Clinical Case Report

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Intraparenchymal schwannoma with calcification of the temporal lobe

Case report and literature review

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Abstract

Rationale: Intracranial schwannomas most frequently arise from the trigeminal nerve and the vestibular nerve. Schwannomas within the cerebral parenchyma are exceedingly rare. Additionally, calcification is an uncommon histopathological and radiological characteristic in schwannomas.

Patient concerns: A 46-year-old man presented to us with sudden onset epileptic seizure and a 3-month history of intermittent headache. After admission, the physical and neurological examinations were all normal. Brain CT revealed an irregular, well-defined, hyperdense mass in the right temporal lobe. MRI showed a solid mass appearing iso- to hypointensity on T1-weighted imaging and heterogeneous intensity on T2-weighted imaging in the right temporal lobe; after Gd-DTPA administration, the lesion showed heterogeneous enhancement.

Diagnosis: Histopathological examination revealed hyperchromatic nuclei and loose intercellular matrix with calcification. Immunohistochemical analysis demonstrated that the tumor was strongly positive for S100 protein but negative for GFAP and CK, which was consistent with a schwannoma.

Interventions and outcomes: A surgical resection via the right temporal approach was performed. Intraoperatively, we noticed that the tumor was grayish yellow, capsuled, and located entirely within the temporal parenchyma. A gross total resection was achieved. The postoperative course was uneventful, and there was no epileptic seizure.

Lessons: Intraparenchymal schwannoma with calcification is an uncommon histopathological and radiological characteristic in schwannomas. Intraparenchymal schwannoma with calcification is extremely rare. The early identification and appropriate surgical treatment should be highlighted.

Abbreviations: CK = cytokeratin, CT = computed tomography, CUSA = cavitron ultrasonic surgical aspirator, CYT = cytochrome, EMA = epithelial membrane antigen, Gd-DTPA = gadolinium-diethylene triamine pentaacetic acid, GFAP = glial fibrillary acidic protein, MRI = magnetic resonance imaging.

Keywords: calcification, case report, intraparenchymal, schwannoma, surgical resection

1. Introduction

Intracranial schwannomas account for 8% of all primary cerebral neoplasms.[1] The tumors most frequently involve the trigeminal nerve and the vestibular nerve, and those arising from other cranial nerves are rare. Especially, schwannomas occurring within the cerebral parenchyma are exceedingly rare.[1,2] According to literatures, approximately 70 cases of schwannomas unrelated to the cranial nerves have been reported.[1] The pathogenesis of intraparenchymal schwannomas remains unknown, as there are no Schwann cells in the cerebral parenchyma.[3] Additionally, calcification is an uncommon histopathological and radiological characteristic in schwannomas; only 16 cases of intracranial schwannomas with calcification were identified in previous reports.

Herein, we reported a case diagnosed as intraparenchymal schwannoma with calcification in the temporal lobe. Moreover, we reviewed the literatures regarding schwannomas with calcification, and the clinicoradiological features were analyzed.

2. Case presentation

A 46-year-old man presented to us with sudden onset epileptic seizure and a 3-month history of intermittent headache. After admission, the physical and neurological examinations were all normal. Brain computed tomography (CT) revealed an irregular,

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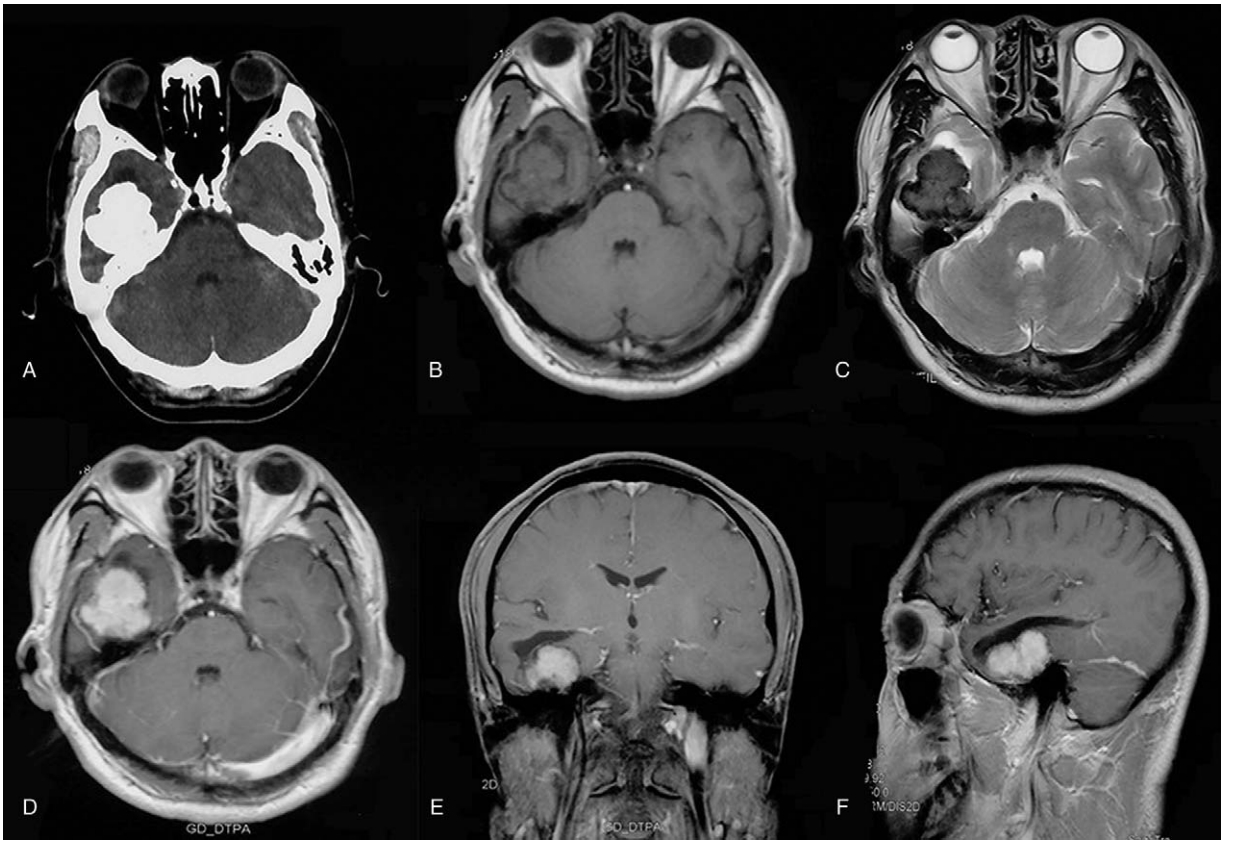

Figure 1. Radiological examinations of the patient. Axial computed tomography (CT) revealed an irregular, well-defined, hyperdense mass in the right temporal lobe (A). On magnetic resonance imaging (MRI), the lesion showed iso- to hypointensity on T1-weighted imaging (B) and heterogeneous intensity on T2-weighted imaging (C); after contrast medium administration, the lesion showed heterogeneous enhancement, and it was located entirely within the temporal parenchyma, not involving the cerebellopontine angle region (D, axial; E, coronal; F, sagittal).

well-defined, hyperdense mass in the right temporal lobe (Fig. 1A). Magnetic resonance imaging (MRI) showed a solid mass appearing iso- to hypointensity on T1-weighted imaging and heterogeneous intensity on T2-weighted imaging in the right temporal lobe; after gadolinium-diethylene triamine pentaacetic acid (Gd-DTPA) administration, the lesion showed heterogeneous enhancement; the tumor did not involve the cerebellopontine angle region (Fig. 1B–F). The operative preliminary diagnosis was meningioma with calcification, and a surgical resection via the right temporal approach was performed. Intraoperatively, we noticed that the tumor was grayish-yellow, capsuled, and located entirely within the temporal parenchyma; there was parenchymal tissue between the tumor and the dura mater, and no significant adhesion with the surrounding tissue was noted. Microscopically, the tumor was resected in piecemeal fashion and the calcified part was removed using cavitron ultrasonic surgical aspirator (CUSA); eventually, a gross total resection was achieved. Histopathological examination revealed hyperchromatic nuclei and loose intercellular matrix with calcification (Fig. 2A). Immunohistochemical analysis demonstrated the tumor was strongly positive for S100 protein (Fig. 2B) but negative for glial fibrillary acidic protein (GFAP) and cytokeratin (CK), which was consistent with a schwannoma. The Ki-67 proliferation index was less than 1%. Immediately after surgery, headache was relieved. The postoperative course was uneventful, and there was no epileptic seizure. After a 6-month follow-up, MRI showed no recurrence (Fig. 3), and the patient remained asymptomatic.

3. Discussion

We retrieved the relevant literatures, and identified 16 cases of schwannomas with calcification. The individual clinical characteristics, MRI features, and surgical outcomes were summarized in Table 1. The locations included cerebellopontine angle region (n=8), sulci olfactorius (n=4), middle cranial fossa (n=2), frontal lobe (n=1), and anterior cranial fossa (n=1). Herein, we reported the first schwannomas with calcification occurring in the temporal lobe. Including the present case, the average age is 41.7 years (range, 14–66 years). There are 9 males and 8 females, yielding a male-to-female ratio of 1:1.25. On MRI, these tumors appeared hypointensity (54.55%), iso- to hypointensity (18.18%), isointensity (9.09%), hyperintensity (9.09%), and heterogeneous intensity (9.09%) on T1-weighted imaging; the tumors appeared hyperintensity (60%), hypointensity (10%), and heterogeneous intensity (30%) on T2-weighted imaging; after contrast medium administration, the tumors appeared heterogeneous (90%) or homogeneous (10%) enhancement. Noteworthily, among these cases, there are 8 patients with calcified schwannomas occurring in the cerebral parenchyma.

The pathogenesis of intraparenchymal schwannomas is still unknown. Generally, schwannomas are considered to originate from the Schwann cells, while the Schwan cells exclusively exist

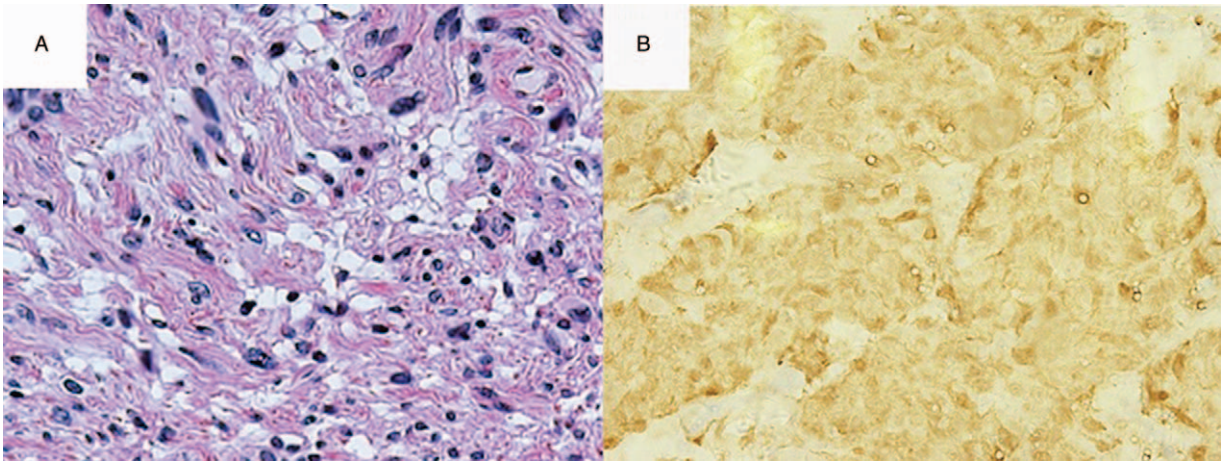

Figure 2. Histopathological examination of the patient. Hematoxylin-eosin staining revealed hyperchromatic nuclei and loose intercellular matrix with calcification (A, ×400). Immunohistochemical analysis demonstrated that the tumor was strongly positive for S100 protein (B, ×400).

outside the pia mater forming nerve sheath and there is no such cell in the cerebral parenchyma.[4] According to literatures, there are four theoretical hypotheses:

(1) intraparenchymal Schwann cells may be transformed from the pial mesodermal cells[4–6];
(2) these aberrant Schwann cells may be differentiated from the pluripotent interstitial cells[3–6];
(3) intraparenchymal schwannoma may arise from the Schwann cells existing within the perivascular nerve plexus and large arteries in the subarachnoid spaces[3,5–7];

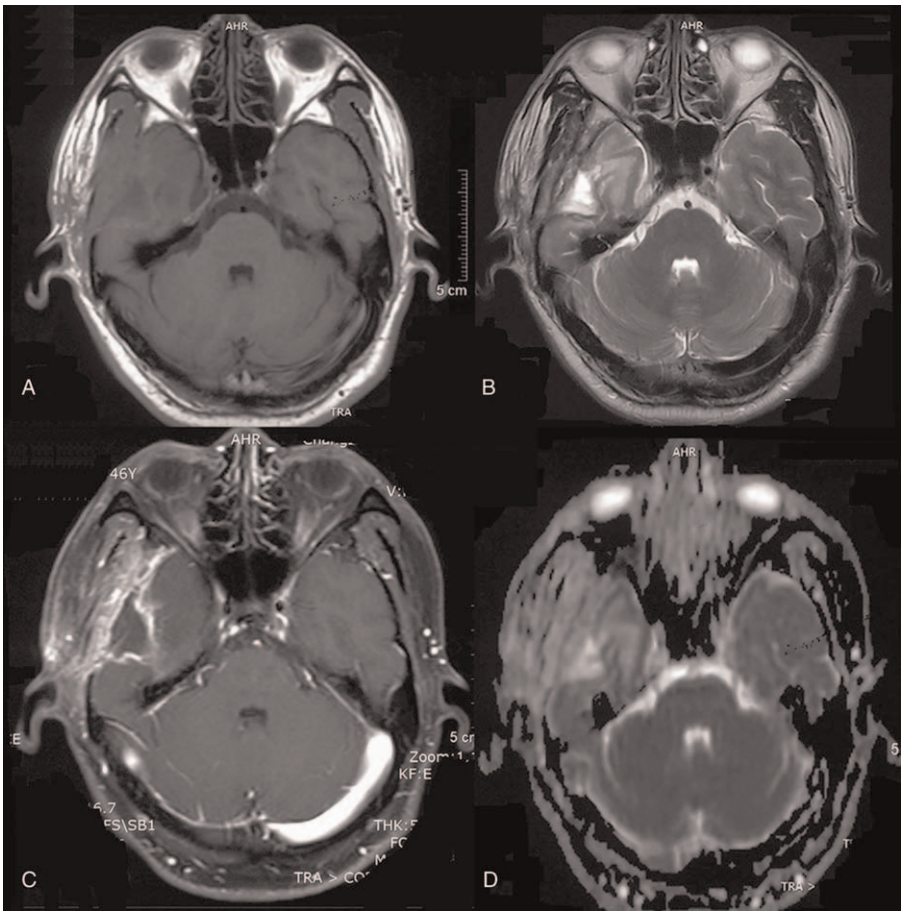

Figure 3. Follow-up magnetic resonance imaging. Axial T1-weighted (A), T2-weighted (B), contrasted (C), and diffusion-weighed (D) imaging showed no recurrence.

Table 1

The clinical and radiological profiles of schwannomas with calcification.

Author	Year	Gender	Age	Symptoms	Location	MRI features	Resection extent	Follow-up duration (month)	Recurrence
Kusumi et al[16]	2016	F	66	Vertigo	Left middle cranial fossa	T1WI: isointensity T2WI: hyperintensity Gd-DTPA: heterogeneously enhancement	GTR	N.A.	N.A.
Zhang et al[17]	2012	M	48	Hearing loss	Left cerebellopontine angle	T1WI: hypointensity T2WI: heterogeneous intensity Gd-DTPA: heterogeneously enhancement	GTR	6	No
Li[4] et al	2012	F	16	Epileptic seizure	Right frontal lobe	T1WI: N.A. T2WI: hyperintensity Gd-DTPA: heterogeneously enhancement	GTR	N.A.	N.A.
Gopalakrishnan et al[18]	2011	M	65	Hearing loss, facial numbness, asymmetry	Left cerebellopontine angle	T1WI: hypointensity T2WI: hyperintensity Gd-DTPA: heterogeneously enhancement	GTR	N.A.	N.A.
		F	31	Hearing loss, facial numbness	Right cerebellopontine angle	T1WI: hypointensity T2WI: hypointensity Gd-DTPA: homogeneously enhancement	GTR	N.A.	N.A.
Choi et al[19]	2009	F	39	Anosmia, intermittent frontal headache	Right anterior cranial fossa	T1WI: N.A. T2WI: N.A. Gd-DTPA: heterogeneously enhancement	GTR	N.A.	N.A.
Saberi et al[20]	2008	F	35	Headache, diplopia, epileptic seizure	Sulci olfactorius	T1WI: hypointensity T2WI: hyperintensity Gd-DTPA: heterogeneously enhancement	GTR	1	No
Katoh et al[21]	2007	F	59	Epileptic seizure	Left cerebellopontine angle	T1WI: heterogeneous intensity T2WI: heterogeneous intensity Gd-DTPA: heterogeneously enhancement	STR	N.A.	N.A.
Adachi et al[22]	2007	F	22	Convulsion	Sulci olfactorius	T1WI: N.A. T2WI: N.A. Gd-DTPA: heterogeneously enhancement	GTR	N.A.	N.A.
Yako et al[23]	2005	M	14	Headache, vomiting	Sulci olfactorius	T1WI: hyperintensity T2WI: N.A. Gd-DTPA: N.A.	GTR	12	No
Tosaka et al[24]	2002	M	36	Hearing loss	Left cerebellopontine angle	T1WI: iso- to hypointensity T2WI: hyperintensity Gd-DTPA: N.A.	STR	N.A.	N.A.
Bando et al[25]	1992	F	28	Visual field deficiency	Sulci olfactorius	T1WI: hypointensity T2WI: N.A. Gd-DTPA: N.A.	GTR	N.A.	N.A.
Atlas et al[26]	1992	M	50	Hearing loss	Right cerebellopontine angle	N.A.	GTR	N.A.	N.A.
Beskin et al[27]	1989	M	47	Hearing loss	Left cerebellopontine angle	T1WI: hypointensity T2WI: hyperintensity Gd-DTPA: N.A.	GTR	N.A.	N.A.
Horimoto et al[28]	1987	M	63	Facial weakness	Right middle cranial fossa	N.A.	STR	N.A.	N.A.
Thomsen et al[29]	1984	M	44	Hearing loss	Right cerebellopontine angle	N.A.	GTR	N.A.	N.A.
Present case	–	M	46	Intermittent headache, epileptic seizure	Right temporal lobe	T1WI: iso- to hypointensity T2WI: heterogeneous intensity Gd-DTPA: heterogeneously enhancement	GTR	6	No

F = female, Gd-DTPA = gadolinium-diethylene triamine pentaacetic acid, GTR = gross total resection, M = male, N.A. = not available, STR = subtotal resection, T1WI = T1-weighted imaging, T2WI = T2-weighted imaging.

(4) developmental anomaly may result in heterotopic Schwann cells.[3,7] The definitive oncogenesis mechanism still needs further research.

The clinical manifestations of intraparenchymal schwannomas are nonspecific, localization-related space-occupying symptoms. The common symptoms include headache, nausea, vomiting, sensorimotor dysfunctions, and epileptic seizures. The duration of clinical course before diagnosis mainly depends on the location and size of the tumor; the tumor involving the functional areas may be associated with relatively short duration.

MRI is the first choice for the preoperative diagnosis of intraparenchymal schwannomas. Noteworthily, for intraparenchymal schwannomas with calcification, CT has unique superiority. The MRI characteristics of intraparenchymal schwannomas are similar to those of the extraparenchymal counterparts, appearing variable intensity on T1-weighted imaging and hyperintensity on T2-weighted imaging; the tumors are usually well circumscribed with homogeneous enhancement, and cystic changes can be visible.[5]

The differential diagnosis of intraparenchymal schwannomas with calcification should include the following entities: (1) psammomatous meningiomas, which usually show calcification on CT and dural tail sign on MRI[8]; (2) oligodendrogliomas, which appear hypointensity on T1-weighted imaging and hyperintensity on T2-weighted imaging and calcification can be visible.[9]

Pathologically, intraparenchymal schwannomas exhibit dense spindle-shaped Schwann cells arranged in interlacing fascicles with hyperchromatic nuclei.[3,10] Immunohistochemical staining can facilitate the diagnosis, which are positive for S-100 protein and cytochrome (CYT), but negative for CK, GFAP, epithelial membrane antigen (EMA), desmin, and Bcl-2.[11–14] The Ki-67 proliferation indexes are usually less than 1%, suggesting a benign behavior.

Since intraparenchymal schwannomas are usually well defined with capsule and there is no adhesion with the surrounding tissue, surgical complete resection can be feasible. However, when calcification is present, the tumor may be hard in nature, which may bring challenges in tumor removal.[15] In the current case, we attempted to use CUSA for the surgical resection. In literatures, no recurrence was noted. However, we recommend a much longer follow-up.

Author contributions

Fan Chen conceived the concept and approved the final manuscript. Shuai Zhao and Junguo Cao are responsible for the design. Dawei Chen is responsible for the data analysis.
Resources: Shuai Zhao, Ying Yu, Dawei Chen.
Writing – original draft: Fan Chen.

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