



Intracranial Intracerebral Schwannoma: a Case Report and Review of the Literature

Mohamed Henia¹ · Stefan Linsler¹ · Walter J. Schulz-Schaeffer² · Steffi Urbschat¹ · Julia Becker-Ketter² · Malvina Garner³ · Joachim Oertel¹ · Ralf Ketter¹

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Abstract

Intracranial schwannomas are relatively uncommon, accounting for approximately 8% of all intracranial tumors, while intracerebral schwannomas represent an even rarer entity, responsible for roughly 1% of all intracranial schwannomas. After reviewing the relevant literature, we discussed the clinical journey of a 74-year-old woman who presented with a 3-week history of dizziness and nausea. Magnetic resonance imaging revealed a right temporal mass lesion with perifocal edema. The initial suspicion was the diagnosis of a glioblastoma or metastasis, prompting surgical intervention. During the surgery, a gross total resection of a noninvasive tumor was successfully performed. The patient's postoperative recovery was uneventful. Histopathological examination and confirmatory immunohistochemistry played a crucial role in reaching the final diagnosis of an intracerebral temporal schwannoma, highlighting the diagnostic challenges posed by radiologically indistinguishable features from metastasis and gliomas. Despite these challenges, complete surgical removal remains the most preferred treatment option, resulting in a favorable long-term prognosis without the need for adjuvant or neoadjuvant chemotherapy. Intracerebral schwannomas are exceedingly rare brain tumors, often found on the brain's surface or adjacent ventricles. Early and accurate diagnosis can be challenging due to radiological features overlapping with other intracranial pathologies. Nonetheless, histopathological examination and immunohistochemistry remain indispensable tools in establishing a definitive diagnosis and guiding effective treatment strategies. With complete surgical excision, patients with intracerebral schwannomas can expect a positive outcome and a promising long-term prognosis. Further research and case studies are warranted to enhance our understanding of these rare tumors and improve patient outcomes.

Keywords Case report · Brain tumor · Intracerebral schwannoma · Review

Abbreviations

EMA	Epithelial membrane antigen
GFAP	Glial fibrillary acidic protein
GTR	Gross total resection
N.A	Not applicable
NF2	Neurofibromatosis type 2
STAT6	Signal transducer and activator of transcription 6
STR	Subtotal resection

TOF	Time of flight
VGLL	Vestigial-like family

Introduction

Schwannomas are a type of tumor that originates from Schwann cells, which form the myelin sheath of peripheral nerves. They account for approximately 8% of all primary brain tumors, with the majority (80–90%) being located in the cerebellopontine angle in association with the vestibular nerve [1]. However, when schwannomas occur without any relation to cranial nerves, they are considered intracerebral schwannomas (IS) [2]. These tumors are extremely rare and are often associated with neurofibromatosis type 2 (NF2). These intracerebral schwannomas account for less than 1% of all intracranial schwannomas. The first documented case

✉ Mohamed Henia
Mohamed.Henia@uks.eu

¹ Department of Neurosurgery, Saarland University Medical Centre, Building 90.5, 66421 Homburg, Saarland, Germany

² Institute of Neuropathology, Saarland University Medical Centre, Building 90.5, 66421 Homburg, Saarland, Germany

³ Department of Neuroradiology, Saarland University Medical Centre, Building 90.5, 66421 Homburg, Saarland, Germany

of intracerebral schwannoma was reported by Gibson et al. in 1966 [2].

To the best of our knowledge and according to the latest literature review made in 2021, there were 150 reported cases in the literature with a majority occurring before the age of 30 and a slight male predominance. Only 7 cases of patients older than 70 years have been reported [3].

In our manuscript, we report a case of 74-year-old women with temporomesial schwannoma. This case report follows the CARE Guidelines [4].

Methods

In the present study, we report a new case of intracerebral schwannoma. In addition, we performed a literature research using the PubMed electronic database using the following medical subject heading (Mesh) terms: intracerebral schwannoma, schwannoma, brain tumor. The keywords were used separately and/or in different combinations. Only cases report of intracerebral schwannoma in old patients (≥ 70 years old) were considered.

Case Description

History and Presentation

The patient is a 74-year-old female presented after a 3-week history of dizziness and nausea. The dizziness was described as a feeling of unsteadiness, exacerbated when lying down and turning to the side while lying down, leading to accompanying nausea and vomiting. Initial evaluation revealed a right temporal mass lesion with perifocal edema on a CT scan and MRI. For further investigation, the patient was transferred to our department (Fig. 1). It is also important

to note that our patient suffered from arterial hypertension, hypercholesterolemia, glaucoma, and reflux esophagitis.

Imaging

An initial imaging (MRI cranial without contrast, MRI cranial with diffusion tensor imaging (DTI) and MRI cranial vascular angiography) confirmed the presence of a contrast-enhancing mass lesion in the right temporomesial region observed in conjunction with the pre-contrast MRI. It measured approximately 36×32 mm in the current T2 axial image. There were hypo-intensities (Fig. 2).

In the right frontal region, there were two small areas with increased signal intensity on diffusion-weighted imaging (DWI), corresponding to decreased apparent diffusion coefficient (ADC) values and increased signal on T2-weighted images.

Compared to the previous images (FIRST MRT), there were a slight increase in the size of the ventricles. Measurement of the lateral ventricle anterior horns on the axial image showed a current width of 41 mm, previously measuring 37 mm. There was no significant midline shift observed. The apical and basal cisterns were clearly delineated (Fig. 2).

The arterial time-of-flight (TOF) angiography showed no abnormalities in the anterior cerebral artery (ACA), middle cerebral artery (MCA), or internal carotid artery (ICA). The vertebral and basilar arteries appeared normal. A small outpouching was seen caudally at the transition from the right P1 to P2 segment of the posterior communicating artery (PCA), measuring approximately 2×2 mm in the TOF angiography (Fig. 2).

Surgical Procedure

The patient underwent a right temporal craniotomy. Guided by neuronavigation, a total microsurgical excision of the lesion

Fig. 1 cMRI (Mprage, T1 weighted). (A Axial, B Coronal, C Sagittal). The images show a right temporal mass lesion (red arrow) with perifocal edema (yellow arrow)



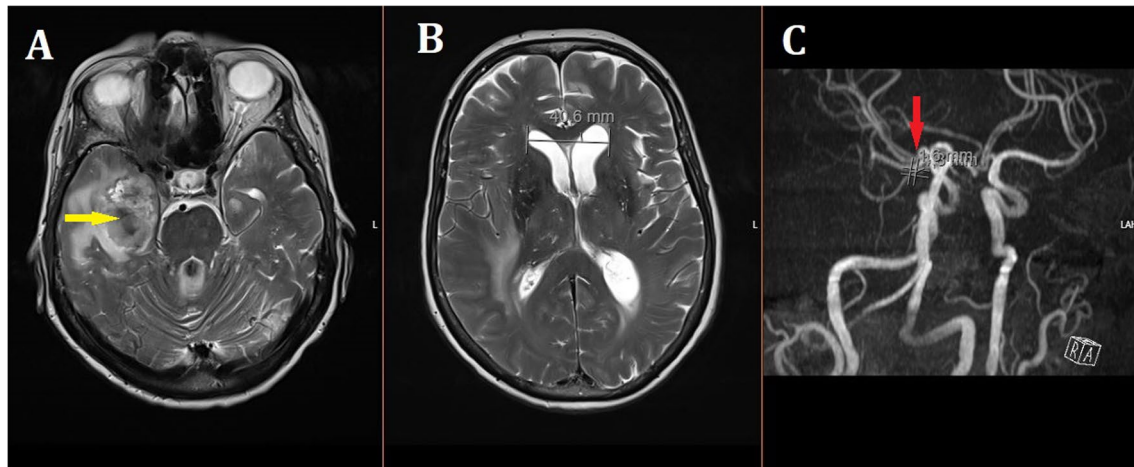


Fig. 2 cMRI (A T2 weighted MRI) shows increased signal on T2 weighted images (yellow arrow). B T2 weighted MRI shows the current size of the ventricles. C Angiography shows a small outpouching in the transition between the right P1 and P2 segment (red arrow)

was achieved. During the surgery, the tumor appeared well demarcated and firm, distinct from the brain tissue. Multiple samples were taken for histopathological examination. The tumor exhibited a high degree of vascularity and was partially firm-grayish-yellow in appearance.

Postoperative Course

After the surgery, the patient was transferred to the monitoring unit. On the following day, with stable neurological and cardiopulmonary condition, the patient was transferred to the general ward. The patient was mobilized at a rapid pace on the ward. The postoperative course was uneventful. No new neurological deficits were observed. The wound was dry and non-irritated. The postoperative cranial CT control showed a regular postoperative status without bleeding or obstruction.

Pathological Findings

The neuropathologist received 10.72 g of tissue, consisting of a large, gray, and firm tissue fragment measuring $3.9 \times 2.4 \times 0.7$ cm.

The histological sections demonstrate a densely cellular mesenchymal tumor. The tumor exhibited both, a compact and a loosely arranged, partially mucinous tissue matrix. Some areas showed a high cellularity. The tumor partly abutted vascular endothelium, while in other areas, tumor vessels demonstrated hyalinized walls and perivascular fibrous cuffs. The tumor cells showed a dense reticulin fiber network (Gomori's silver impregnation). Pleomorphic elongated cell nuclei were observed in some areas. Individual mitotic figures were present. There was no evidence of tumor tissue necrosis (Fig. 3 A, B).

In some areas, the tumor was in direct contact with brain tissue. No leptomeningeal structures were visible between the brain and tumor tissue. A sharp demarcation of the tumor bulk was seen, and the brain tissue showed a reactive astrocytic gliosis with a gemistocytic reaction. Some remaining glia cells and some axons were visible in between the tumor bulk, arguing for an infiltrative growth of the tumor (Fig. 4).

By immunohistochemical antibody staining, it could be demonstrated that the tumor cells showed a nuclear and cytoplasmic expression of S-100 (Fig. 3C), but no expression of the glial fibrillary acidic protein (GFAP) (Fig. 4C), no expression of the epithelial membrane antigene (EMA; data not shown), and no nuclear redistribution of the signal transducer and activator of transcription 6 (STAT6; data not shown). Together with the histological appearance, this identifies the tumor as a schwannoma WHO grade 1 and excludes a glioma, a meningioma, and a solitary fibrous tumor (SFT).

Genetic Findings

For further characterization of the schwannoma, an analysis of allelic losses from tumor cells on the long arm of chromosome 22 was conducted using microsatellite analysis. Four different microsatellites were analyzed for the characterization of a regional loss of chromosome 22, located between 22q11.23 and 22q13.1. The analysis was performed on a high-resolution Spreadex EL 800 gel in the Origins electrophoresis system by Elchrom Systems. The DNA extracted from the schwannoma tissue shows an allelic loss on the long arm of chromosome 22 in comparison to DNA extraction from patients' blood (Fig. 5).

To confirm these findings, fluorescence in situ hybridization (FISH) was performed in native tumor tissue.

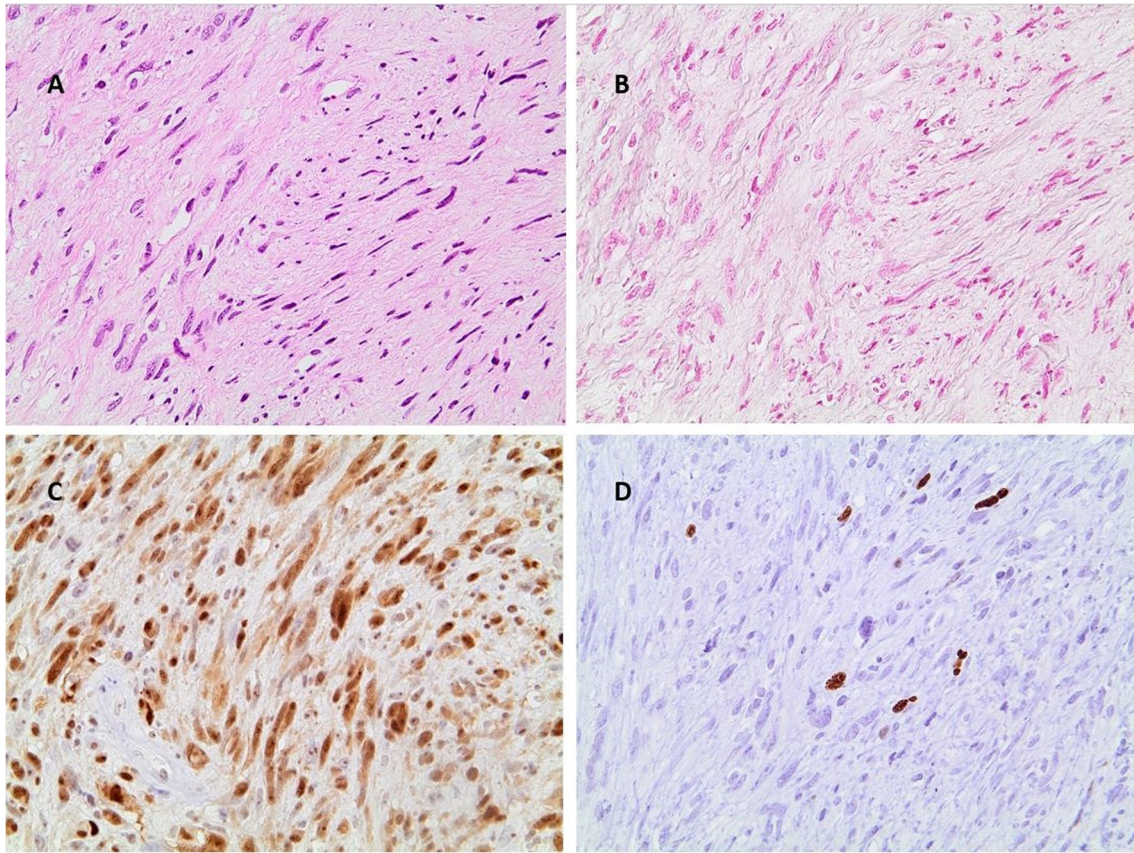


Fig. 3 Histology of the intracerebral schwannoma. The tumor is mainly composed of mesenchymal cells with elongated nuclei (H&E-stain, **A**) that exhibited subtle basal membranes, as shown by the silver impregnation according to Gomori as fine lines around the cells

(**B**). The immunohistochemical characterization is the nuclear and cytoplasmic S-100 antibody reactivity, shown by the brown color reaction (**C**). The proliferative activity, demonstrated with the antibody Ki67 (=MIB1) as nuclear brown staining is moderate (**D**)

Two-color hybridization DNA probes were used to detect the chromosomal regions 22q11.2 and 1p36.3 (MetaSystems GmbH). The procedure has been performed in previous studies in our neurosurgery research laboratory [5, 6]. Signals were analyzed with an Olympus BX43 fluorescence microscope based on the criteria of Hopman et al. [7].

The outcome of the FISH, as shown in Fig. 6, indicated the deletion of the chromosomal region 22q11.2. We could also observe an internal heterogeneity showing cells with a deletion on chromosome 22q11.2, while others showed an intact 22q11.2. Furthermore, we could demonstrate no deletions of the chromosomal region 1p36.3 in the investigated cells (Fig. 6). Taken together, this suggests that some tumor cells are affected by extensive loss of chromosome 22q, ranging, at least, from 22q11.2 to 22q12.3.

Literature Review

A total of 6 articles describing 7 patients were included in our study (Table 1) [3, 8–12]. Among these seven patients (male = 2, female = 5), three underwent a subtotal resection of the tumor, and the rest underwent complete resection or gross total resection of the tumor. One of the patients was treated, after the gross tumor resection, by intensity-modulated radiation therapy (IMRT) with 37.5 Gy, delivered to the resection cavity in 15 fractions because a malignant intracerebral nerve sheath tumor was diagnosed. This entity is extremely rare compared to benign intracerebral schwannoma [12]. The vast majority of tumor cells show a lack of differentiation by S-100 negativity.

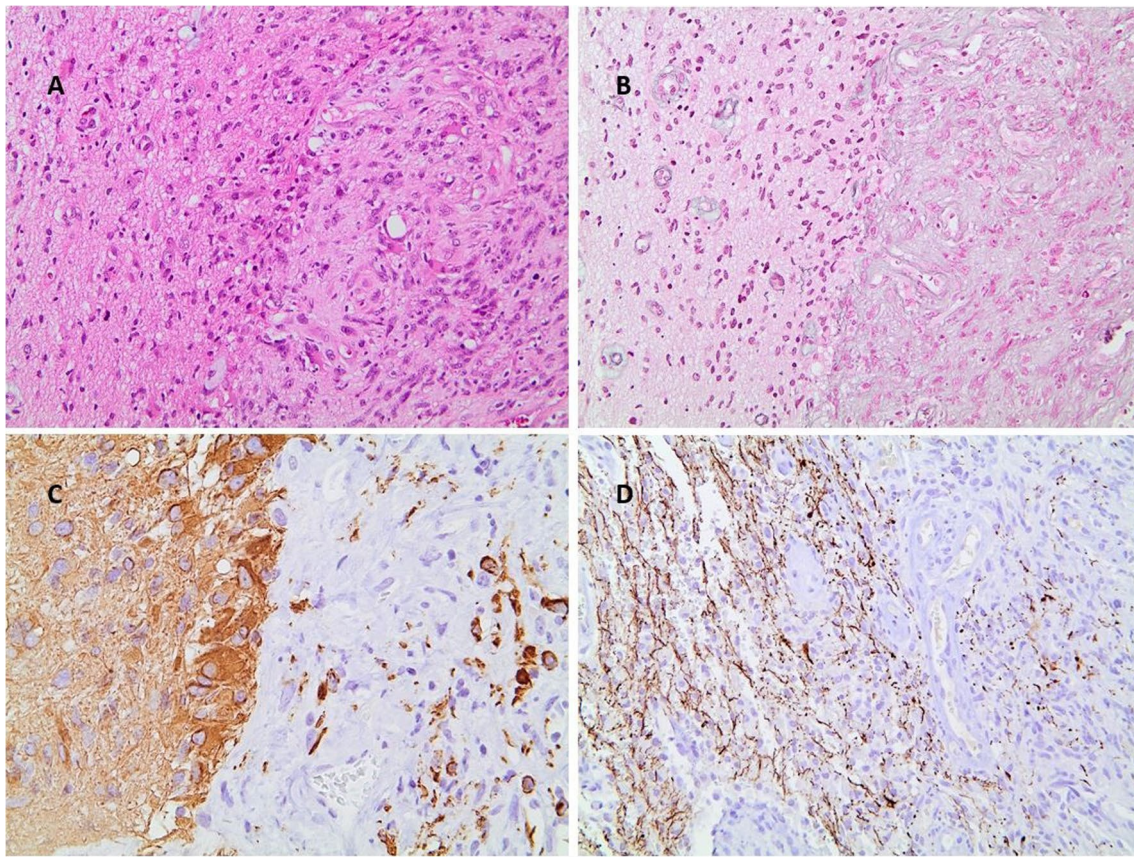


Fig. 4 Sharp border between brain tissue (left) and tumor bulk (right, H&E stain, **A**). Whereas the silver impregnation according to Gomori demonstrates the subtle basal membranes in the tumor tissue, the brain tissue is free of it (**B**). Densely arranged clumsy astrocytes, immunoreactive for the glial fibrillary acidic protein (GFAP, brown

color), are seen in the brain tissue (left) adjacent to the tumor (right) and as single cells in the tumor (**C**). Whereas a dense axon staining is seen in the brain tissue using an antibody against neurofilament, only some axons are visible in the tumor tissue (**D**)

The reason for a subtotal resection was the tumor location. Weiner et al. reported a tumor intimately adjoining the brainstem [10], and Casedei et al. reported tumors extending to the medulla and into basal ganglia [8].

Discussion

Schwannomas originating from Schwann cells, which typically form the myelin sheath of peripheral nerves, are commonly found in cranial and peripheral nerves. However, intracerebral schwannomas are rare and have attracted speculation regarding their origin [8]. Various theories have been proposed to explain the development of this tumor, categorized as developmental and non-developmental theories. The developmental theory suggests distorted embryogenesis as the source of aberrant foci of Schwann cells in the brain parenchyma. This may involve the transformation of developed mesenchymal pial cells into Schwann cells, differentiation of multi potential mesenchymal elements, ectopic migration of neural crest cells, or misplaced

myelinated nerve fibers [13, 14]. Conversely, the non-developmental theory proposes that intracerebral Schwann cells arise from the perivascular nerve plexus of parenchymal arterioles [15]. Schwannomas are frequent in patients carrying an autosomal dominant mutation in the gene *NF2*, which codes for the tumor-suppressor protein merlin (Moesin-Ezrin-Radixin-like protein). The gene of this protein is located on chromosome 22q.12.2 in proximity to the microsatellite marker D22S268. With two independent methods, we can demonstrate a somatic heterozygote loss of chromosome 22q in the tumor of the present patient, which may contribute to the tumor development. In the case of intracerebral schwannoma, which comprises less than 1% of surgically treated central and peripheral nervous system schwannomas, the lesions are predominantly supratentorial, either superficial or periventricular [3]. Most cases in the last literature review have been reported in younger individuals, with the majority presenting below the age of 30 [3]. Uncommon locations of ectopic schwannomas have also been described, such as the third and fourth ventricles, intrasellar region, and

Fig. 5 PCR-based microsatellite analysis reveals heterozygous allelic losses in the tumor tissue in for at least 3 (D22S445, D22S684, D22S268) out of 4 microsatellite markers (A). The location of the microsatellites is indicated by black lines on chromosome 22 (B). Legend: B, blood DNA; T, tumor DNA, M, marker of size

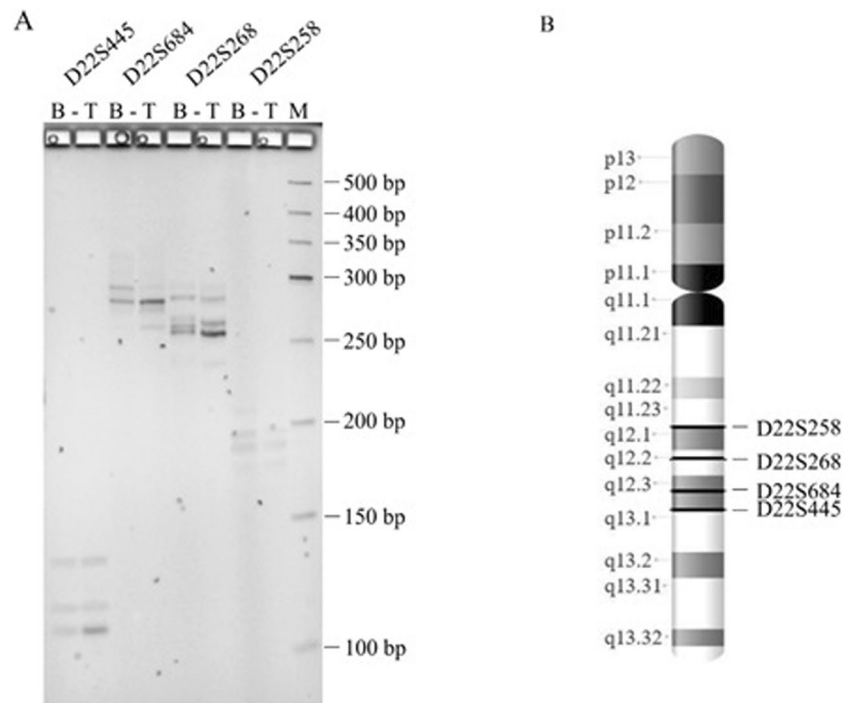
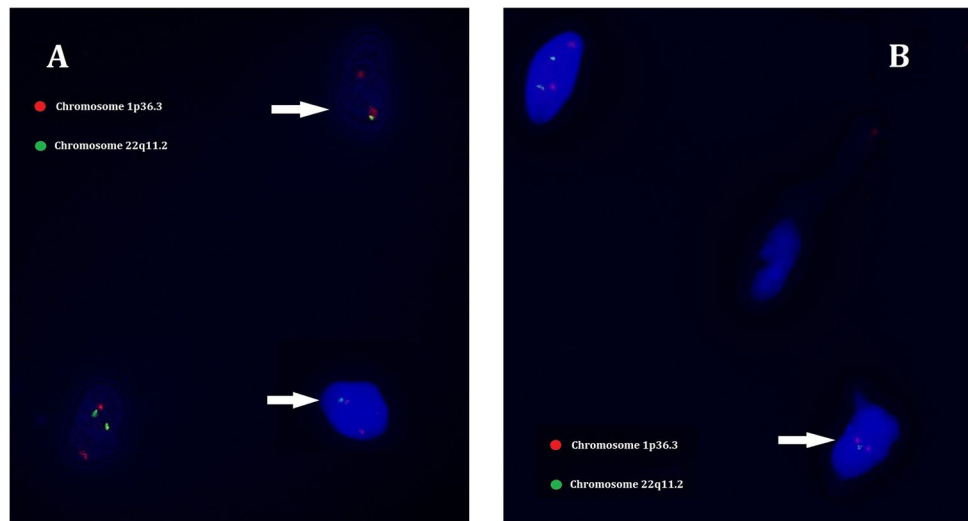


Fig. 6 Our FISH findings showing one normal cell and two heterozygous deletions of 22q11.2 (white arrow) (A) and also one normal cell and one heterozygous deletion of 22q11.2 (white arrow) (B)



pontomedullary cistern with extension into the dorsal spinal cord. Radiological features of intracerebral schwannoma on CT and MRI include cystic formations, calcifications, and peritumoral edema. However, these imaging findings are not pathognomonic, leading to occasional misdiagnoses, such as mistaking schwannoma for meningiomas based on tentorial involvement and the presence of a dural tail sign [16, 17]. Actually, the diagnosis of this intracerebral schwannoma is almost never made preoperatively.

Histopathological analysis is crucial for distinguishing IS from other tumors, such as pilocytic astrocytoma, pleomorphic xanthoastrocytoma, ganglioglioma, solitary fibrous tumors,

and meningioma. Differential diagnosis can be challenging, particularly between fibroblastic meningioma, the solitary fibrous tumor and an Antoni A cell-predominant schwannoma, or pilomyxoid/pilocytic astrocytoma and Antoni B cell-predominant schwannoma. Additional diagnostic tools, i.e., immunohistochemical analyses, are necessary to differentiate these entities. Intracerebral schwannoma typically exhibits positive staining for S-100, SOX10, and calretinin, while staining negative for EMA, GFAP, and STAT6 [18–20]. In addition, in the current epigenetic Heidelberg Brain Tumor Classifier v12.8, “MC CNS Schwannoma, VGLL-fused” is introduced as a provisional new class. The editors of this brain

Table 1 Summary of eight cases (including our case) of intracerebral schwannomas in old patients

Authors and year	Sex and age	Symptoms	Localization	Histological findings	Radiological diagnosis	Treatment
Casadei et al. [8]	F, 84	Hemiparesis	Temporal	S-100 +, GFAP +, EMA-	N.A.	STR
Casadei et al. [8]	F, 79	Ataxia	Cerebellar	S-100 +, GFAP +, EMA-	N.A.	STR
Sharma et al. [9]	F, 73	Gait disturbance, headache, vomiting	Cerebellar	S-100 +, GFAP -, EMA-	N.A.	GTR
Weiner et al. [10]	F, 78	Facial nerve spasm diplopia	Brainstem	S-100 +, GFAP -	Ependymoma, glioma, plexus papilloma	STR
Luo et al. [11]	M, 72	Hemiparesis, headache	Parieto-occipital	S-100 +, GFAP -	N.A.	CR
Barnard et al. [12]	F, 75	Personality changes and dysphasia	Frontal	S-100 -, GFAP -, EMA-, CD34 -, a-SMA -	Glioblastoma	GTR + RT
Kovalainen et al. [3]	M, 74	Gait disturbance	Pons	S-100+	Hemangioblastoma	GTR
Current case	F, 74	Dizziness and nausea	Temporal	S-100 +, GFAP -, EMA-, STAT6	Hemangioblastoma	GTR

tumor classifier explained that this kind of tumor represents a very rare group of intracranial tumors with morphological and immunohistochemically features of schwannoma. These tumors often display fusions of the VGLL gene family (VGLL2 or VGLL3), which have also been reported for a subset of peripheral schwannoma. This type of schwannoma is a provisional class, which is still being further characterized [21].

For intracerebral schwannoma, surgery is the primary treatment, with complete resection often resulting in a favorable long-term outcome. Total removal of the tumor is typically curative, as evidenced in cases where long-term follow-up demonstrated no recurrence. A recurrence rate of 5.3% has been documented after GTR primarily attributed to the presence of malignant histopathology within the tumor [3]. In cases that underwent subtotal resection, 20% of the tumors with no malignant pathology needed a second surgery, proposing that recurrence is due to incomplete tumor removal [3]. In the presented case, total tumor removal was achieved, as confirmed by postoperative CT scan.

Conclusion

In conclusion, intracerebral schwannoma represents a rare variant of schwannoma, characterized by its intracerebral location and atypical features. The origin of these tumors remains an area of ongoing research, with both developmental and non-developmental theories proposed. Accurate diagnosis requires careful evaluation of histopathological features and the use of ancillary diagnostic techniques. Surgical resection is the preferred treatment approach, aiming for complete removal whenever possible, which often leads to favorable long-term outcomes. Further studies are warranted to elucidate the underlying mechanisms and optimal management strategies for this rare entity.

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Code Availability Not applicable

Author Contributions Scientific support and paper writing: MH (1) Surgery and paper writing: SL (1) FISH based analysis and paper writing: SU (1) Pathological analysis and LOH: WJS-S, JB-K (2) Magnet resonance imaging: MG (3) Principal investigator: JO (1) Idea, scientific support, statistical analysis, surgery and paper writing: RK (1)

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Data Availability All data underlying the results are available as part of the article and no additional source data are required.

Declarations

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

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

Conflict of Interest The authors declare no competing interests.

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