Intraparenchymal Schwannoma of Temporal Lobe: A Case Report and Review of the Literature

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Abstract

Intracranial schwannomas (ISs) account for approximately 8% of intracranial tumors, while IS, a rare entity, is responsible for roughly 1% of IS. A 33-year-old man with a 3-month headache and sudden onset seizure was referred to our clinic. Preoperative magnetic resonance imaging revealed a contrast-enhancing mass accompanied by cystic components in the right temporal lobe. Ganglioglioma, metastasis, or glioblastoma multiforme was suspected, and surgery was advised. During surgery, gross total resection of a noninvasive tumor was conducted. Postoperative recovery was uneventful. Based on histopathological examination and confirmatory immunohistochemistry, the intraparenchymal temporal tumor was diagnosed as schwannoma. ISs are extremely scarce brain tumors mainly located on the surface of the brain or adjacent brain ventricles. The definite preoperative diagnosis of schwannoma cannot be readily established due to radiologically indistinguishable features from metastasis and gliomas; however, histopathology and immunohistochemistry are of great assistance. Complete surgical removal is the most preferred treatment alternative with a long-term favorable prognosis without adjuvant and neoadjuvant chemotherapy requirements.

Keywords

► brain tumors
► case report
► intraparenchymal schwannoma
► review
► temporal lobe

Introduction

Schwann cells are a type of glial cells present in the peripheral nervous system (PNS). Generally, Schwann cells are categorized into two types, including myelinating and nonmyelinating cells, which have essential functions in maintaining and regenerating the axons of peripheral nerves.1 Schwannoma (World Health Organization [WHO] grade I) is an almost benign primary tumor of Schwann cells, which accounts for 89% of all neural sheath tumors, and vestibular schwannoma represents approximately 60% of schwannomas.2 Schwannoma can occur in intra- and extracranial peripheral nerves, and intracranial schwannoma (IS) constitutes 8% of cerebral tumors. Roughly 60% of ISs are related to the cranial nerves and are commonly present in the vestibular

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cranial nerve VIII) and trigeminal (cranial nerve V) nerves, respectively. Even though Schwann cells are not prevailing in the brain parenchyma, intraparenchymal schwannoma is extremely rare and responsible for less than 1% of IS.

The definitive preoperative diagnosis of IS cannot be clearly established, and postoperative histopathological evaluations mainly confirm the diagnosis. Here, we sought to present the clinical, radiological, and histopathological features of a young patient diagnosed with temporal IS and a thorough literature review so as to shed light on some untouched aspects of ISs.

Case Report

A 33-year-old male with the presentation of a 3-month headache and sudden onset epileptic seizure was referred to our neurosurgery clinic. He had one focal seizure with impaired consciousness without aura that lasted roughly 2 minutes. During the attack, lip smacking and repetitive speech were observed by his family, followed by confusion and inability to remember the seizure. The patient had neither relevant medical and family history nor cutaneous stigmata of neurofibromatosis type I. The neurological examination revealed no focal neurological deficits. On brain magnetic resonance imaging (MRI), the lesion had a cystic component and peritumoral edema with hypo-signal intensity in T1 and hypersignal intensity in T2. Homogenous enhancement after contrast injection was observed in the lesion (►Fig. 1).

The patient underwent a right-sided temporal craniotomy under general anesthesia with a differential diagnosis of ganglioglioma, metastasis, or glioblastoma multiforme. The tumor was then resected completely via a transcortical approach under microscopic view with a Cavitron ultrasonic surgical aspirator and electrocautery. Intraoperative findings were a well-demarcated, firm, round tumor lying in the right temporal lobe. The lesion was elastic, hard, and yellowish in color. Postoperative MRI revealed complete tumor removal, and the patient had no neurological symptoms over a follow-up of 62 months (►Fig. 2).

Histological findings demonstrated the proliferation of spindle cells with elongated-looking nuclei in a hyalinized and myxoid background with foci of lymphocytic infiltration. Further immunohistochemical (IHC) examination revealed diffuse and strong positivity for S-100 protein. Although glial fibrillary acidic protein (GFAP) glial marker and vimentin were positive in tumoral cells, epithelial membrane antigen (EMA) was negative (►Fig. 3). Based on the above-mentioned findings, the tumor was diagnosed as a WHO grade I intraparenchymal schwannoma.

Discussion

ISs are exceedingly rare primary brain tumors located within the brain parenchyma and account for 1% of IS. Schwannomas are mainly originated from the Shawn cells of the sheet
of cranial and peripheral nerves related to the PNS. Since Schwann cells are present in the PNS and are not histologically present in the parenchyma of central nervous system, it is difficult to explain the cause of IS. However, some developmental and non-developmental theories have been proposed to elucidate the cause of IS. The developmental theory reveals that the Schwann cells underwent ectopic migration during embryonic neurogenesis, rendering it possible for them to exist in the brain parenchyma. Moreover, the proliferation and differentiation of pial mesenchymal stem cells to the histologically Schwann-like cells support the hypothesis of developmental theory.

Several case reports indicated the different locations of IS that are not related to the intra-crani al macrovasculature and can also associate with the perivascular nerve plexus adjacent to the large intracranial deep arteries. The non-developmental theory claims that the intraparenchymal Schwann cells stem from the perivascular nerve plexus of parenchymal arterioles. Therefore, the vast majority of ISs are supratentorial and present on the brain surface and adjacent to the ventricles. Although different theories have been proposed for the tumorigenesis of IS, the exact mechanism is also unclear, and further studies are suggested. To date, 150 confirmed cases ISs are reported, among which 65% were located supratentorially and 35% infratentorially.

There is no significant difference in the sex and site of such tumors; however, an infinitesimal predominance in men has been reported. Ten et al reported that the prevalence of IS among young men was slightly higher than among women, whereas the result was converse in patients aged over 40 years. Moreover, Kovalainen et al reviewed the 150 cases of IS and corroborated previous findings that there was a minute tendency to the male gender in patients with IS (male-to-female ratio: 1.27). Although no definite age-specific presentations were proposed, young adults mainly suffered from headaches and seizures similar to our patient, whereas the primary manifestation of the elderly has been reported to be focal neurological deficits. The clinical manifestations of IS are not specific and depend on the size and location of the tumor; however, headache and seizure are the most common ones. Reviewing the literature showed that the frontal and temporal lobes are the most common sites for developing IS.

There are no specific radiological features for the IS, and imaging findings are thought to be similar to the ISs. These features include calcification, cystic components, and edema. Even though computed tomographic scans can more potentially detect the calcification of IS tumors, MRI is the superior modality to diagnose IS. On brain MRI, the lesion appears hypointense and hyperintense on T1- and T2-weighted sequences, respectively. The solid portion and cyst wall often show a homogenous enhancement following contrast administration.

It is not straightforward to make a preoperative diagnosis of IS based on clinical and radiological features, and the combination of histopathological examinations and IHC can be of great assistance. The IHC analysis of schwannomas represents a strong positivity of S-100 protein and vimentin filament and no reactivity for EMA and GFAP markers. The IHC test of our patient revealed strong reactivity of S-100, vimentin, and GFAP with no reactivity for EMA, which is almost in line with pertinent literature.

ISs are benign tumors and require no adjuvant chemotherapy, and the primary strategy has been the complete removal of the tumor. Previous studies have explicated that clinical manifestations would gradually disappear after GTR. Similarly, our patient's symptoms faded following the surgery, and he experienced no neurological deficits during a long-term follow-up. Moreover, a recurrence rate of 5.3% has been reported after GTR owing to malignant histopathology of the tumor. In cases that underwent subtotal resection, only four patients with no malignant pathology needed a second surgery, proposing that recurrence is due to incomplete tumor removal. We have summarized the
Table 1: Summary of 20 cases of temporal lobe intraparenchymal schwannomas

<table>
<thead>
<tr>
<th>Author(s)/year</th>
<th>Age (y)/sex</th>
<th>Symptoms</th>
<th>Duration of symptoms presentation</th>
<th>Size</th>
<th>Cystic component</th>
<th>Preoperative radiological diagnosis</th>
<th>EOR</th>
<th>IHC confirmed diagnosis</th>
<th>Follow-up</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present study/2022</td>
<td>33/M</td>
<td>Headache, seizure</td>
<td>3 months (headache), sudden onset seizure</td>
<td>4 × 4 × 3 cm³</td>
<td>Yes</td>
<td>Ganglioglioma, metastasis, GBM</td>
<td>GTR</td>
<td>Yes (S-100 + Vimentin + GFAP = EMA –)</td>
<td>62 months</td>
<td>No</td>
</tr>
<tr>
<td>Patankar et al. /2019</td>
<td>20/M</td>
<td>Headache, vomiting</td>
<td>3 months</td>
<td>4 × 5 cm</td>
<td>No</td>
<td>Meningioma</td>
<td>GTR</td>
<td>Yes (S-100 + Vimentin + GFAP –)</td>
<td>6 months</td>
<td>No</td>
</tr>
<tr>
<td>Chen et al. /2019</td>
<td>46/M</td>
<td>Seizures, headache</td>
<td>3 months (headache), sudden onset seizure</td>
<td>NA</td>
<td>No</td>
<td>Meningioma</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP –)</td>
<td>6 months</td>
<td>No</td>
</tr>
<tr>
<td>Wilson et al. /2016</td>
<td>34/M</td>
<td>Asymptomatic</td>
<td>Incidental finding</td>
<td>2.2 × 2.1 × 1.9 cm³</td>
<td>Yes</td>
<td>Ganglioglioma, oligodendroglioma, post infectious</td>
<td>GTR</td>
<td>Yes (S-100 + EMA + GFAP – CD34 +)</td>
<td>NA</td>
<td>No</td>
</tr>
<tr>
<td>Al Batly et al. /2014</td>
<td>49/F</td>
<td>Headache, gait disturbance</td>
<td>Long-term headache</td>
<td>NA</td>
<td>Yes</td>
<td>Astrocytoma, GBM</td>
<td>STR</td>
<td>Yes (S-100 + GFAP –)</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Luo et al. /2013</td>
<td>51/M</td>
<td>Headache</td>
<td>1 month</td>
<td>NA</td>
<td>Yes</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP –)</td>
<td>NA</td>
<td>No</td>
</tr>
<tr>
<td>Guha et al. /2012</td>
<td>51/F</td>
<td>Seizures</td>
<td>4 years</td>
<td>1.2 × 1.3 × 0.9 cm³</td>
<td>No</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP –)</td>
<td>6 months</td>
<td>No</td>
</tr>
<tr>
<td>Bhatte et al. /2003 (abstract only)</td>
<td>50/M</td>
<td>Seizures, headache</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + Vimentin + EMA –)</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Sharma et al. /1998 (abstract only)</td>
<td>8/F</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>Esthesioneuroblastoma, fungal granuloma, nasoethmoid carcinoma</td>
<td>GTR</td>
<td>Yes (S-100 –)</td>
<td>NA</td>
<td>No</td>
</tr>
<tr>
<td>Sharma et al. /1996</td>
<td>8/M</td>
<td>Seizures</td>
<td>4 years</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP –)</td>
<td>NA</td>
<td>No</td>
</tr>
<tr>
<td>0.5/F</td>
<td>Seizures, hemiparesis, vomiting</td>
<td>2 months</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP –)</td>
<td>NA</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Casadei et al. /1993</td>
<td>16/M</td>
<td>Asymptomatic</td>
<td>Incidental finding</td>
<td>1.5 cm</td>
<td>No</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP – EMA –)</td>
<td>2 years</td>
<td>No</td>
</tr>
<tr>
<td>17/M</td>
<td>Seizure</td>
<td>6 months</td>
<td>1.2 cm</td>
<td>No</td>
<td>NA</td>
<td>STR</td>
<td>Yes (S-100 + GFAP – EMA –)</td>
<td>1 year</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>23/F</td>
<td>Headache</td>
<td>10 days</td>
<td>4</td>
<td>No</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP – EMA –)</td>
<td>2 months</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>49/F</td>
<td>Headache</td>
<td>2 months</td>
<td>1.8 cm</td>
<td>No</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + GFAP – EMA –)</td>
<td>2 years</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>84/F</td>
<td>Mental change, hemiparesis</td>
<td>3 weeks</td>
<td>6 cm</td>
<td>Yes</td>
<td>NA</td>
<td>STR</td>
<td>Yes (S-100 + GFAP – EMA –)</td>
<td>2 years</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Firm et al. /1992</td>
<td>11/F</td>
<td>Seizures</td>
<td>NA</td>
<td>NA</td>
<td>Yes</td>
<td>NA</td>
<td>GTR</td>
<td>Yes (S-100 + Vimentin + GFAP – EMA –)</td>
<td>15 months</td>
<td>No</td>
</tr>
<tr>
<td>Kasantikul et al. /1981</td>
<td>21/M</td>
<td>Seizures</td>
<td>5 years</td>
<td>5.5 cm</td>
<td>No</td>
<td>NA</td>
<td>Temporal lobectomy</td>
<td>No</td>
<td>NA</td>
<td>No</td>
</tr>
<tr>
<td>Van Rensburg et al. /1975</td>
<td>21/M</td>
<td>Seizures, headache, amnesia</td>
<td>7 years</td>
<td>2 cm</td>
<td>Yes</td>
<td>Glioma, calcified hamartoma</td>
<td>GTR</td>
<td>No</td>
<td>21 months</td>
<td>No</td>
</tr>
<tr>
<td>Gibson et al. /1966</td>
<td>6/M</td>
<td>Seizures</td>
<td>12 months</td>
<td>8.6 × 4.5 cm³</td>
<td>No</td>
<td>NA</td>
<td>GTR</td>
<td>No</td>
<td>6 months</td>
<td>No</td>
</tr>
</tbody>
</table>

Abbreviations: EMA, epithelial membrane antigen; EOR, extent of resection; F, female; GBM, glioblastoma multiforme; GFAP, glial fibrillary acidic protein; GTR, gross total resection; IHC, immunohistochemistry; M, male; NA, not available; STR, subtotal resection.
details of patients harboring temporal ISs, including our case so as to provide a thorough review of their clinical, radiological, and histopathological characteristics (►Table 1).

Conclusion
ISs are rare low-grade tumors easily cured with complete removal. Preoperative diagnosis is not readily established, and histopathology and confirmatory IHC play a pivotal role among diagnostic modalities. Notwithstanding the predominance of IS in young adults, it cannot be excluded in older individuals. To select the best surgical strategy, ISs should be taken into consideration preoperatively when radiological characteristics such as peritumoral edema, calcifications, and cystic components are observed.

Informed Consent
Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

Authors’ Contributions
S.S. and M.A.H. contributed to writing the paper, data collection, interpretation, and leadership responsibility for the research activity planning and execution, including mentorship external to the core team. M.E., F.K., and M.M.V. contributed to data collection and interpretation. R.Z. contributed to the study concept or design and interpretation.

Conflict of Interest
None declared.

References