The surgical management of trigeminal schwannomas
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Introduction
Schwannomas are tumors of the nerve sheath that usually exhibit benign behavior\(^1,5,8\). Benign trigeminal schwannoma of the trigeminal nerve comprises only 0.2% to 0.4% of all intracranial tumors and primarily arises in the gasserian ganglion\(^2,9,11,12\). Trigeminal schwannomas are benign tumors of schwann cell origin, are relatively rare and much less common than acoustic neuroma\(^5,8,12\). Most trigeminal neurinomas irrespective of the site of spread have an association with this region of the nerve\(^5\). The tumor grows larger and spreads in the available spaces\(^7\).

The Meckel's cave can accommodate a large amount of the tumor, which bloats up the cave\(^7\). The tumor being soft is unable to open up the dural sheath beyond the ganglion into the roots. This may be the reason that in most of the cases tumor does not extend beyond the dilated cave\(^3,7\). The tumor presses the adjacent normal fifth nerve, most of which is clinically involved by direct pressure of the tumor\(^7\). In the posterior fossa the trigeminal neurinomas are located intradurally\(^7\). The part in proximity to the brain stem is in most cases like any other extra-axial tumor with a well-defined plane of cleavage\(^7,9\). In general these tumors involve the adjoining cranial nerves, blood vessels and brain only by displacement and not by invasion\(^7,10\). We retrospectively analyzed the clinical profiles of 6 patients who were surgically treated for trigeminal schwannomas. The aim of this study was to analyze the presenting clinical and radiological features of these tumors, to establish factors that might affect surgical decision-making, to critically evaluate the appropriate surgical route, depending on tumor location and to assess the long-term outcomes after radical tumor resection.

Surgical anatomy
Recent advances in understanding the microsurgical anatomy of skull base structures are hallmarks of modern neurosurgery\(^6\). The trigeminal nerve has an extensive anatomic course\(^2,6\). Comprehensive knowledge of trigeminal nerve anatomy facilitates understanding of the relationship between the brainstem, skull base and facial area\(^2\). The trigeminal nerve trifurcates into ophthalmic, maxillary and mandibular nerves distal to the trigeminal ganglion\(^2\). The ophthalmic nerve passes forward in the lateral wall of the cavernous sinus. It gains access into the orbit via the superior orbital fissure. The ophthalmic nerve then divides to supply sensation to the eyeball, lacrimal glands, conjunctiva, part of the nasal mucosa, skin of the nose, eyelid and forehead\(^3,6\).

Classification of tumor extension
Trigeminal schwannomas may originate from the root, the ganglion or the peripheral branches of the trigeminal nerve\(^8\). Jefferson initially divided these tumors into 4 groups depending on their anatomical location: Posterior fossa (Root type), Combined posterior fossa-middle fossa (Dumbbell type), Middle fossa (Ganglion type), and Peripheral (Division type)\(^8\). Samii et al\(^13\) classified the tumor extension into 4 categories based on radiological findings: Type A- Intracranial tumor predominantly in the middle fossa; Type B- Intracranial tumor predominantly in the posterior fossa; Type C- Intracranial dumbbell-shaped tumor in the middle and posterior fossa; and Type D- Extracranial tumor with intracranial extensions\(^13\).
Case Report

Methods
This series includes 6 patients who were surgically treated between 2005 and 2008 at BSMMU. No patients received a diagnosis of NF2.

Results
Total tumor excision was possible in 4 patients, whereas total removal were not achieved in 2 patients. The extent of resection was graded according to the surgeons' impressions, confirmed by postoperative imaging in all patients. "Total" resection was defined as complete resection of the tumor and its capsule. "Radical subtotal" was assigned to the resection when tumor capsule fragments remained on vital structures. When the tumor capsule remained in the cavernous sinus or on the brain stem, the resection was graded as "Subtotal". Out of the six patients one undergone subtotal resection, two patient required a staged procedure with a large dumbbell type lesion. There were no operation related death or mortality.

Table 1: Results of surgical procedures and outcomes after surgery of patients with TSs

<table>
<thead>
<tr>
<th>Operative approaches</th>
<th>No. of patients(%)</th>
<th>Operative outcomes</th>
<th>No. of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retrosigmoid approach</td>
<td>3 (50%)</td>
<td>Total resection</td>
<td>4 (66.6%)</td>
</tr>
<tr>
<td>Subtemporal transtentorial</td>
<td>3 (50%)</td>
<td>Radical subtotal</td>
<td>1 (16.6%)</td>
</tr>
<tr>
<td>approach</td>
<td></td>
<td>Subtotal</td>
<td>1 (16.6%)</td>
</tr>
<tr>
<td>Subtemporal interdural</td>
<td>2 (33.3%)</td>
<td>Recurrence</td>
<td>1</td>
</tr>
<tr>
<td>approach</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fronto temporopolar extradural</td>
<td>1 (16.6%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

"Functional outcome was assigned a grade of 'excellent' if the patient returned to his or her preoperative employment and was living independently". It would be preferable to compare the real functional outcome (i.e., according to the functions of the fifth nerve and/or other neural structures of the neighboring areas). Hence, an excellent result is achieved when there are no new neurological deficits.

Discussion
Trigeminal neurinomas are relatively rare tumors and represent 0.2% of all intracranial tumors. Trigeminal neurinomas usually arise from the Schwann cells of the sensory root and can originate in any section of the fifth cranial nerve and correspondingly a variety of symptoms and signs may develop.

Clinical presentation
There was significant sex variation in our series, with the male/female ratio being 2:1. The age of presentation varied from 12 years to 55 years.

The clinical presentation was usually in the form of paresthesia or numbness, often in more than one division of the nerve. Severe or neuralgic pain was uncommon and was not observed in any cases. Wasting of the temporalis and pterygoid muscles were common and occurred in 66.6 % of cases. The corneal reflex was depressed or absent for all.
patients. The symptoms of involvement of adjacent cranial nerves in the cavernous sinus and in the cerebellopontine angle have been frequently reported in 2 cases. These symptoms probably because of the large sizes of the tumors encountered in this series.

The large tumor size was also responsible for the relatively infrequently encountered symptoms of increased intracranial pressure and ophthalmoscopically demonstrated papilledema, which were observed in 4 cases (66.6%). One patient demonstrated contralateral hemiparesis and pyramidal signs related to severe compression of the brainstem. The unusual symptom of pathological laughter was observed in one case of large, dumbbell-shaped tumors. The clinical features of slowly progressive symptoms and the predominant presence of trigeminal nerve-related symptoms of numbness and muscle wasting are usually diagnostic.

Table 2: Preoperative clinical symptoms in 6 patients with TSs

<table>
<thead>
<tr>
<th>Symptom</th>
<th>No. of Patients(%)</th>
<th>Symptom</th>
<th>No. of Patients(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trigeminal hypesthesia</td>
<td>6(100%)</td>
<td>Diplopia</td>
<td>2(33.3%)</td>
</tr>
<tr>
<td>Facial hypesthesia</td>
<td>4(66.6%)</td>
<td>Ataxia</td>
<td>2(33.3%)</td>
</tr>
<tr>
<td>Headache</td>
<td>3(50%)</td>
<td>Pathological laughter</td>
<td>1(16.6%)</td>
</tr>
<tr>
<td>Hearing symptoms</td>
<td>3(50%)</td>
<td>Hemiparesis &amp; increased ICP with papilledema</td>
<td>2(33.3%)</td>
</tr>
<tr>
<td>Seizure</td>
<td>Nil</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 3: Shows the distribution of cases

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age/sex</th>
<th>Presentation</th>
<th>Location</th>
<th>Name of operation</th>
<th>Complications</th>
<th>Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>28/F</td>
<td>Headache, facial hypoesthesia</td>
<td>Middle fossa</td>
<td>Fronto tempopolar extradural approach</td>
<td>No</td>
<td>Lost from follow up</td>
</tr>
<tr>
<td>2</td>
<td>55/M</td>
<td>Deafness, facial hypoesthesia, ataxia</td>
<td>Posterior fossa</td>
<td>Stages 1.Petrosigmoid approach 1.Subtemporal transtentorial</td>
<td>No</td>
<td>3 yrs no recurrence</td>
</tr>
</tbody>
</table>
Case Report

The postcontrast T1-weighted image (Figure 5) reveals a mass lesion in the left cerebellopontine angle extending anteriorly. The lesion is predominantly hyperintense on T2-weighted image (Figure 6), and enhances homogenously with CT of the skull base shows the grossly enlarged left foramen ovale.

The characteristic anatomical location, extent and signal characteristics make the diagnosis of trigeminal nerve schwannoma almost certain.

Although there have been reports of malignant trigeminal neuromas, none of the patients in our series had a malignant neuroma.

Several contemporary series have demonstrated no deaths or major surgical complications with radical removal of TSs. In a classic series of 44 patients reported by Dolenc, total resection was achieved in 100% of the patients, including 5 who had undergone incomplete resection elsewhere and underwent another surgery to excise the remainder of the tumor. The authors recommend an epidural approach to schwannomas originating in the fifth cranial nerve peripherally to the Gasserian ganglion and either an epidural-transdural or an epidural-transdural-transpetrous approach to lesions originating in the Gasserian ganglion or in the root of the fifth nerve.

Surgical strategy

Recent reports demonstrate a higher percentage of tumor resection, a low surgical morbidity rate and a lower rate of recurrence. Various reports have stressed the need for radical surgery, because total resection leads to tumor cure and the recurrence rate for cases with partial resection is relatively higher for trigeminal neuromas, compared with acoustic neuromas.

Konovalov et al, on the basis of a comparatively large experience, the authors demonstrate some important points for trigeminal neuroma surgery, as follows: 1) These tumors are usually well separated from the cavernous sinus and the carotid artery, which facilitates their radical removal. 2) Usually the tumor does not completely destroy the trigeminal nerve, and some of its fibers can be preserved (preservation of the first trigeminal nerve division, if possible, is especially important). 3) The basal extradural approach (which the authors call "interdural") is an effective way to reach and remove small and medium-size trigeminal tumors. There is a behavioral difference between neuromas and neurofibromas, which may infiltrate the nerve. Tumors may spread along the nerve division, far from the Meckel cavity and other approaches may be necessary for their removal. In every case, selection of an appropriate approach should be strictly individualized.

The major impediment to complete removal is inadequate exposure. Because of the location within the layers of the dura in the middle fossa, there was no specific need to achieve proximal control of the carotid artery, as would probably be necessary for some other lesions in this location. Under surgical technique, the authors state, "a temporal craniectomy (if needed) is performed to..."
obtain a flat viewing angle across the floor of the middle fossa." Naturally, it is always needed, because this provides better access to the parasellar region and does not necessitate any retraction of the brain. Another statement, "If necessary, the middle meningeal artery is ligated to increase exposure of the lateral middle fossa floor"3-5.

**Retrosigmoid approach**
This approach is performed by placing the patient in the dorsal (mastoid) position with the head turned to the opposite side and the ipsilateral shoulder elevated. A linear incision is placed 4 cm behind the external auditory canal. The asterion is exposed to determine the junction of the transverse and sigmoid sinuses. A craniectomy 4 cm in diameter is performed, with the superior and anterior margins bordering the transverse and sigmoid sinuses, respectively. The dura mater is opened parallel to the sigmoid sinus; CSF is drained from the cerebellomedullary cistern; and CNs VII-XI are identified. The tumor is thereby exposed near the tentorium margin. After intracapsular tumor debulking, microsurgical radical removal is accomplished3.

**Lateral basal subtemporal approach**
The skin incision extends from in front of the ear and travels superiorly and posteriorly. The branches of the facial nerve are saved by working deep to the fascial layers. The exposure is centered on the external ear canal. The temporalis muscle is elevated and displaced anteriorly. The basal extension of the exposure was achieved by resection of the roots of the zygomatic arch, roof of the external ear canal and superior third of the mastoid bone (Figure 7). The temporalis muscle was rotated anteriorly and was thus away from the field. The exposure was centered over the external ear canal in line with the petrous apex. The direction of the approach to the tumor was the shortest and perpendicular from the surface and avoided any neural or vascular manipulation. Inclusion of mastoidectomy in the exposure added the advantages of petrosal approach3-7.

**Subtemporal transtentorial approach**
This approach provides a limited view of the underside of the tumor and its relationship to the vessels and cranial nerves below. A better view of the tumor's relationship to cranial nerves VI, VII, and VIII, the vertebrobasilar system and the anteroinferior cerebellar artery is provided via the suboccipital portion of the combined petrosal approach3,5,7,14.

**Combined (retrosigmoid with subtemporal transtentorial approach)**
This is the combination of subtemporal and lateral suboccipital approach for dumbbell shaped schwannoma. This approach do not need drilling of petrosal bone and exposure of sigmoid sinus, hence less time consuming, disadvantage is cerebellar retraction3,5,7.

**Frontotemporal extradural temporopolar approach**
Lesions of the orbital-cavernous and ganglion types were approached via an extradural approach to the cavernous sinus. With the head in three-pin fixation, supine, rotated approximately 30 degrees, a pterional skin incision is made. The scalp and temporalis muscle are then reflected anteriorly in one layer. In selected cases, a two-layer scalp flap is fashioned to retract the temporalis muscle inferiorly and posteriorly. This maneuver provides a widened corridor to the anterior middle fossa, necessary in approaching larger tumors. A pterional craniotomy is performed, which typically measures 3 x 5 cm. A temporal craniectomy (if needed) is performed to obtain a flat viewing angle across the floor of the middle fossa. The dura is then elevated from the sphenoid ridge and
medial frontal fossa. Elevation of the dura of the middle fossa proceeds laterally, to the foramen spinosum. If necessary, the middle meningeal artery is ligated to increase exposure of the lateral middle fossa floor.2,3,5,7

Combined petrosal approach
Patients with lesions that involve both the cavernous sinus and posterior fossa, the so-called "dumbbell" type, underwent surgical resection via the combined petrosal approach. The patient is placed in the three-quarter lateral position. An "L" shaped craniotomy is made around the ear to expose temporal and retromastoid dura. A mastoidectomy is then performed, preserving the structures of the bony labyrinth. Extradurally, the petrous apex medial to the internal auditory canal is fenestrated to create a window via the middle fossa trajectory to the posterior fossa.3,7,14

Conclusion
On the basis of our limited experience, we believe that the best treatment for TSs is complete microsurgical removal of the lesion and that this treatment should be considered the gold standard therapeutic modality for the majority of cases. Hypesthesia, to some degree, is common after surgery, at least in the early postoperative period.

Bibliography