Presigmoid Transpetrosal Approach for the Treatment of a Large Trochlear Nerve Schwannoma

—Case Report—

Toru MATSUI, Eiharu MORIKAWA, Tadashi MORIMOTO, and Takao ASANO

Department of Neurosurgery, Saitama Medical Center/School, Kawagoe, Saitama

Abstract

A 61-year-old man presented with a rare, large trochlear nerve schwannoma manifesting as left-sided weakness and hypesthesia, bilateral bulbar pareses, and trochlear nerve paresis persisting for 3 months. T1-weighted magnetic resonance imaging with gadolinium revealed an intensely enhanced, well-circumscribed lesion with multicystic formation occupying the prepontine and interpeduncular cisterns and compressing the pons and midbrain with greater extension to the right. The mass was completely removed through the presigmoid transpetrosal approach with preservation of the posterior cerebral, superior cerebellar, and basilar arteries and their branches. Neuroradiological examination after 3 years demonstrated no recurrence. Enlargement of a tumor in the cisternal portion is inclined to involve and/or encase the adjacent major arteries and their branches. The presigmoid transpetrosal approach is one of the best surgical routes to remove a large trochlear nerve schwannoma safely and completely.

Key words: trochlear nerve schwannoma, presigmoid transpetrosal approach

Introduction

Trochlear nerve sheath tumors are uncommon, with only 15 cases reported. Intracranial schwannomas occur in a number of mixed and purely motor cranial nerves, especially in association with neurofibromatosis type 2. Total removal of a large tumor in the region associated with the trochlear nerve requires retraction of the temporal lobe to obtain an adequate operative field. Therefore, the use of a modern cranial base approach is thought to be preferable to a simple subtemporal approach. Previous cases involved a relatively mid-sized tumor, so the subtemporal approach was used to obtain total or subtotal removal. However, removal of a large tumor sometimes leads to unexpected postoperative neurological deficits, possibly due to the intraoperative compression of the temporal lobe.

Here, we present an extremely rare case of a large trochlear nerve schwannoma without neurofibromatosis treated by the presigmoid transpetrosal approach, which allowed complete removal.

Case Report

A 61-year-old man presented in December 1997 with left-sided weakness and hypesthesia, bilateral bulbar pareses, and trochlear nerve paresis which had persisted since September 1997. During the 4 weeks before his admission, he had had difficulty in swallowing with intake of liquid and needed support because of cerebellar ataxia. He had no family history of neurofibromatosis.

Examination found no cutaneous stigmata of neurofibromatosis. His visual fields were full. The pupils were equal and reacted directly and indirectly to light. He had diplopia on downward gaze to the right, suggesting right trochlear nerve paresis. Corneal reflexes were brisk bilaterally, and facial sensation was normal. The functions of the VIIth to XIIth cranial nerves were intact. He demonstrated mild left-sided weakness of the upper motor neuron type, with hyperreactive reflexes. There was no convincing sensory loss. T1-weighted magnetic resonance (MR) imaging with gadolinium revealed an intensely enhanced, well-circumscribed lesion with multicystic formation occupying the prepontine and interpeduncular cisterns. The midbrain and upper pons were compressed and shifted to the left by the...
large extension of the tumor, and the tumor was attached to the upper third clivus and petrous bone (Fig. 1). T2-weighted MR imaging clearly demonstrated involvement of the basilar artery (BA) and the neighboring major arteries and their branches (Fig. 1). Cerebral angiography demonstrated no tumor blush. The preoperative diagnosis was trigeminal neurinoma rather than petroclival meningioma. Therefore, we decided to approach this lesion via the presigmoid transpetrosal approach with the goal of removing bone to preserve the brain.18)

The patient was positioned for the right presigmoid transpetrosal approach. A U-shaped skin incision starting two fingerbreadths anterior to the external auditory canal (EAC) was carried out superiorly and then curved posteriorly three fingerbreadths superior to the EAC and curved again downward to the suboccipital area, two fingerbreadths posterior to the mastoid process. A temporo-occipito-suboccipital craniotomy with mastoidectomy was then performed. The mastoidectomy was designed preoperatively to preserve the vestibulocochlear apparatus, based on computed tomography (CT) (Fig. 2). Postoperative hearing loss should thus be avoided through rongeuring off the mastoid bone. Following the craniotomy, the presigmoid and temporal dura was incised, and then the superior petrosal sinus and tentorium cerebelli were coagulated and cut. Evacuation of cerebrospinal fluid from the ambient cistern was helpful to obtain a large space for surgery by mild retraction of the temporal lobe and cerebellum (Fig. 3). First of all, the arachnoid membrane surrounding the petrosal vein was incised to expose the trigeminal nerve. The trigeminal, facial, and acoustic nerves were easily identified and dissected from the tumor, but the branches of the superior cerebellar artery (SCA) were overlying the tumor. The distal portion of the trochlear nerve was identified on the inner aspect of the tumor. A large mass was located at the ambient portion of the trochlear nerve. Yellowish-green fluid was evacuated from the tumor but the lumen was multilobulated. Unexpectedly, the tumor was composed of parenchymatous as well as cystic components. The tumor was removed by piecemeal resection to preserve the small branches from the major arteries such as the BA, posterior cerebral artery (PCA), and SCA. A large tumor located in the cisternal region is likely to involve and/or encase vascular components, so careful attention was paid to preserving any vessels. Finally, complete excision of the tumor including part of the trochlear nerve was obtained. Subsequently, the oculomotor nerve and the abducens nerve were identified in the ventral region of the BA-PCA junction and in the ventral site of the SCA and the trigeminal root, respectively. These observations suggested that the tumor origi-
Fig. 3 Intraoperative photograph and sketch showing that minimal retraction of temporal lobe (Temp) and cerebellum (cerebelli) allowed exposure of the important anatomical details of the cerebellopontine angle such as the trochlear nerve (Tro), tumor (Tum), branches of the superior cerebellar artery (SCAb), petrosal vein (Pv), and the trigeminal nerve (Tri). The tumor clearly originated from the trochlear nerve. P: rongeured petrous bone covered with cotton plugs, R1 and R2: Sugita retractors, S: sucker.

Fig. 4 Postoperative T1-weighted magnetic resonance images with gadolinium (left) within a month after surgery revealing complete resection, and (right) at 3 years after surgery showing no recurrence of the tumor. The brainstem dislocated by the large mass has normalized within 3 years.

Discussion

Preoperative localization of intraaxial or extraaxial lesions located in the parapeduncular region was very difficult before the development of high-resolution CT and/or MR imaging, but now only small iso-intense lesions present problems. The present tumor appeared with the following characteristics of meningioma and neurinoma. The mass was iso-intense on T1- and T2-weighted MR images and brightly enhanced with gadolinium. An extremely peculiar case of cystic trochlear nerve neurinoma mimicked a brainstem tumor. Generally, trochlear nerve neurinoma is preoperatively identified as trigeminal neurinoma, meningioma, or epidermoid cyst. In almost all cases, direct surgery revealed trochlear nerve neurinoma.

Trochlear nerve paresis was present in eight of 16 reported cases, including this case, and five of these 16 cases manifested as involvement of other cranial nerves (Table 1). The unilateral combination of hemiparesis, cerebellar ataxia, and sensory disturbance in the presence of extraaxial mass at the tentorial notch is more strongly suggestive of trochlear nerve neurinoma. However, these symptoms remain silent until the tumor size exceeds more than 3–4 cm diameter. A trochlear nerve neurinoma of 1.5 cm diameter, although extremely rare, was found at autopsy.

No preoperative trochlear nerve involvement was present in at least 45% of previous cases. The most frequently used surgical approach was the subtem-
### Table 1  Previous cases of trochlear nerve schwannoma

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age/ Sex</th>
<th>n.IV sign</th>
<th>Other symptoms</th>
<th>Tumor site</th>
<th>Surgery</th>
<th>Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>King (1976)</td>
<td>55/F</td>
<td>+</td>
<td>rt hyperesthesia, n.V and n.VII</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>18 mos</td>
</tr>
<tr>
<td>2</td>
<td>Boggan et al. (1979)</td>
<td>32/F</td>
<td>+</td>
<td>lt hemiparesis, amenorrhea, n.V and n.VII</td>
<td>cisternal</td>
<td>subtemporal, and transtentorial</td>
<td>8 mos</td>
</tr>
<tr>
<td>3</td>
<td>Leunda et al. (1982)</td>
<td>54/M</td>
<td>+</td>
<td>headache</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>1 yr</td>
</tr>
<tr>
<td>4</td>
<td></td>
<td>16/F</td>
<td>+</td>
<td>headache</td>
<td>cisternal and cavernous</td>
<td>subtemporal and transtentorial</td>
<td>6 mos</td>
</tr>
<tr>
<td>5</td>
<td>Yamamoto et al. (1987)</td>
<td>37/F</td>
<td>+</td>
<td>increased ICP, temporal headache</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>5 yrs</td>
</tr>
<tr>
<td>6</td>
<td>Garen et al. (1987)</td>
<td>18/F</td>
<td>-</td>
<td>diplopia, n.III</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>?</td>
</tr>
<tr>
<td>7</td>
<td>Tokuriki et al. (1988)</td>
<td>43/M</td>
<td>-</td>
<td>lt numbness, gait disturbance, n.V and n.VII</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>1 mo</td>
</tr>
<tr>
<td>8</td>
<td>Maurice-Williams (1989)</td>
<td>56/M</td>
<td>-</td>
<td>rt hemiparesis, diplopia</td>
<td>cisternal</td>
<td>suboccipital</td>
<td>2 yrs</td>
</tr>
<tr>
<td>9</td>
<td>Celli et al. (1992)</td>
<td>51/M</td>
<td>+</td>
<td>lt hemiparesis, lt cerebellar ataxia</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>5 yrs</td>
</tr>
<tr>
<td>10</td>
<td>Jackowski et al. (1994)</td>
<td>26/F</td>
<td>+</td>
<td>lt facial numbness</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>6 mos</td>
</tr>
<tr>
<td>11</td>
<td>Abe et al. (1994)</td>
<td>60/M</td>
<td>-</td>
<td>lt hemiparesis</td>
<td>cisternal</td>
<td>suboccipital</td>
<td>?</td>
</tr>
<tr>
<td>12</td>
<td></td>
<td>57/M</td>
<td>-</td>
<td>lt facial numbness</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>?</td>
</tr>
<tr>
<td>13</td>
<td>Dolenc and Coscia (1996)</td>
<td>68/M</td>
<td>-</td>
<td>lt hemiparesis, diplopia (transient)</td>
<td>cisternal and cavernous</td>
<td>subtemporal and suboccipital</td>
<td>2 mos</td>
</tr>
<tr>
<td>14</td>
<td>Santoreneos et al. (1997)</td>
<td>35/F</td>
<td>-</td>
<td>lt hemiparesis, bil bulbar pareses, emotional disorder, n.VII</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>23 mos</td>
</tr>
<tr>
<td>15</td>
<td>Nadkarni and Goel (1999)</td>
<td>48/F</td>
<td>-</td>
<td>pathological laughter</td>
<td>cisternal</td>
<td>subtemporal and transtentorial</td>
<td>6 mos</td>
</tr>
<tr>
<td>16</td>
<td>Present case</td>
<td>60/M</td>
<td>+</td>
<td>lt hemiparesis, diplopia, dysphagia</td>
<td>cisternal</td>
<td>presigmoid and transpetrosal</td>
<td>2.5 yrs</td>
</tr>
</tbody>
</table>


The presigmoid transpetrosal approach combined with tentorial incision (Table 1), and the tumor was almost totally excised. However, postoperative complications included temporary oculomotor and abducens nerve pareses, temporary hemiparesis and expressive dysphasia due to intraoperative compression of the temporal lobe and pons, and/or subsequent contusion.

Even if long-term follow up indicated recovery of the preoperative deficit, modern cranial base surgery is preferable to the simple subtemporal approach, but the presigmoid transpetrosal approach has never been used, possibly due to the low incidence of trochlear nerve schwannoma. The preoperative diagnosis was large trigeminal nerve schwannoma, so the presigmoid transpetrosal approach was performed in the present case. At 2 weeks after surgery, dysphagia and left-sided weakness had completely disappeared. The presigmoid transpetrosal approach provides excellent exposure of the cerebellopontine angle and pons, so removal of the tumor compressing the pons is unlikely to cause neurological deterioration, if the plane between the tumor and the pons is successfully dissected. To remove bone and preserve brain function, cranial base surgery technique is essential, and the total removal of such a large trochlear nerve schwannoma located in the cisternal portion is a good indicator of the effectiveness of this approach.

Oculomotor, trochlear, and abducens nerve schwannomas generally arise far from the glial Schwann sheath junction, at not more than 1 mm from the neuraxis. Trochlear nerve schwannomas arise almost exclusively from the cisternal segment. Therefore, the tumor grows in the space occupied by...
the PCA, SCA, BA, and perforators. Trochlear nerve schwannoma in the basal cistern is likely to form severe adherences to these anatomical structures, making total removal more difficult.

We started to treat large lesions located in the petroclival region employing the presigmoid transpetrosal approach in 1988 according to Hakuba et al. Acoustic neurinoma could be basically removed via a lateral suboccipital route, but such tumor with severe upward compression of the tentorium cerebelli and extension into the petroclival region was treated via the presigmoidal route. Postoperatively, MR imaging was repeated to detect recurrence of the tumor every one year, and MR angiography was performed to investigate the patency of the lateral and sigmoid sinuses, due to development of the compensatory venous drainage system following gradual obstruction of the sinuses. Thus, this approach is thought to be safe.

The possibility of total removal depends upon the severity of adhesion of the tumor to the brainstem and/or important vessels in the deep brain region, and can be well predicted by preoperative MR imaging, as the presence of thin low-intense areas on T1-weighted MR imaging, as the presence of thin low-intense areas on T1-weighted MR imaging, suggesting cerebrospinal fluid collection, between the tumor and brainstem.

Furthermore, T2-weighted MR imaging shows the presence of thin low-intense areas on T1-weighted MR imaging, suggesting cerebrospinal fluid collection, between the tumor and brainstem. Postoperatively, MR imaging was repeated to detect recurrence of the tumor every one year, and MR angiography was performed to investigate the patency of the lateral and sigmoid sinuses, due to development of the compensatory venous drainage system following gradual obstruction of the sinuses. Thus, this approach is thought to be safe.

The present case indicates that the presigmoid transpetrosal approach is the first choice to treat a patient with a large trochlear nerve schwannoma located in the ambient, parapeduncular, interpeduncular, or prepontine cistern.

References


Address reprint requests to: T. Matsui, M.D., Department of Neurosurgery, Saitama Medical Center/School, 1981 Kamoda, Kawagoe, Saitama 350–8550, Japan. e-mail: matsui@ns2.saitama-med.ac.jp.