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

Sajjad Saghebdoust[1] Mohammad Amin Habibi[2] Mehran Ekrami[3] Farshid Khadivar[4] Mohammad Moein Vakilzadeh[1] Reza Zare[1]

[1] Department of Neurosurgery, Razavi Hospital, Mashhad, Iran
[2] Faculty of Medicine, Qom University of Medical Sciences, Qom, Iran
[3] Department of Neurosurgery, Faculty of Medicine, Mashhad University of Mashhad, Mashhad, Iran
[4] Department of Clinical Sciences, Faculty of Veterinary Medicine, Ferdowsi University of Mashhad, Mashhad, Iran

Address for correspondence Reza Zare, MD, Department of Neurosurgery, Razavi Hospital, MashhadMashhad, 9189845940, Iran (e-mail: drrezazare@gmail.com).

AJNS 2023;18:191–195.

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

article published online
April 3, 2023

DOI https://doi.org/10.1055/s-0043-1763525.
ISSN 2248-9614.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

(cranial nerve VIII) and trigeminal (cranial nerve V) nerves, respectively.[3] Even though Schwann cells are not prevailing in the brain parenchyma, intraparenchymal schwannoma is extremely rare and responsible for less than 1% of IS.[3]

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**).

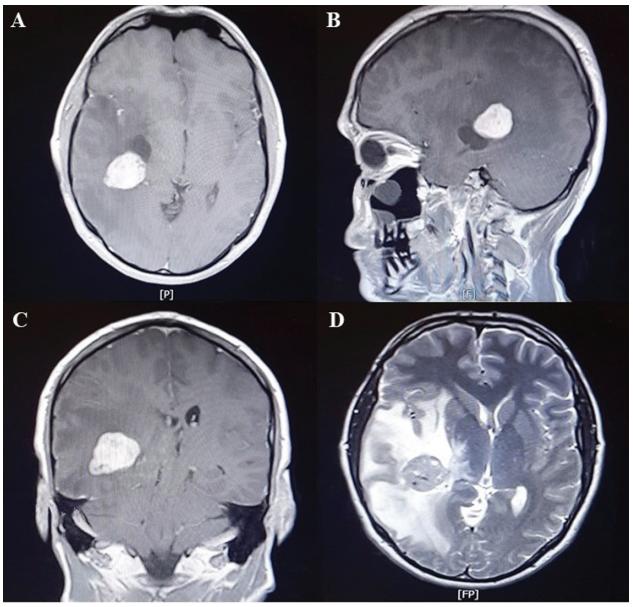

Fig. 1 Preoperative magnetic resonance imaging, including axial (**A**), sagittal (**B**), and coronal (**C**) postcontrast T1-weighted images showed a homogenously enhanced solid mass accompanied by cystic components in the right temporal lobe. Preoperative axial T2-weighted image (**D**) revealed a hyperintense lesion with marked peritumoral edema.

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

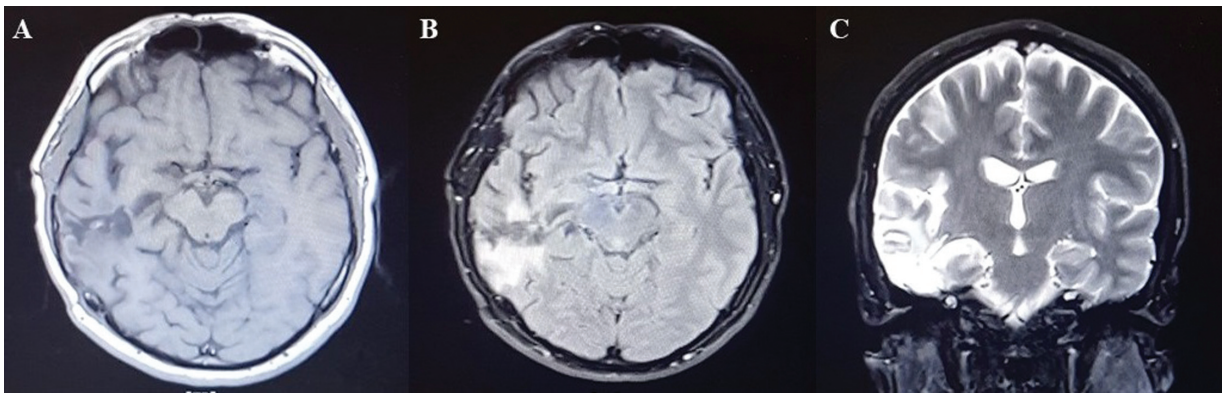

Fig. 2 Postoperative axial T1-weighted (**A**), fluid-attenuated inversion recovery-weighted (**B**), and coronal T2-weighted images showed complete tumor removal (**C**).

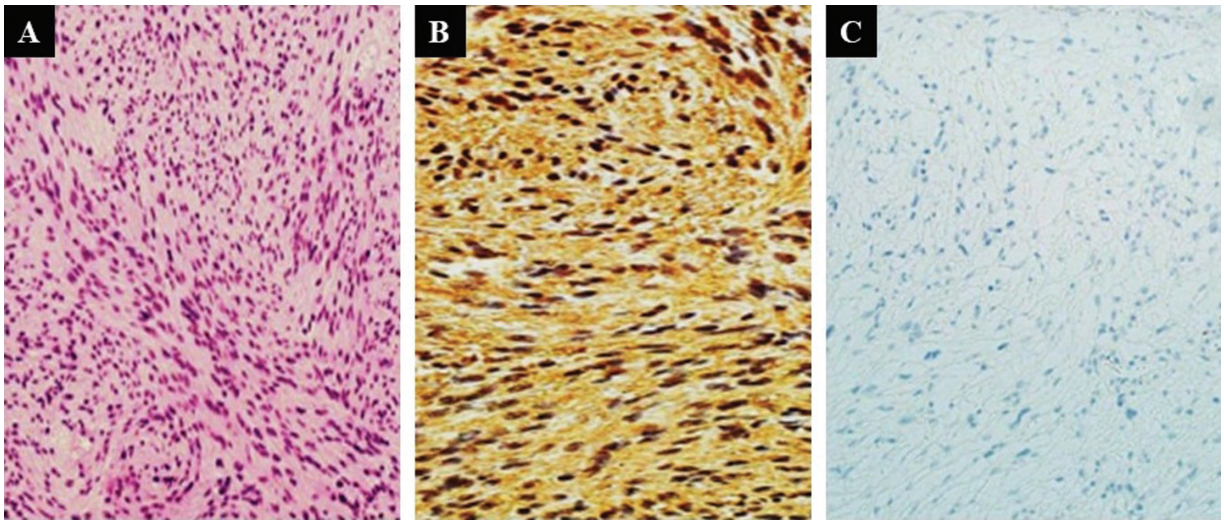

Fig. 3 Elongated spindle cells are shown in hematoxylin and eosin (H&E) staining (**A**). Strong positivity for S-100 protein was noted following immunohistochemistry staining (**B**). Epithelial membrane antigen (EMA) protein staining was negative (**C**). (**A**: H&E 200 ×; **B**: S-100 400 ×; **C**: EMA 100 ×).

of cranial and peripheral nerves related to the PNS.[4] 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.[5]

Several case reports indicated the different locations of IS that are not related to the intra-cranial 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.[6] Therefore, the vast majority of ISs are supratentorial and present on the brain surface and adjacent to the ventricles.[7] 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 supratentrorially and 35% infratentorially.[8]

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[9] 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[8] 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.[10] 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.[11] Reviewing the literature showed that the frontal and temporal lobes are the most common sites for developing IS.[8]

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.[12] 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.[13]

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.[8] The IHC analysis of schwannomas represents a strong positivity of S-100 protein and vimentin filament and no reactivity of EMA and GFAP markers.[8] 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.[14,15] 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.[8] We have summarized the

Table 1 Summary of 20 cases of temporal lobe intraparenchymal schwannomas

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

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

1 Bhatheja K, Field J. Schwann cells: origins and role in axonal maintenance and regeneration. Int J Biochem Cell Biol 2006;38 (12):1995–1999

2 Propp JM, McCarthy BJ, Davis FG, Preston-Martin S. Descriptive epidemiology of vestibular schwannomas. Neuro-oncol 2006;8 (01):1–11

3 Deora H, Srinivas D, Beniwal M, Vikas V, Rao KVLN, Somanna S. Rare cranial nerve schwannomas: a retrospective review of non-trigeminal, nonvestibular cranial nerve schwannomas. J Neurosci Rural Pract 2018;9(02):258–263

4 McLendon R, Rosenblum M, Bigner D. Russell & Rubinstein's Pathology of Tumors of the Nervous System. 7th ed. London: CRC Press; 2006

5 Casadei GP, Komori T, Scheithauer BW, Miller GM, Parisi JE, Kelly PJ. Intracranial parenchymal schwannoma. A clinicopathological and neuroimaging study of nine cases. J Neurosurg 1993;79(02):217–222

6 Guha D, Kiehl T-R, Krings T, Valiante TA. Intracerebral schwannoma presenting as classic temporal lobe epilepsy. J Neurosurg 2012;117(01):136–140

7 Paredes I, Jimenez Roldán L, Ramos A, Lobato RD, Ricoy JR. Intraparenchymal schwannomas: report of two new cases studied with MRI and review of the literature. Clin Neurol Neurosurg 2012;114(01):42–46

8 Kovalainen A, Haeren R, Paetau A, Lehecka M. Brainstem intraparenchymal schwannoma: a case report and literature review. Surg Neurol Int 2021;12:508

9 Ten H, Adachi K, Yamaguchi F, Matsuno A, Teramoto A, Morita A. Occipital lobe epilepsy was presented in a patient with intracerebral schwannoma: a case report and literature review. Int J Neurosci 2019;129(03):308–312

10 Gao Y, Qin Z, Li D, et al. Intracerebral schwannoma: a case report and literature review. Oncol Lett 2018;16(02):2501–2510

11 Sharma MC, Karak AK, Gaikwad SB, Mahapatra AK, Mehta VS, Sudha K. Intracranial intraparenchymal schwannomas: a series of eight cases. J Neurol Neurosurg Psychiatry 1996;60(02):200–203

12 Lipper MH, Eberhard DA, Phillips CD, Vezina L-G, Cail WS. Pleomorphic xanthoastrocytoma, a distinctive astroglial tumor: neuroradiologic and pathologic features. AJNR Am J Neuroradiol 1993;14(06):1397–1404

13 Zagardo MT, Castellani RJ, Rees JH, Rothman MI, Zoarski GH. Radiologic and pathologic findings of intracerebral schwannoma. AJNR Am J Neuroradiol 1998;19(07):1290–1293

14 Bruner J. Immunocytochemistry of recurring intracerebral nerve sheath tumor. J Neuropathol Exp Neurol 1984;43:296

15 Rodriguez-Salazar A, Carrillo R, de Miguel J. Intracerebral schwannoma in a child: report of a case. Childs Brain 1984;11(01):69–72

