Intramedullary Hemangioblastoma
of the Medulla Oblongata
—Two Case Reports and Review of the Literature—

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Abstract

We determined the treatment modality of hemangioblastoma of the brain stem on the bases of our two cases and 31 cases searching from the literature since 1960 which were treated surgically. Hemangioblastomas of the brain stem were categorized according to one of three locations: hemangioblastoma of the fourth ventricle attached to the floor of the ventricle (Type A), hemangioblastoma of the fourth ventricle partially embedded in the floor of the ventricle (Type E), and intramedullary hemangioblastoma of the medulla oblongata (Type I); and were evaluated their clinical features including the operative mortality and morbidity of each location. In our two cases of Type I hemangioblastoma, Case 1, removed partly, died due to sleep apnea and Case 2 lead to normal school life after hemangioblastomas were removed radically. In our review of the 33 surgically-treated cases, radical excision was carried out in 29 cases (87.9%). The mortality was 24.2% overall; that of Type A was 25%, Type E was 28.6%, and Type I was 14.3%. In terms of postoperative mortality, the location of the hemangioblastoma was irrelevant and radical excision was much better than partial removal. Hemangioblastomas of the brain stem could be removed radically by meticulous dissection of the tumor on distinct cleavage, even in cases of intramedullary location. Microsurgical dissection of medullary hemangioblastomas with low morbidity is feasible and prudent postoperative care is mandatory to reduce the operative mortality and morbidity.

Key words: hemangioblastoma, medulla oblongata, intramedullary location, fourth ventricle, total removal, surgical result

Introduction

Hemangioblastoma of the brain stem may be located in the fourth ventricle or the brain stem. These two locations may be distinguished by the operative risk and the functional quality of the clinical outcome, but whether surgical procedures and the results vary significantly with site remains unclear. Present microsurgical techniques can achieve total removal in most cases, but prudent postoperative care is mandatory due to the likelihood of respiratory and circulatory disorders, and bulbar palsy due to damage to the respiratory and vasomotor centers such as the vagal nuclei and tractus solitarius. We describe two cases of hemangioblastoma of the brain stem located in the medulla oblongata, one resulting in death due to sleep apnea and one in return to normal life.

Case Reports

Case 1: A 33-year-old male was admitted in October 1987, with headache, nausea, hiccups, anorexia, dysphagia, staggering gait, and profound weight loss, which had all progressed over the preceding 17 months. Neurological examination disclosed advanced papilledema, horizontal nystagmus on the right, left lateral gazes, absent gag reflex, dysphagia, ataxic gait, unsteadiness when standing on one foot, and motor incoordination of the upper and lower extremities.

Magnetic resonance (MR) imaging demonstrated a mixed-intensity mass occupying the caudal part of
the fourth ventricle and the dorsal medulla, associated with a cystic component at the caudal portion and displacing the inferior vermis posteriorly and superiorly (Fig. 1). T2-weighted MR imaging showed many linear flow voids in the parenchymatous portion. Vertebral angiography revealed a prominent retromedullary tumor stain, which is typical of a hemangioblastoma of the dorsal medulla.

Ventricular drainage was carried out through the posterior horn of the right lateral ventricle with the patient in the prone position. Suboccipital craniectomy and resection of the posterior arch of the atlas were performed. After opening the dura, dilated abnormal red veins appeared on the dorsal surface of the medulla oblongata, and the dorsal medulla protruded conspicuously, obstructing the foramen of Magendie and suggesting an intramedullary tumor. After incising the inferior vermis slightly, a cherry red, highly vascular mass was found embedded in the dorsal medulla, identifiable through the thin brain tissue. Under the operating microscope, the tumor was dissected from the brain stem and was gently retracted laterally. The ventral feeding arteries were identified, coagulated with bipolar forceps, and divided. A plane of cleavage was formed by gliotic interspace between the tumor and the brain. The tumor was gently rolled back and forth, progressively interrupting its blood supply and shrinking the size by half, and the fourth ventricle was opened. However, the latter bled profusely, with a blood loss of about 1000 ml. Because of the amount of lost blood and time, the procedure was abandoned and a second surgery was planned.

Postoperatively, he regained full consciousness without respiratory distress or dysarthria. His vital signs were stable, so he was extubated and brought back to the recovery room. Although he was cooperative and responded well, 2 hours later he was found to be apneic and in cardiac arrest, probably due to a sleep apnea attack. The patient was resuscitated, but he expired 2 months later due to repeated pneumonia.

**Case 2:** A 16-year-old school girl was admitted with vomiting, loss of appetite, imbalance, occipitalgia, and staggering gait persisting for 18 months. Neurological examination showed early papilledema, difficulty in tandem gait, and unsteadiness when standing on the left foot alone. Computed tomography showed hydrocephalus, so ventriculoperitoneal shunting was undertaken immediately.

T1-weighted MR imaging revealed a large isointense mass lesion in the dorsal medulla, associated with a caudal low intensity area and displacing the vermis superiorly and both tonsils posterolaterally (Fig. 2). T2-weighted MR imaging revealed a cord-like flow void sign in the hyperintense mass lesion, and except for the cystic component at the caudal

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**Fig. 1 Case 1.** T1-weighted magnetic resonance images showing a tumor located in the dorsal medulla and protruding into the fourth ventricle. The caudal medulla was displaced inferiorly and dorsally and a cyst is present at the basal pole of the mass.

**Fig. 2 Case 2.** T1-weighted magnetic resonance images showing the tumor located at the dorsal medulla, protruding into the fourth ventricle, and associated with a cyst at the basal pole of the mass and a syringomyelic cavity below C-2 (left column), and intense enhancement of the mass and cord-like flow voids in the mass after gadolinium administration, with another enhanced mass in the spinal cord at the C-2 level (right column).
part, intense enhancement by gadolinium. Another small enhanced mass lesion was disclosed at the C-2 level and was associated with syringomyelia below the mass. Vertebral angiography showed dense tumor staining, fed from the third and fourth segments of the vertebral arteries and the supra- and retrotonsillar segments of the posterior inferior cerebellar arteries. The vein of the lateral recess of the fourth ventricle drained into the petrosal vein and superior petrosal sinus, which served as the draining veins of the tumor. Staining revealed another tumor, fed from the posterior spinal artery on the right, located at the C-2 level. These findings are typical of hemangioblastoma of the dorsal medulla oblongata and spinal cord, and associated syringomyelia.

Suboccipital craniectomy and resection of the C-1 posterior arch were performed in October 1994. After opening the dura, dilated red veins were located on the dorsal medulla, the inferior vermis was displaced upwardly, and both tonsils were displaced dorsolaterally. The caudal floor of the fourth ventricle had protruded, and a cherry red, vascular-rich mass was seen through the thin dorsal medulla, suggesting an intramedullary mass (Fig. 3). Under the operating microscope, a midline incision was made in the dorsal medulla, and the tumor was dissected from the brain stem. There was a delicate gliotic cleavage between the tumor and the surrounding brain tissue. The tumor was gently rolled back, forth to coagulate, and separate the feeding arteries. The tumor shrank and was removed totally en bloc. Venous oozing from the tumor bed was controlled by oxycel and cottonoid packing.

Postoperatively she became fully conscious but developed bulbar palsy and hypoventilation. Assisted ventilation was required for 4 days, followed by tracheostomy. Gastrointestinal bleeding was treated with medication. One and a half months later the patient's dysphagia began to diminish, and 3 months later she could eat and speak normally. On March 7, 1995, a hemangioblastoma of the upper cervical cord was removed totally without any additional neurological deficit. The patient returned to normal school life in April 1995.

Review of the Literature

We reviewed 33 surgically-treated cases of hemangioblastoma of the brain stem, 31 collected from the literature available to us and the two cases described above (Table 1A-C).1-3,6,8-10,12,14-16,18,23,26-33) We categorized them by tumor location: 12 cases (36.4%) of hemangioblastoma of the fourth ventricle attached to the floor of the ventricle (Type A), 14 cases (42.4%) of hemangioblastoma partially embedded in the floor of the ventricle (Type E), and seven cases (21.2%) of intramedullary hemangioblastoma of the medulla oblongata (Type I). Cases of hemangioblastoma of the fourth ventricle were more frequent than cases of intramedullary hemangioblastoma: 79% of the total cases involved the fourth ventricle. These tumors were most frequently located in the dorsal medulla oblongata, especially in the area postrema.2'9'14 Hemangioblastoma appeared at varying patient ages, from 15 years to 68 years; average age was 33 years, with the greatest frequency in the 3rd and 4th decades. In 55% of the cases the patients were males.

The frequency of symptoms stemming from intracranial hypertension (headache, nausea, and vomiting) were 83%, 50%, and 86% for Type A, E, and I hemangioblastomas, respectively. Eighty-three percent of the patients with Type A hemangioblastoma had unsteady gait, and 33% had numbness of the extremities. Thirty-six percent of the patients with Type E hemangioblastoma had ataxia, and 29% had diplopia. In the patients with Type I hemangioblastoma, 86% showed ataxic gait and/or incoordination, and 43% had dysphagia and/or dysarthria. All these symptoms were more frequent in the Type I group than in the others. The mean durations of the preoperative symptoms for the Type A, E, and I groups were 8.8, 20.5, and 9 months, respectively.

In neurological examinations, cerebellar signs were present most frequently, followed by papilledema in 42%, 29%, and 50% in Type A, E, and I cases, respectively. Bulbar signs appeared most often in Type I (67%). The tumor was solid in 87% of all cases reviewed and was associated with small cysts or

![Fig. 3 Case 2. Intraoperative photograph of an intramedullary tumor showing bulging of the dorsal medulla and the discolored mass just below the vermis and the left tonsil, visible through thin brain tissue.](image-url)
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author [Year]</th>
<th>Age/ Sex</th>
<th>Tumor location</th>
<th>Tumor size</th>
<th>Preoperative symptoms</th>
<th>Duration</th>
<th>Preoperative neurological findings</th>
<th>Extirpation</th>
<th>Outcome and comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Archer et al. (1972)²⁵</td>
<td>33/F</td>
<td>fourth ventricle</td>
<td>solid: 15 mm (cyst)</td>
<td>headache, nausea, vomiting, unsteady gait</td>
<td>1 mo</td>
<td>nystagmus; IX, XII cr n palsy; ataxic gait</td>
<td>total</td>
<td>uneventful; VHL disease</td>
</tr>
<tr>
<td>2</td>
<td>Nishimoto and Kawakami (1980)²⁶</td>
<td>31/M</td>
<td>fourth ventricle</td>
<td>solid: 35 × 40 × 25 mm</td>
<td>fatigue, headache, tinnitus, tendency to veer to the left</td>
<td>1 yr</td>
<td>papilledema; cerebellar sign</td>
<td>total</td>
<td>unstable vital signs; trachceostomy; GI bleeding; died 3 mos after surgery</td>
</tr>
<tr>
<td>3</td>
<td></td>
<td>26/M</td>
<td>fourth ventricle</td>
<td>solid: 20 × 15 × 30 mm</td>
<td>dysphagia, headache, dysarthria, gait disturbance, numbness of rt lower leg</td>
<td>3 yrs</td>
<td>IX, X, XII cr n palsy; hypesthesia of rt lower leg; ataxia</td>
<td>total</td>
<td>unstable vital signs; respiratory distress; trachceostomy; paralytic ileus; improved and working 4 yrs and 1 mo after surgery</td>
</tr>
<tr>
<td>4</td>
<td></td>
<td>21/F</td>
<td>fourth ventricle</td>
<td>solid: 30 × 30 × 40 mm</td>
<td>headache, nausea, vomiting, urinary incontinence, convulsion, somnolence</td>
<td>2-3 mos</td>
<td>papilledema; ventricular drainage, total</td>
<td></td>
<td>abducens nerve palsy; bII nystagmus; slight nystagmus; mild ataxic gait 6 mos after surgery</td>
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<tr>
<td>5</td>
<td>Tognetti et al. (1986)²⁸</td>
<td>68/M</td>
<td>fourth ventricle</td>
<td>solid: 30 mm</td>
<td>alternating sensory loss, unstable gait, hiccup</td>
<td>3 mos</td>
<td>loss of deep sensation; upper extremity dysmetria; ataxia</td>
<td>total</td>
<td>progressive coma; died 8 days after surgery</td>
</tr>
<tr>
<td>6</td>
<td>Sanford and Smith (1986)²⁶</td>
<td>15/F</td>
<td>fourth ventricle</td>
<td>solid</td>
<td>headache, imbalance</td>
<td>3 mos</td>
<td>papilledema; truncal and extremity ataxia</td>
<td>total</td>
<td>hypertension; cerebellar signs; pneumonia; neurogenic hypertension remained after 5 mos</td>
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<tr>
<td>7</td>
<td></td>
<td>28/F</td>
<td>fourth ventricle</td>
<td>solid 30 × 40 mm</td>
<td>nausea, vomiting, headache, loss of weight, gait disturbance</td>
<td>6 mos</td>
<td>somnolence; extremity and truncal ataxia</td>
<td>VP shunt, total</td>
<td>hypoventilation while sleeping; hypertension; ataxia explored 11 mos postoperatively; VP shunt; removal of a 3 mm residual tumor; requires a walker and unable to perform household tasks</td>
</tr>
<tr>
<td>8</td>
<td>Tobiyama et al. (1990)²⁹</td>
<td>51/M</td>
<td>fourth ventricle</td>
<td>solid: 30 × 30 mm (cyst)</td>
<td>headache, unsteady gait</td>
<td>truncal ataxia</td>
<td>total</td>
<td>uneventful; died 1 yr 2 mos after surgery due to putaminal hemorrhage (metastasis)</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Yoshino et al. (1990)³¹</td>
<td>34/F</td>
<td>fourth ventricle</td>
<td>solid: 40 × 35 × 15 mm (cyst)</td>
<td>anorexia, weight loss, unsteadiness, vertigo, gait disturbance, headache</td>
<td>1 yr 6 mos</td>
<td>rt hemiparesis; hyperreflexia; wide-based gait; tremor</td>
<td>VP shunt, total</td>
<td>Lt hemiparesis; Lt homisensory loss; dysphagia; trachceostomy; GI bleeding; improved; lead a home life with slight dyssarthria and Lt hemihypesthesia</td>
</tr>
<tr>
<td>Case No.</td>
<td>Author (Year)</td>
<td>Age/ Sex</td>
<td>Tumor location</td>
<td>Tumor size</td>
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<td>10</td>
<td>Kuwahara et al. (1991)</td>
<td>25/F</td>
<td>fourth ventricle</td>
<td>solid</td>
<td>headache, nausea, ear obstruction, diplopia</td>
<td>7 mos</td>
<td>papilledema; VIII cr n palsy; bil nystagmus; truncal ataxia; nuchal rigidity papilledema</td>
<td>total (2-stage)</td>
<td>tachycardia; hearing difficulty; truncal ataxia; improved</td>
</tr>
<tr>
<td>11</td>
<td>Ehrenpreis et al. (1994)</td>
<td>35/M</td>
<td>fourth ventricle</td>
<td>solid (cyst)</td>
<td>decreased vision, occipitalgia, dizziness, rt hemiparesis, gait disturbance</td>
<td>4 mos</td>
<td>total</td>
<td>facial nerve palsy; diminished gag reflex; bil internuclear ophthalmoplegia; tracheostomy; died 4 wks after surgery improved</td>
<td></td>
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<tr>
<td>12</td>
<td>Masuda and Fujikawa (1996)</td>
<td>47/M</td>
<td>dorsal surface of the medulla, lt</td>
<td>cyst: 15 × 20 mm (solid mass: 6 mm)</td>
<td>numbness of lt upper extremity, dysarthria, unsteady gait, hiccup</td>
<td>4 mos</td>
<td>decreased vibration of occipital area and upper extremity, lt</td>
<td>total</td>
<td></td>
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</table>


**Table 1B Clinical summary of hemangioblastoma of the brain stem, embedded in medulla oblongata**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age/ Sex</th>
<th>Tumor location</th>
<th>Tumor size</th>
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<th>Extirpation</th>
<th>Outcome and comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>13</td>
<td>Archer et al. (1972)</td>
<td>28/F</td>
<td>medulla</td>
<td>solid (many cysts)</td>
<td>neck pain</td>
<td>3 yrs 6 mos</td>
<td>normal papilledema; disconjugate gaze; unsteady gait nystagmus;</td>
<td>cyst: drained</td>
<td>uneventful; VHL disease</td>
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<tr>
<td>14</td>
<td></td>
<td>23/F</td>
<td>medulla, lt cbl</td>
<td>large cyst (solid: 15 mm)</td>
<td>headache, ataxia, disconjugate gaze</td>
<td>2 yrs</td>
<td>total</td>
<td>uneventful; sister of patient 1</td>
<td></td>
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<tr>
<td>15</td>
<td>Yasargil et al. (1976)</td>
<td>27/M</td>
<td>medulla-cervical cord</td>
<td>occultal and nuchal pain, vertigo, ataxia, blurred vision</td>
<td>6 mos</td>
<td>total</td>
<td>improved; died 1.8 yrs after surgery due to bil IX, X, XI cr n palsy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>16</td>
<td></td>
<td>44/F</td>
<td>medulla-cervical cord</td>
<td>solid: plum sized (cyst)</td>
<td>headache</td>
<td>5 yrs</td>
<td>total</td>
<td>improved and working 2 yrs after surgery</td>
<td></td>
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<tr>
<td>17</td>
<td>Chou et al. (1975)</td>
<td>18/F</td>
<td>brain stem</td>
<td>solid: 20 mm</td>
<td>intracranial hypertension</td>
<td></td>
<td>total</td>
<td>nystagmus; unsteady gait; normal 5 yrs after surgery; family history dysmetria; normal 3 mos after surgery; cervical cord lesion</td>
<td></td>
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<tr>
<td>18</td>
<td>Rawe et al. (1978)</td>
<td>17/M</td>
<td>brain stem, cbl, spinal cord</td>
<td>cyst 25 mm (solid: 10 mm)</td>
<td>transient visual obscuration, headache, lt incoordination, lt perioral paresthesia, unsteady gait</td>
<td>1 mo</td>
<td>papilledema; nystagmus;</td>
<td>total</td>
<td></td>
</tr>
<tr>
<td>Case No.</td>
<td>Author (Year)</td>
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<td>Duration</td>
<td>Preoperative neurological findings</td>
<td>Extirpation</td>
<td>Outcome and comments</td>
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<tr>
<td>19</td>
<td>Helle et al. (1980)</td>
<td>27/M</td>
<td>medulla</td>
<td>solid (cyst)</td>
<td>headache, anorexia, nausea, vomiting, diplopia, weight loss</td>
<td>1 yr 3 mos - 4 yrs</td>
<td>rt V, VII, IX, XII cr n palsy; ataxia</td>
<td>VA shunt, cyst drainage, irradiation (60 Gy) total</td>
<td>removed under cardio-pulmonary bypass; hypothermia; hypotension with cardiac standstill; apnea spell; dysarthria; dysphagia; mild quadripareisis; extremity ataxia; died 82 days after surgery due to sleep apnea</td>
</tr>
<tr>
<td>20</td>
<td>Silverberg et al. (1981)</td>
<td>29/M</td>
<td>medulla</td>
<td>hemorrhage, fourth ventricle</td>
<td>head</td>
<td></td>
<td></td>
<td></td>
<td>total</td>
</tr>
<tr>
<td>21</td>
<td>Nakamura et al. (1985)</td>
<td>42/M</td>
<td>medulla</td>
<td>solid: chestnut 20 mm</td>
<td>dysphagia, dysarthria, hoarseness, bil sensory loss</td>
<td>1 yr 4 mos</td>
<td>rt V, IX, X, XI, lt VII, IX, X cr n palsy; sensory loss; bil extremity ataxia</td>
<td></td>
<td>no deficit; removed under hypothermia and cardiac arrest final sleep apnea; hypotension; bulbar palsy; ataxia; lt hemisensory loss; died 1.5 yrs after surgery due to airway obstruction</td>
</tr>
<tr>
<td>22</td>
<td></td>
<td>41/F</td>
<td>medulla</td>
<td>solid: 25 mm</td>
<td>difficulty with handwriting, numbness of the rt arm, gait disturbance, vomiting</td>
<td>6 mos</td>
<td>lt V cr n palsy; lt pyramidal sign; sensory loss; bil extremity ataxia</td>
<td>partial</td>
<td>unstable vital signs; dysphagia; lt hemisataxia; bil sensory loss leads a normal life except bil sensory disorders</td>
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<tr>
<td>23</td>
<td>Tognetti et al. (1980)</td>
<td>24/M</td>
<td>rt medulla</td>
<td>solid: 10 mm</td>
<td>headache, photophobia</td>
<td>3 days</td>
<td>papilledema; nystagmus; hyperrelexia; ataxia</td>
<td>total</td>
<td>uneventful</td>
</tr>
<tr>
<td>24</td>
<td></td>
<td>50/F</td>
<td>medulla</td>
<td>solid: 15 mm (cyst)</td>
<td>headache, vomiting</td>
<td>8 mos</td>
<td>papilledema; nystagmus; hyperrelexia; ataxia</td>
<td>partial</td>
<td>immediate coma; died 7 days after surgery</td>
</tr>
<tr>
<td>25</td>
<td></td>
<td>41/M</td>
<td>rt medulla, pons</td>
<td>solid: 25 mm</td>
<td>neck pain, vomiting, dysphagia, slurred speech, hiccup</td>
<td>5 yrs</td>
<td>nystagmus; rt IX, X cr n palsy; decreased superficial reflexes; gait disturbance; rt ataxia</td>
<td>total</td>
<td>lower cr n palsy; incoordination; improved 4 yrs after surgery</td>
</tr>
<tr>
<td>26</td>
<td></td>
<td>24/M</td>
<td>rt medulla, vermis, lt cbl</td>
<td>solid</td>
<td>headache, nausea, diplopia, gait disturbance</td>
<td>8 mos</td>
<td>vertical nystagmus; hypotonia; rt flexion and rotation of head</td>
<td>total</td>
<td>excellent (progressive neurological recovery); asymptomatic 5 yrs after surgery</td>
</tr>
</tbody>
</table>

### Table 1C Clinical summary of hemangioblastoma of the brain stem, intramedullary

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age/Sex</th>
<th>Tumor Location</th>
<th>Tumor size</th>
<th>Preoperative Symptoms</th>
<th>Duration</th>
<th>Preoperative neurological findings</th>
<th>Exirpation</th>
<th>Outcome and comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>27</td>
<td>Bird and Mendelow (1960)</td>
<td>26/F</td>
<td>medulla, bil cbl</td>
<td>solid</td>
<td>headache, vomiting, tendency to fall incoordination, headache, nausea, vomiting</td>
<td>1 yr</td>
<td>extremity ataxia</td>
<td>total</td>
<td>satisfactory; symptom-free; cerebellar sign; gait disturbance; work as an automechanic 2 yrs after surgery; C6–7 hemangioblastoma excision; disabled and received VP shunt; VHL disease respiratory distress; Lt ataxia; gait disturbance; improved 4 mos after surgery</td>
</tr>
<tr>
<td>28</td>
<td>Chou et al. (1975)</td>
<td>25/M</td>
<td>middle cerebellar peduncle, bil cbl</td>
<td>solid: 25 mm</td>
<td>il cerebellar sign</td>
<td></td>
<td>papilledema;</td>
<td>total</td>
<td></td>
</tr>
<tr>
<td>29</td>
<td>Djindjian (1986)</td>
<td>54/M</td>
<td>Lt nucl cuneatus</td>
<td>large cyst [solid: plum sized]</td>
<td>lt paresthesia, headache, nausea, dysphagia, dysarthria, dys equilibrium</td>
<td>4 mos</td>
<td>deterioration; nystagmus; V, VII, IX, X, XII cr n palsy; Lt ataxia; Lt respiratory distress</td>
<td>total</td>
<td></td>
</tr>
<tr>
<td>30</td>
<td>Abyai et al. (1990)</td>
<td>43/M</td>
<td>medulla</td>
<td>solid: 34 × 23 × 21 mm</td>
<td>confusion, disorientation dysphagia, unsteady gait, headache, vertigo</td>
<td></td>
<td>nystagmus; diplia; IX, X, XII cr n palsy; rt hemiparesis; rt hemisensory loss; truncal ataxia</td>
<td>total</td>
<td>psychosyndrome; complete recovery; nystagmus; vertigo; lower cr n palsy; improved; sensory impairment; unsteady gait; housework</td>
</tr>
<tr>
<td>31</td>
<td>Kohno et al. (1993)</td>
<td>34/F</td>
<td>medulla</td>
<td>solid: 35 × 25 mm (cyst)</td>
<td>headache, vomiting, dysarthria, unsteady gait</td>
<td>6 mos</td>
<td>papilledema; nystagmus; IX, X cr n palsy; truncal and extremity ataxia</td>
<td>partial</td>
<td>sleep apneic attack 2 hrs after surgery; died 2 mos later due to pneumonia</td>
</tr>
<tr>
<td>32</td>
<td>Present Case 1</td>
<td>33/M</td>
<td>dorsal medulla</td>
<td>solid: 35 × 25 mm (cyst)</td>
<td>headache, vomiting, dysarthria, unsteady gait</td>
<td>1 yr</td>
<td>papilledema;</td>
<td>total</td>
<td>unstable vital signs; IX, X, XII cr n palsy; tracheostomy; recovered to normal life</td>
</tr>
<tr>
<td>33</td>
<td>Present Case 2</td>
<td>16/F</td>
<td>dorsal medulla, upper cervical cord</td>
<td>solid: 40 × 30 mm (cyst)</td>
<td>headache, vomiting, unsteady gait</td>
<td>6 mos</td>
<td>papilledema;</td>
<td>total</td>
<td></td>
</tr>
</tbody>
</table>

peritumoral cysts in 10 of 27 cases (37%). Tumor size exceeded 25 mm in diameter in seven of nine Type A cases. Of the 14 Type E cases, the tumor was smaller than 25 mm in diameter in all but two cases. In three of the four Type I cases, the tumor was rather large, exceeding 30 mm. Cystic hemangioblastoma has been reported in only four cases, and the mural nodules in these cases were between 6 and 20 mm in diameter.

In terms of surgical procedures, radical excision was carried out in 29 cases (87.9%), partial removal in three cases, and cyst drainage in one case. Partial removal was done in two cases of Type E and in one case of Type I hemangioblastoma. Surgical mortality was found in 24.2% of all the cases under review; in 25% of the Type A group, 28.6% of the Type E group, and 14.3% of the Type I group. In two of the three cases involving partial removal of the tumor, the patients died soon after surgery. In the Type A group (Table 1A), the tumor was radically excised in 12 cases; three patients died postoperatively, and one died 1 year and 2 months later due to tumor metastasis in the basal ganglia. In the Type E group (Table 1B), the tumor was radically excised in 11 cases; four patients died postoperatively due to bulbar palsy and unstable vital signs. Among the mortalities, the tumor had been radically excised in three patients and partially excised in the fourth. One patient underwent cyst drainage and had an uneventful recovery after surgery. Of the seven Type I cases (Table 1C), the tumor was radically excised in six patients, and four of them were able to resume normal life. In the one case in which the tumor was partially excised, the patient died postoperatively due to a sleep apnea attack.

### Table 2 Characteristics of patients with hemangioblastoma of the brain stem

<table>
<thead>
<tr>
<th>Tumor location</th>
<th>Attached to fourth ventricle floor</th>
<th>Embedded in medulla oblongata</th>
<th>Intra-medullary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total patients [M/F]</td>
<td>12 (6/6)</td>
<td>14 (8/6)</td>
<td>7 (4/3)</td>
</tr>
<tr>
<td>Age of onset [mean]</td>
<td>15-68 (35) yrs</td>
<td>17-50 (31) yrs</td>
<td>20-54 (33) yrs</td>
</tr>
</tbody>
</table>

**Presenting symptoms**
- headache, nausea, vomiting: 10, 7, 6
- unsteady gait: 10, 3, 3
- numbness: 4, 2, 0
- ataxia: 2, 5, 6
- diplopia: 1, 4, 0
- dysphagia/dysarthria: 2, 2, 3

**Mean duration of preoperative symptoms**
- 8.8 mos, 20.5 mos, 9 mos

**Neurological signs**
- cerebellar: 9, 9, 6
- papilledema: 5, 4, 3
- bulbar: 2, 3, 4

**Tumor characteristics**
- solid: 11, 10, 5
- cystic: 1, 2, 1

**Size**
- 6-40 mm, 10-25 mm, 25-40 mm

**Surgical plane**
- cleavage plane: 4, 9, 4

**Surgical procedure**
- radical: 12, 11, 6
- partial: 0, 2, 1
- cyst drainage: 1, 2, 0
- VP or VA shunt: 2, 1, 0

**Postoperative condition**
- bulbar palsy: 5, 5, 3
- unstable vital signs: 6, 4, 3

**Outcome**
- improved: 7, 10, 6
- unchanged: 2, 0, 0
- worsened: 3, 4, 1
- Mortality: 3, 4, 1

VA: ventriculotrial, VP: ventriculoperitoneal.

### Discussion

Hemangioblastoma of the brain stem is rare, with only 33 cases, including our cases, surgically treated since 1960 (Table 1). Anatomically, the dorsal medulla oblongata and the fourth ventricle are the most frequent sites. The dorsal medulla oblongata is very important because of the critical respiratory, vasomotor, and vagal centers. Hemangioblastoma of the fourth ventricle originates from a vascular mesenchymal plate in or adjacent to the posterior end of the floor of the ventricle (posterior medullary velum), which develops in the 3rd month of fetal life. Therefore, this caudal floor of the fourth ventricle, i.e., posterior medullary velum, is another common site. Clinically and surgically, hemangioblastoma of the brain stem is a highly vascular tumor, and was solid in 26 of 30 cases (Table 2). Cystic hemangioblastoma of the brain stem is rare compared with the high frequency in cerebellar hemangioblastoma (64.7–74%), with only four cases reported. The blood supply to the tumor originates from the posterior inferior cerebellar artery, which lies deep in the tumor, and dilated red veins are located dorsally. The tumor is often not easy to excise completely without causing neurological deficit, due to the hypervascularization of the tumor and its critical location with respect to blood supply and draining. Patients often suffer from lower cranial nerve involvement and damage to their respiratory and circulatory centers.

Surgical intervention for hemangioblastoma of the brain stem is controversial, with high mortality associated with single biopsy and/or decompression varying from 33% to 50%. Hemangioblastoma of the brain stem can be divided according to different criteria.
to two locations, fourth ventricle and intramedullary, with differences in the operative risk and functional quality of the clinical results. We classified the locations of 33 cases of hemangioblastoma into: attached to the floor of the fourth ventricle, partially embedded in the dorsal medulla oblongata, and intramedullary; and evaluated the operative risk and the functional quality of the clinical results (Table 2). The overall mortality in the 33 surgically-treated cases was 24.2% (8/33 cases). The postoperative mortality was not significantly different at any location, although the mortality of cases located in the fourth ventricle was somewhat higher than in intramedullary cases (Table 2). A cleavage plane is often present between the tumor and the floor of the fourth ventricle. Small or medium-sized cysts are often present at the external and basal poles of the tumor and extend caudally into the inferior aspect of the fourth ventricle. This cleavage plane often present as a cleavage plane between the nervous parenchyma and intramedullary or partially embedded tumor. Small- or medium-sized cysts are often present at the external and basal poles of the tumor and extend caudally into the inferior aspect of the fourth ventricle. This cleavage plane separates the tumor from the brain parenchyma and facilitates removal of the tumor. The presence of deep feeding arteries and the volume of venous drainage are major threats to hemostasis despite the distinct cleavage plane. Successful removal of the highly vascular tumor requires deliberate dissection of the tumor on a distinct cleavage plane, minimization of brain damage, preferential division of feeding arteries at an early stage, preservation of the draining veins during dissection of the tumor, strict management of respiration, and circulation during the intraoperative and postoperative periods.

Midline incision on the intercolumnar space of the medulla oblongata is recommended for good access to a large intramedullary tumor. Lateral incision is usually performed to avoid damage to nuclear structures when the tumor is located laterally in the caudal medulla oblongata. Six of seven intramedullary tumors were radically excised without mortality, with five patients leading a useful life, and one patient becoming disabled due to a secondary cervical cord lesion during the long-term follow-up period.

Surgical intervention in cases of hemangioblastoma of the brain stem involves some risks even using microsurgical techniques. Vital signs may fluctuate during dissection of the tumor from the brain stem, necessitating termination of the procedure. Tachycardia or bradycardia may occur intermittently, and hypotension is often difficult to control. Total tumor excision has been achieved under conditions of artificial hypothermia and cardiac arrest, and these techniques are recommended as an ad-junct to neurosurgery.

Postoperative complications are common. Eight of the 33 patients died of serious complications such as sleep apnea and bulbar palsy, and two patients became disabled. As in our Case 1, a sleep apnea attack is likely to occur right after the surgery despite good recovery from anesthesia. Respiration maintenance is mandatory, and mechanical control for respiration should be used postoperatively for several days. Patients should be monitored carefully for sleep apnea for at least 1 month. As in our Case 2, bulbar palsy dysfunction may recover about 1.5 months postoperatively. Tracheostomy and/or gastrostomy should be performed if necessary.

Preoperative irradiation may decrease the operative risk. Preoperative irradiation with 6000 rads reduced a tumor by about 45% after 44 months. Gamma knife radiosurgery of 11 cases of solitary small- or medium-sized hemangioblastoma caused shrinkage or stopped growth with a marginal dose of 10 to 15 Gy, but the adjoining cyst did not respond and required later evacuation. Treatment of a hemangioblastoma of the fourth ventricle by radiosurgery resulted in no change in tumor size in 2 years of follow up. Further investigation of the effect of radiosurgery is needed.

Hemangioblastoma of the brain stem can be removed radically by meticulous dissection of the tumor along a distinct cleavage using microsurgical techniques, even in intramedullary locations. Treatment of patients with medullary hemangioblastomas is possible but prudent postoperative care is mandatory to reduce the risk of mortality and morbidity.

References


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