Surgical treatment for extremely rare solitary fibrous tumors of the central nervous system originating from cranial nerve VIII: new clinicopathological findings. Illustrative case

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BACKGROUND Reports of solitary fibrous tumors (SFTs) of the central nervous system (CNS) originating from cranial nerves are extremely rare. The origins of these neurogenic SFTs of the CNS have been determined only by intraoperative findings, and there is no pathological evidence of whether they really originated from cranial nerves.

OBSERVATIONS A 54-year-old female with hearing loss and facial paralysis presented with a giant right cerebellopontine angle tumor. She was diagnosed with a petrous meningioma based on preoperative imaging, and the tumor was removed via the retrosigmoid approach after embolization of the tumor-feeding vessels. Intraoperatively, the tumor was not attached to the dura mater but extended from the internal auditory canal to the cisternal portion. The acoustic nerve was not identified, but it was possible to separate the tumor from the facial nerve. The tumor was removed as an acoustic schwannoma intraoperatively. Postoperative pathological examination revealed an SFT. Immunostaining revealed peripheral nerve bundles entrapped within the tumor tissue. The patient was diagnosed with an SFT of the CNS originating from the acoustic nerve.

LESSONS A neurogenic SFT of the CNS was diagnosed based on both intraoperative and pathological findings.

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KEYWORDS solitary fibrous tumors; central nervous system; schwannoma; meningioma

Solitary fibrous tumors (SFTs) of the central nervous system (CNS) were first described by Carneiro et al.1 in 1996. Most SFTs of the CNS reportedly arise from the meninges. The various origin sites have included the supra- and infratentorial locations, orbit, and spinal cord.2,3 However, there have been extremely rare reports of SFTs of the CNS originating from the cranial nerves, which have minimal relation with meningotheial tissue. The origin of these neurogenic SFTs of the CNS have been determined only by intraoperative findings (Table 1),4–14 and there is no pathological evidence of whether they really originate from cranial nerves. Here, we report the first case of a neurogenic SFT of the CNS arising from the acoustic nerve that was diagnosed based on both intraoperative and pathological findings. SFTs of the CNS require aggressive resection owing to the high local recurrence rate and metastasis rate to other organs. We also briefly discuss the surgical difficulty, especially when the tumor arises from cranial nerves.

Illustrative Case
A 54-year-old female presented to our hospital with gradually progressive right-sided hearing loss, facial palsy, and gait disturbances for several years. She presented with a House-Brackmann grade 2 facial palsy, Gardner-Robertson grade 3 hearing loss, right facial hypesthesia, cerebellar ataxia, and headache. Cranial contrast-enhanced magnetic resonance imaging (MRI) revealed a giant mass lesion with a uniform contrast pattern in the right cerebellopontine angle (CPA) and a dural tail sign along the petrous bone. The mass significantly compressed the brainstem and cerebellum, resulting in obstructive hydrocephalus (Fig. 1). Although the tumor extended into the internal auditory canal,
head computed tomography showed no canal enlargement. Angiography revealed tumor staining from the petrosal branch of the middle meningeal artery (MMA) and inflow from the surface of the pyramidal bone on the cranioventral side of the internal auditory canal (Fig. 2). Pial supply from the anterior inferior cerebellar artery (AICA) was also observed. Following coil embolization of the MMA, the patient underwent tumor resection. Auditory brainstem response, monopolar mapping of the facial nerve, and eye movement monitoring were conducted. The tumor was removed via the retrosigmoid approach. The tumor was not clearly attached to the dura mater, but it seemed to extend from the internal auditory canal into the cisternal portion (Fig. 3). The internal auditory canal was filled with the tumor, and when the tumor was dissected from the fundus to the lateral-to-medial region, the bundle of the facial nerve was clearly identified. However, the vestibular and cochlear nerves were not identified. Intraoperatively, the tumor was presumed to be an acoustic schwannoma rather than a meningioma. The facial nerve was fanning and firmly attached to the tumor, and its route within the cisternal segment was not confirmed. There were strong adhesions to the brainstem and cerebellum. Moreover, because of unexpected bleeding from the tumor, partial resection was inevitable. Postoperatively, all symptoms except for her hearing improved. After rehabilitation and a full-body examination, she was discharged to home 1-month postsurgery. Histopathology showed proliferating tumor cells with oval to spindle-shaped nuclei, an intercellular fibrous matrix, and scattered thin-walled blood vessels with a dilated lumen. Fewer than 1 fission cell was observed per 10 intensely magnified views, and no necrotic cells were observed. Immunostaining showed that the tumor cells were positive for STAT-6 and CD34. Ki-67 positivity was less than 1%. The tumor was negative for EMA and SSTR2A. It should be noted that peripheral nerve bundles entrapped by tumor cells were identified, so the Schwann cells were positive for S-100, SOX, and Schwann2E (Fig. 4). Moreover, the peripheral nerve axons were positive for neurofilaments.

TABLE 1. Reports of SFTs and HPCs of the central nervous system originating from cranial nerves

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Authors &amp; Year</th>
<th>Age (yrs)/Sex</th>
<th>Nerve of Origin</th>
<th>Pre-Tx</th>
<th>Extent of Resection</th>
<th>Additional Tx</th>
<th>Pathological Dx</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Tan et al., 2001 🅃️</td>
<td>41/M</td>
<td>Trigeminal nerve</td>
<td>—</td>
<td>NA</td>
<td>NA</td>
<td>HPC</td>
</tr>
<tr>
<td>2</td>
<td>Badion et al., 2003 🅃️</td>
<td>47/F</td>
<td>Hypoglossal nerve</td>
<td>—</td>
<td>Complete resection</td>
<td>NA</td>
<td>SFT</td>
</tr>
<tr>
<td>3</td>
<td>Caroli et al., 2004 🅃️</td>
<td>38/M</td>
<td>Trigeminal nerve</td>
<td>—</td>
<td>Subtotal resection</td>
<td>—</td>
<td>SFT</td>
</tr>
<tr>
<td>4</td>
<td>Waldron et al., 2006 🅃️</td>
<td>29/F</td>
<td>Abducent nerve</td>
<td>Embolization</td>
<td>Small remnant involving the cavernous sinus &amp; Mecckel’s cave</td>
<td>Radiation</td>
<td>SFT</td>
</tr>
<tr>
<td>5</td>
<td>Motoori et al., 2010 🅄️</td>
<td>49/F</td>
<td>Trigeminal nerve</td>
<td>Biopsy</td>
<td>Complete resection</td>
<td>—</td>
<td>SFT</td>
</tr>
<tr>
<td>6</td>
<td>Scholsem &amp; Scholtes, 2012 🅃️</td>
<td>36/F</td>
<td>Vagus nerve</td>
<td>—</td>
<td>Complete resection</td>
<td>—</td>
<td>SFT</td>
</tr>
<tr>
<td>7</td>
<td>Teoh et al., 2014 🅄️</td>
<td>24/F</td>
<td>Acoustic nerve s/o</td>
<td>—</td>
<td>Almost complete resection</td>
<td>Radiation</td>
<td>HPC</td>
</tr>
<tr>
<td>8</td>
<td>Yue et al., 2020 🅄️</td>
<td>66/M</td>
<td>Acoustic nerve s/o</td>
<td>—</td>
<td>NA</td>
<td>—</td>
<td>SFT/HPC</td>
</tr>
<tr>
<td>9</td>
<td>Oishi et al., 2020 🅄️</td>
<td>65/M</td>
<td>Trigeminal nerve</td>
<td>—</td>
<td>Partial resection</td>
<td>Radiation → op</td>
<td>SFT/HPC</td>
</tr>
<tr>
<td>10</td>
<td>Li et al., 2021 🅄️</td>
<td>41/M</td>
<td>Hypoglossal nerve</td>
<td>Radiation</td>
<td>Complete resection</td>
<td>Radiation</td>
<td>SFT/HPC</td>
</tr>
<tr>
<td>11</td>
<td>Frantzias et al., 2022 🅄️</td>
<td>32/M</td>
<td>Trigeminal nerve</td>
<td>—</td>
<td>NA</td>
<td>—</td>
<td>HPC</td>
</tr>
<tr>
<td>12</td>
<td>Present case 2023 🅄️</td>
<td>54/F</td>
<td>Acoustic nerve</td>
<td>—</td>
<td>Partial resection</td>
<td>—</td>
<td>SFT</td>
</tr>
</tbody>
</table>

Dx = diagnosis; NA = not applicable; s/o = suspect of; Tx = treatment.

FIG. 1. MRI revealed right CPA tumor with dural tail sign along the petrous bone (A), which significantly compressed the brainstem and cerebellum (B), resulting in obstructive hydrocephalus (C). The tumor extended into the internal auditory canal (A).
Based on both intraoperative and pathological findings, the patient was diagnosed with an SFT of the CNS originating from the acoustic nerve (World Health Organization [WHO] grade 1). Systemic examination revealed a liver lesion. A subsequent biopsy revealed an SFT, with characteristics similar to the CNS lesion, suggesting liver metastasis from the CNS lesion. Although additional resection was recommended for both the CNS and liver lesions, the patient requested to be placed under close observation.

**Patient Informed Consent**

The necessary patient informed consent was obtained in this study.

**Discussion**

**Observations**

In the fourth edition of the WHO classification (2016), SFT and hemangiopericytoma (HPC) were integrated because of the identification of NAB2/STAT6. Then, in the fifth edition of the WHO classification of brain tumors (2021), SFT and HPC were merged into SFT because distinguishing between the two entities was not significant. These tumors were collectively referred to as tumors arising outside the CNS. Neurogenic SFT of the CNS is extremely rare, with only a few reported cases. Ours is the first report of a neurogenic SFT of the CNS, supported by the identification of peripheral nerve bundles in the tumor tissue, as well as the intraoperative findings. Certainly, distinguishing whether a tumor such as the one in our case originates from the nerve itself or the structures surrounding the nerve is challenging. However, pathological findings revealed a clear proliferation of peripheral nerve fibers within the tumor tissue, leading us to suspect a neurogenic SFT of the CNS.

SFTs of the CNS have a higher local recurrence rate (61%–76%) and metastasis rate (23%–64%) than schwannomas and meningiomas, which are differential diagnoses. Thus, it is crucial to confirm the diagnosis because immediate and complete removal of the tumor is ideal for SFTs. Previous studies have proposed the application of preoperative diagnostic imaging, but there have been no cases of neurogenic SFTs of the CNS that were definitively diagnosed preoperatively or intraoperatively. In our case, the patient was also diagnosed and treated for a schwannoma or meningioma. In cases of an SFT of the CNS arising at the CPA, strong adhesions to the developing and surrounding nerves are problematic. Li et al. reported a case of HPC of the jugular foramen that regrew 6 years after the patient had received radiotherapy for a glomus tumor. Because of the significant adhesions to the hypoglossal nerve, it had to be sacrificed. In this case, the tumor was considered to originate from the acoustic nerve. Although it was possible to separate the tumor from the facial nerve in the internal auditory canal, it was impossible to separate it from the cisternal portion owing to the strong adhesions. Neuroradiography was performed to prevent new neurological symptoms, and partial resection was performed. Hypervascularity was another factor that made the resection difficult. Oishi et al. reported that the initial resection of SFT/HPC in the CPA and Meckel’s cave had been terminated and only a partial resection was performed owing to heavy intraoperative hemorrhage. In the present case, only tumor embolization of the external carotid artery system was performed preoperatively, and the vertebrobasilar system feeder remained, resulting in unexpected bleeding. Some reports recommend multistage surgery for hypervascularized acoustic schwannomas to allow for hemorrhage control. Thus, planned multistage surgery should be explored as a possible management option for this disease.

**Lessons**

A neurogenic SFT of the CNS was diagnosed based on both intraoperative and pathological findings. Neurogenic SFTs of the CNS should be treated carefully because of the high likelihood of...
incomplete resection because of diagnostic fluidity, strong adhesion to the surrounding nerve, and hemorrhage control. A prompt full-body examination may be recommended when diagnosing intracranial SFTs.

References

Disclosures
The authors report no conflict of interest concerning the findings specified in this paper.

Author Contributions
Conception and design: Arai, Shimizu. Acquisition of data: Arai. Analysis and interpretation of data: Arai. Drafting the article: Arai. Critically revising the article: Arai, Shimizu. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Arai. Statistical analysis: Arai. Administrative/technical/material support: Arai. Study supervision: Arai, Shimizu.

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