), intracapsular debulking of the tumor may provide several years of symptom-free survival.

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