Schwannoma of the medulla oblongata

Case report

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A case of intraparenchymal schwannoma of the medulla oblongata is presented. The radiographic and pathological characteristics of this rare tumor are discussed, and the world literature regarding intracerebral intraparenchymal schwannomas is reviewed. The current etiological theories of these intra-axial nerve-sheath tumors are reviewed. The importance of early identification and differentiation of these potentially curable tumors from malignant glial tumors is emphasized.

KEY WORDS • brain neoplasm • schwannoma • neurinoma • medulla oblongata

INTRAMEDULLARY schwannomas are rare tumors of the central nervous system (CNS). They develop most frequently in the spinal axis, with only occasional reports of an intracranial intraparenchymal location. A search of the literature revealed only 11 cases of intracranial intraparenchymal schwannomas. Prakash, et al., reported a pontine intraparenchymal schwannoma in 1980, and Sarkar, et al., recently described an intracerebellar schwannoma. A case of an intraparenchymal schwannoma of the medulla oblongata is reported in a 50-year-old woman and the radiographic and pathological characteristics of this lesion are discussed. A review of the relevant literature is presented.

Case Report

This 50-year-old right-handed white woman noted the onset of episodic nausea and emesis associated with the occurrence of retro-orbital headaches approximately 6 weeks before admission. Her symptoms progressed in severity, and over the course of the next 3 weeks she noted the gradual development of left-sided facial numbness which spread to involve the left side of the neck, shoulder, and proximal arm in a cape-like distribution. She also noted occasional diplopia and, according to family members, developed slight slurring of speech. She admitted to recent difficulty in swallowing solid food as well as progressive unsteadiness of gait. Her medical history was noncontributory.

Examination. The general physical examination was normal, and there was no physical evidence or family history of neurofibromatosis. Detailed neurological examination at the time of admission revealed an alert woman. Mild bilateral nystagmus evoked by upward and right-lateral gaze was present. There was decreased light touch, temperature, and pinprick sensation in the left trigeminal distribution. The left corneal reflex was depressed. Mildly decreased hearing was noted in the left ear, and Weber and Rinne tests were normal. On examination, gag and swallow reflexes were grossly normal; however, the patient reported subjective difficulty in swallowing sips of water. Her speech was remarkable for a mild nasal twang. Strength was normal in both upper and lower extremities. Sensory examination showed decreased pinprick and light-touch sensation in a left C2–6 distribution as well as in the entire right hand. Proprioception was normal. Deep-tendon reflexes were 2+ and symmetrical; no pathological reflexes were present. The patient had minimal ataxia, which was more pronounced on tandem gait.

The diagnostic impression on initial neurological consultation was of an intrinsic brain-stem mass, and accordingly a magnetic resonance (MR) study of the head was obtained (Fig. 1). Sagittal T₁-weighted images revealed a 1.5-cm hypointense intraparenchymal mass in the medulla with surrounding areas of isointensity. On axial T₂-weighted images, this area was hyperintense. This MR picture, coupled with the subacute nature of the patient's presentation, resulted in the tentative preoperative diagnosis of cystic brain-stem glioma.
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**FIG. 1.** Sagittal T₁-weighted magnetic resonance image (spin-echo sequence: pulse relaxation time (TR) 55 msec, echo delay time (TE) 26 msec) revealing a 1.5-cm hypointense mass in the medulla. Axial T₂-weighted images (spin-echo sequence: TR 2000 msec, TE 120 msec) demonstrated a hyperintense mass (not shown).

**Operation.** The patient underwent a suboccipital craniectomy and posterior fossa exploration in the prone position. After the dura was opened, the cerebellar tonsils were retracted laterally and superiorly revealing the lower vermis and floor of the fourth ventricle. The latter was noted to be asymmetrical, with a noticeable bulging of the right side of the brain stem. This bulging was most prominent just rostral to the obex and toward the right. It was at this point that a small incision was made in the floor of the fourth ventricle. A cystic cavity containing several milliliters of a thick yellowish fluid was encountered. This cavity was irrigated copiously, and biopsies for frozen and permanent section were taken from the cyst wall. The cyst wall was distinct from the surrounding brain parenchyma, and was dissected free in toto and sent for pathological evaluation. The initial pathology report from the frozen-section specimens was of a low-grade brain-stem glioma.

**Postoperative Course.** The patient awoke immediately postoperatively without obvious deficit. However, in the next several hours mild left upper-extremity weakness was noted which did not progress. By the time of her discharge from the hospital on the 11th postoperative day, this weakness had resolved, and she reported improvement in left-sided numbness and swallowing difficulties. A follow-up computerized tomography scan 1 week postoperatively and an MR study 9 months after surgery revealed no residual mass. The patient has continued to do well clinically and exhibited no problems at examination 9 months after surgery.

**Pathological Examination.** The tumor was studied by light and electron microscopy. Hematoxylin and eosin-stained sections through the tumor (Fig. 2) revealed areas of spindle cells with elongated nuclei and eosinophilic cytoplasm surrounding relatively acellular areas. These were areas corresponding to the Antoni A and B patterns characteristic of schwannomas. Frequent areas of exuberant collagen formation were noted. Reticulin staining revealed an extensive network of reticulin. Glial fibrillary acidic protein (GFAP) staining was negative; however, the tumor did stain diffusely positive for S-100, which is characteristic of schwannomas. Electron microscopy showed basement membranes and long spacing collagen fibers, also typical of Schwann cells.

**Discussion**

Intracranial schwannomas represent 7% to 8% of primary brain tumors. Although these tumors may arise from any Schwann cell-myelinated nerve, by far the majority arise from the vestibular division of the eighth cranial nerve and are termed "acoustic neuromas." Intraparenchymal CNS schwannomas are unusual, as Schwann cells are not normally found in the brain or spinal cord parenchyma. In our case, radiological and pathological studies as well as surgical exploration confirmed the intraparenchymal nature of this tumor.

Intramedullary schwannomas have been reported by most authors to be more common in the spinal cord than in other CNS locations. Ross, et al., in 1986 and Drapkin, et al., in 1985 independently reported 27 cases of intramedullary spinal schwannomas. These authors reported a mean age at presentation of 44 years and an average duration of symptoms of 25 months for these patients. Ross, et al.,
briefly reviewed intracerebral schwannomas, but did not comment on the similarities or differences between them and their more common intraspinal counterparts.

The characteristics of all known reported intracranial parenchymal schwannomas are presented in Table 1. As can be seen, one striking difference between patients with intraspinal versus intracranial schwannoma is their age at the time of presentation. Excluding the patient in Case 3 (reported by Ghatak, et al.), who is uncharacteristic because of the extremely long duration of symptoms without treatment (over 40 years), all patients previously reported were under 40 years old at the time of initial presentation (mean age 16 years). This difference between age of presentation in patients with intracerebral as opposed to intraspinal schwannomas may reflect a basic difference in etiology of these two conditions (see below). These two patient groups appear similar in other respects, such as average duration of symptoms (2.1 years), male:female ratio (56%;44%), and general absence of clinical history of neurofibromatosis. The majority of cases of intraparenchymal schwannomas have been reported in the last 10 years; this suggests that improved clinical and pathological diagnostic capabilities may prove that such lesions are less rare than previously thought. The recognition of this curable tumor and its differentiation from brain-stem glioma is of obvious importance.

Other than single reports of intracerebellar and intrapontine schwannomas, all reported cases of intraparenchymal intracranial schwannomas have been supratentorial. Frontal and temporal locations were most frequent. In patients with supratentorial tumors the most common presenting symptom was a history of seizures, most probably related to focal gliosis and irritation in the cortex surrounding the tumor. In contrast, previously reported intraparenchymal infratentorial schwannomas appeared to cause symptoms related most directly to their mass effect (headaches, ataxia, and cranial nerve palsies). Our patient, the 12th reported with an intracranial intraparenchymal schwannoma, represents the first published case of an intraparenchymal schwannoma of the medulla. The location of her tumor and her relatively advanced age at the time of presentation make her case a somewhat atypical example of intracerebral schwannoma. In these respects, this case is more characteristic of high intraspinal schwannomas than those of intracerebral location.

The preoperative MR study correlated well with our intraoperative findings; in particular, it indicated a cystic component to the tumor which was confirmed at surgery. This study did not, however, allow a distinction between intraparenchymal schwannoma and the much more common brain-stem glioma. The report of brain-stem glioma based on the initial frozen section further misled us, and might have caused incomplete resection and repeat surgery had the tumor capsule not been so easily removable. In common with other authors, we emphasize the necessity for correct frozen-section diagnosis in the proper surgical resection of these lesions.

Table 1

Summary of reported intracranial parenchymal schwannomas

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors &amp; Year</th>
<th>Sex &amp; Age (yrs)</th>
<th>Location of Tumor</th>
<th>Duration of Symptoms</th>
<th>Clinical Presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Gibson, et al., 1966</td>
<td>M, 6</td>
<td>temporal</td>
<td>1 yr</td>
<td>rt focal seizure, rt-sided weakness</td>
</tr>
<tr>
<td>2</td>
<td>New, 1972</td>
<td>M, 8</td>
<td>parietal</td>
<td>2 yrs</td>
<td>grand mal seizure, headache, emesis</td>
</tr>
<tr>
<td>3</td>
<td>Ghatak, et al., 1975</td>
<td>F, 63</td>
<td>parietal</td>
<td>40+ yrs</td>
<td>lt focal seizures, hemiparesis</td>
</tr>
<tr>
<td>4</td>
<td>Van Rensburg, et al., 1975</td>
<td>M, 21</td>
<td>temporal</td>
<td>7 yrs</td>
<td>partial complex seizures, headache</td>
</tr>
<tr>
<td>5</td>
<td>Komminoth, et al., 1977</td>
<td>M, 15</td>
<td>cerebellar</td>
<td>2 yrs</td>
<td>lt facial palsy, exophtalmos, lt-sided dysmetria</td>
</tr>
<tr>
<td>6</td>
<td>Prakash, et al., 1980</td>
<td>F, 14</td>
<td>pontine</td>
<td>3 yrs</td>
<td>6th &amp; 7th cranial nerve palsies, ataxia</td>
</tr>
<tr>
<td>7</td>
<td>Shalit, et al., 1982</td>
<td>F, 29</td>
<td>temporal</td>
<td>6 mos</td>
<td>syncpope, headache, blurred vision</td>
</tr>
<tr>
<td>8</td>
<td>Bruni, et al., 1984</td>
<td>M, 39</td>
<td>frontal</td>
<td>2 yrs</td>
<td>neurofibromatosis, generalized seizures</td>
</tr>
<tr>
<td>9</td>
<td>Gökay, et al., 1984</td>
<td>F, 16</td>
<td>frontopontine</td>
<td>3 yrs</td>
<td>generalized seizures, rt hemiparesis</td>
</tr>
<tr>
<td>10</td>
<td>Rodríguez-Salazar, et al., 1984</td>
<td>F, 10</td>
<td>frontal</td>
<td>3 mos</td>
<td>rt focal seizures</td>
</tr>
<tr>
<td>11</td>
<td>Sarkar, et al., 1987</td>
<td>M, 24</td>
<td>cerebellar</td>
<td>3 mos</td>
<td>headache, emesis, diplopia, ataxia</td>
</tr>
<tr>
<td>12</td>
<td>Aryanpur &amp; Long, 1988</td>
<td>F, 50</td>
<td>medullary</td>
<td>1 mo</td>
<td>headache, lt facial &amp; upper-extremity numbness, lower cranial nerve palsies, ataxia</td>
</tr>
</tbody>
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cconversion of developed pial cells into Schwann cells may be a source for the intramedullary Schwann cells from which an intramedullary schwannoma could arise. Others\textsuperscript{11,27} have speculated that these tumors may originate distally on the dorsal root and then grow centrally along the root to an intramedullary location.

Available evidence does not allow direct verification of any of these theories. Certainly, theories postulating that these tumors arise from dorsal roots are not adequate to explain intraparenchymal intracerebral schwannomas. Similarly, hyperplasia of the perivascular nerve plexus or aberrant nerve fiber growth secondary to chronic inflammation would require a long pre-morbid interval exclusive of that required for tumor growth itself. Given the relatively young age of most of the patients involved, these seem unlikely etiologies. Indeed, more than anything else a younger mean age of patients with intracerebral schwannomas favors a developmental cause for these tumors, and suggests the possibility of different etiologies for intracerebral versus intraspinal parenchymal schwannomas. Based on the clinical heterogeneity of these tumors, it is not unreasonable to conclude that several different etiological forces may be possible, and that the above theories may represent different pathways to the final common outcome of intraparenchymal schwannoma.

References


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