

Radiologic and Pathologic Findings of Intracerebral Schwannoma

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Summary: We report the radiologic and pathologic findings of an intracerebral schwannoma. MR imaging studies showed a superficially located cystic mass with an enhancing nodule and evidence of peritumoral edema or gliosis.

Intraparenchymal schwannomas of the brain and spinal cord are rare. Gibson et al (1) reported the first English-language account of an intraparenchymal intracerebral schwannoma in 1966. Since then, at least 33 additional cases of this neoplasm have been reported, but little has been written about its imaging appearance (2-24). We describe the MR imaging and pathologic findings in a patient with intracerebral schwannoma and, through a review of the literature, discuss the characteristic imaging features of this neoplasm.

Case Report

A 15-year-old boy was admitted to the hospital after accidental head trauma with loss of consciousness. His family history was noncontributory and there was no evidence of neurofibromatosis. Physical examination revealed a left-sided parietal scalp hematoma and laceration without focal neurologic deficit. Laboratory findings were normal. MR imaging studies showed a heterogeneous right-sided parietal mass (Figs 1A-D). The patient underwent a stereotactic biopsy, which yielded amber-colored cystic fluid. Microscopic analysis of solid tissue showed a benign spindle-cell neoplasm, consistent with a low-grade tumor, possibly a pilocytic astrocytoma. Ten days later, the patient underwent a right-sided parietooccipital craniotomy with resection of the tumor. Sonography was used to identify a solid nodule located immediately beneath the cortical surface with a prominent cystic component along the posterolateral aspect of the nodule. The nodule was gently separated from the overlying brain and was removed in one piece. The cyst, which contained dark-greenish fluid, was decompressed. There was no visible evidence of residual tumor. Microscopic analysis of the tissue showed areas of nuclear palisading, typical of a schwannoma. Densely cellular tumor alternating with loosely textured myxoid tumor was present in equal portions, consistent with Antoni type A and Antoni type B tissue, respectively. A distinct interface between tumor and surrounding brain was present in some sections (Fig 1E). Immunohistochemical testing for S-100 protein was diffusely positive, whereas glial fibrillary acidic protein (GFAP) was negative in the tumor cells, confirming the diagnosis of schwannoma. An MR imaging study performed 1 day later showed no evidence

of residual tumor. The patient remains asymptomatic 2½ years later.

Discussion

Intraparenchymal schwannomas of the brain are rare neoplasms, with only 35 cases reported over the last 30 years. Careful analysis of clinical and radiologic findings of these cases, including one of our own, has disclosed several characteristic features. Unlike vestibular schwannomas, those in the intraparenchymal brain exhibit no female predominance (female 17, male 18) (25). Furthermore, the majority of cases have occurred in children and young adults, with median and average ages at presentation of 21 and 29 years, respectively. Seventy percent of the reported tumors were clinically apparent before patients had reached the age of 30. In contrast, vestibular schwannomas rarely occur in children unless in association with neurofibromatosis type 2 (26). The most common signs and symptoms of intraparenchymal schwannomas are headaches, seizures, and focal neurologic deficits.

Characteristic pathologic and imaging features include calcification, cyst formation, peritumoral edema and/or gliosis, and superficial or periventricular location. Calcification, believed by some to be a characteristic finding in intracerebral schwannomas (21), has been seen radiologically or confirmed histopathologically six times. In contrast, calcification has rarely been seen radiologically or histopathologically in vestibular schwannomas (27). Cysts have been detected on MR images in 20% of vestibular schwannomas (28). Cysts were documented pathologically or by imaging studies in 19 of the cases we reviewed. The cystic component of the tumor may be central and secondary to necrosis. Alternatively, as seen in our case, the cyst may be large and located peripherally, with an associated mural nodule. These extramural/arachnoid cysts may be caused by mechanical trapping of CSF or by leakage of hemorrhagic material from the tumor, causing adhesions and secondary arachnoid cyst formation (28). Extramural cysts seen on MR imaging studies in 13% of acoustic schwannomas were confirmed pathologically

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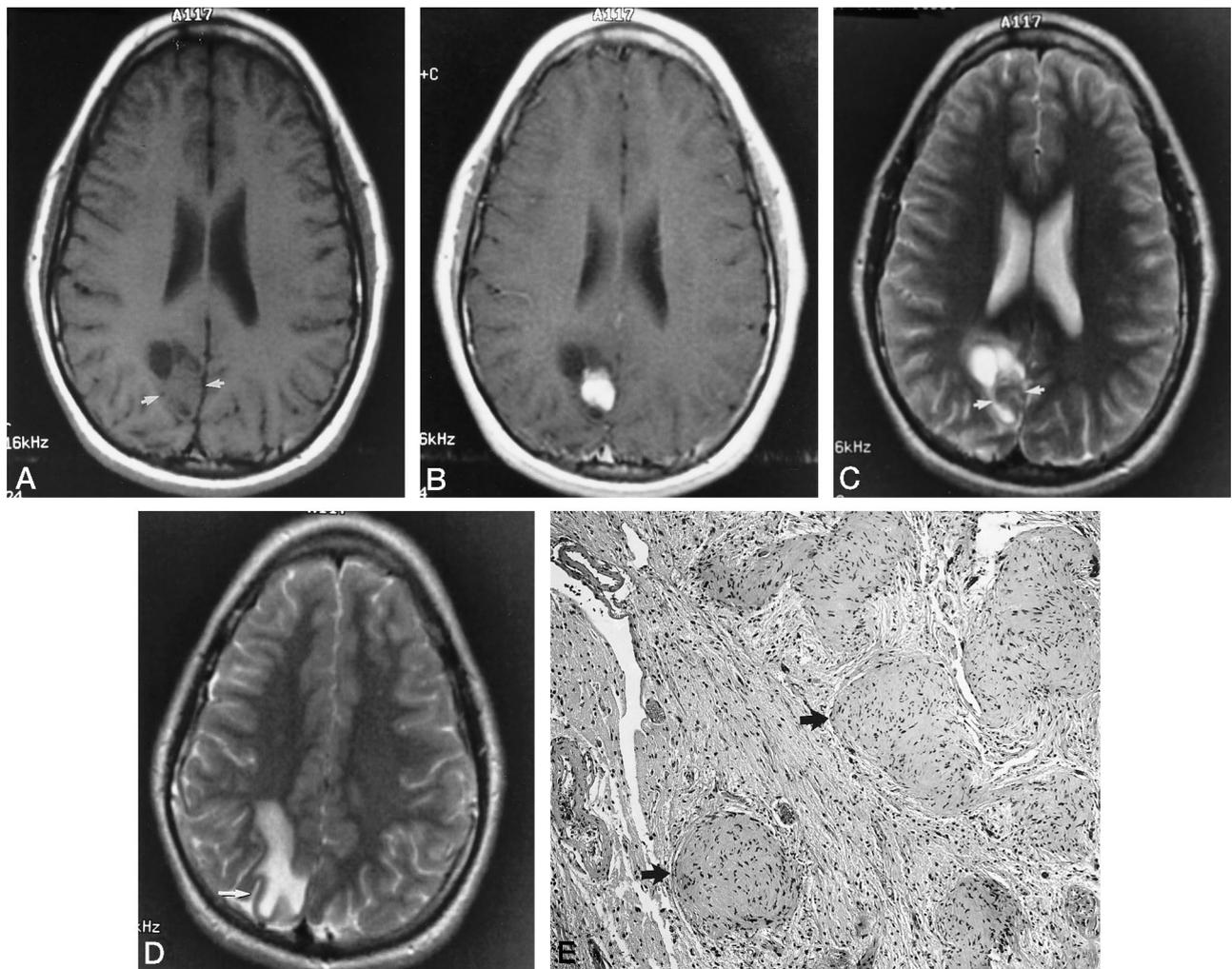


FIG 1. 15-year-old boy who lost consciousness after accidental head trauma.

A, Axial T1-weighted (650/11/1 [TR/TE/excitations]) MR image shows mixed solid/cystic mass in right parietal lobe (precuneus). Solid component (arrows) is superficially located and isointense with cortex.

B, Axial contrast-enhanced T1-weighted (650/11/1) MR image shows intense homogeneous enhancement of solid component. Cystic wall does not enhance.

C, Axial T2-weighted (3500/85/1) MR image shows mixed hypointensity/hyperintensity within solid portion of tumor with minimal central hyperintensity (arrows).

D, Axial T2-weighted (3500/85/1) MR image shows T2 hyperintensity, representing edema and/or gliosis. Note enlarged sulcus (arrow), indicating a component of gliosis and atrophy.

E, Photomicrograph shows the tumor/brain interface. Note the nests of benign Schwann cells extending into the brain in fingerlike projections (arrows). The intervening neural parenchyma was edematous and gliotic (original magnification $\times 100$).

or on imaging studies in 10 intracerebral schwannomas. The solid component of intracerebral schwannoma was found superficially or adjacent to the ventricular system in 12 (41%) of 29 cases (six cases were omitted because of data restrictions) and seven (20%) of 35 cases, respectively.

MR or CT studies were performed in 27 of the 35 reported cases. The presence of hypodensity or hyperintensity, representing edema and/or gliosis, was specifically addressed in 22 patients and was found in 17. In our case, peripheral T2 hyperintensity was present with focal prominence of a sulcus (Fig 1D), suggesting atrophy, while the T2 signal abnormalities perhaps represented a combination of edema and gliosis. The cause of the edema surrounding this benign neoplasm is uncertain. Vascular endothelial

growth factor (VGEF) messenger RNA expression has been implicated in edema formation around some meningiomas and may be an important etiologic factor explaining peritumoral edema in other benign neoplasms (29). The presence of VGEF in intracerebral schwannomas has not been confirmed however, and its role in edema formation requires further investigation. Chronic edema can result in degeneration of white matter and gliosis, possibly explaining the frequent association of these findings (30). Of the 27 patients who had MR or CT studies, 21 received contrast material, and the pattern of enhancement was specifically addressed in 20; in 10 of these, the pattern was described as homogeneous.

MR imaging findings of intracerebral schwannoma are varied. DiBiasi et al (23) reported a 1.5-cm well-

circumscribed mass located within the superficial frontal lobe showing hypointense T1 signal and hyperintense T2 signal that enhanced intensely and homogeneously. A 5-cm cystic frontal lobe mass with a solid, homogeneously enhancing component has been reported (21). The solid portion showed T1 hypointensity and mixed T2 hypointensity/hyperintensity. Calcification, seen on CT scans, may have contributed to the hypointense T2 signal. Similar T2 hypointensity was seen in our case and may be explained by the presence of hyalinized stroma and collagen deposition seen at microscopic examination. Cystic and solid brain stem and inferior vermian schwannomas have also been described (15, 20, 22, 24).

The origin of intracerebral schwannomas is uncertain. Schwann cells have been detected around arteries in the subarachnoid space and within the periphery of the brain along perivascular nerve plexus (31). Similar tiny myelinated nerves may reside in the tela choroidea, the embryologic anlage of the choroid plexus. The location of these nerves may help to explain the predilection of the reviewed cases for superficial or deep periventricular sites. The existence of Schwann cells deep within the substance of the human brain remains controversial. The only evidence of the presence of nonneoplastic Schwann cells in brain tissue is the rarely observed peripheral-type myelin in multiple sclerosis plaques or at the edge of old infarcts (32). These Schwann cells may undergo neoplastic conversion (33). Russell and Rubinstein (25) noted a close resemblance of mesodermal pial cells to neuroectodermal Schwann cells, hypothesizing that pial cells may sometimes undergo conversion to Schwann cells, thus serving as the origin of some intracerebral schwannomas.

The differential diagnosis of an intracerebral schwannoma includes several other neoplasms that may occur in children and young adults. These include pilocytic astrocytoma, pleomorphic xanthoastrocytoma, and ganglioglioma. Most pilocytic astrocytomas are discovered during the first two decades of life, and unlike in intracerebral schwannomas, surrounding edema is rarely present (34). Pleomorphic xanthoastrocytoma is a typically benign supratentorial astrocytoma occurring primarily in children and young adults. It affects males and females equally. MR imaging characteristics are variable, with T1 isointensity and hypointensity and mixed T2 isointensity and hyperintensity (35). Similar to intracerebral schwannomas, cysts and mild to moderate edema are common. This neoplasm is also located superficially, involving the cortex and leptomeninges, and may be firmly attached to the dura; however, calcification is rarely seen on CT scans or confirmed histopathologically, which may help differentiate pleomorphic xanthoastrocytoma from intracerebral schwannoma. Ganglioglioma is a rare neuronal tumor most commonly found within the temporal lobes. Cysts, contrast enhancement, and calcification may be present (36, 37), and its appearance may be nearly identical to the tumor seen in our case (38).

Conclusion

Intracerebral schwannoma is a rare, benign neoplasm. It is usually located superficially or adjacent to a ventricle. Characteristic imaging features include cyst formation, calcification, and evidence of peritumoral edema or gliosis. The recognition of this benign and potentially curable neoplasm and its differentiation from other neoplasms, some of which have less favorable outcomes, is of obvious importance.

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References

1. Gibson AAM, Hendrick EB, Conen PE. **Intracerebral schwannoma: report of a case.** *J Neurosurg* 1966;24:552-557
2. New PFT. **Intracerebral schwannoma: case Report.** *J Neurosurg* 1972;36:795-797
3. Ghatak NR, Norwood CW, Davis CG. **Intracerebral schwannoma.** *Surg Neurol* 1975;3:45-46
4. Van Rensburg MJ, Proctor NSF, Danziger J, Orelowitz MS. **Temporal lobe epilepsy due to an intracerebral schwannoma: report of a case.** *J Neurol Neurosurg Psychiatry* 1975;38:703-709
5. Pialat J, Sindow M, Courjon J, Tommosi M, Mansuy L. **Un cas de neurinome intra-cerebral frontal.** *Lyon Med* 1975;234:129-134
6. Komminoth R, Sokie P, Florange W. **Schwannoma intra-cerebelleux.** *Neurochirurgie* 1977;23:81-88
7. Prakash B, Roy S, Tandon NP. **Schwannomas of the brainstem: case report.** *J Neurosurg* 1980;53:121-123
8. Kasantikul V, Brown WJ, Cahan LD. **Intracerebral neurilemmoma.** *J Neurol Neurosurg Psychiatry* 1981;44:1110-1115
9. Shalit MN, Toledo E, Sandbank IU. **Intracerebral schwannoma.** *Acta Neurochir* 1982;54:253-258
10. Auer R, Budny J, Drake C, Ball M. **Frontal lobe perivascular schwannoma.** *J Neurosurg* 1982;56:154-157
11. Bruni P, Esposito, Greco R, Oddi G. **Solitary intracerebral schwannoma in von Recklinghausen's disease.** *Surg Neurol* 1984;22:360-364
12. Gokay H, Izgi N, Bartos O, Erseven G. **Supratentorial intracerebral schwannomas.** *Surg Neurol* 1984;22:69-72
13. Rodriguez-Salazar A, Carillo RM, de Miguel J. **Intracerebral schwannoma in a child: report of a case.** *Childs Brain* 1984;11:69-72
14. Sarkar C, Mehta V, Roy S. **Intracerebellar schwannoma: case Report.** *J Neurosurg* 1987;67:120-123
15. Aryanpur J, Long DM. **Schwannoma of the medulla oblongata: case report.** *J Neurosurg* 1988;69:446-449
16. Schwartz A. **Intracerebral and intracerebellar neurilemmoma.** *South Med J* 1988;81:385-389
17. Rhouma B, Bouzakoura CH, Bondaouara MA, Mhiri C, Hentati K. **Schwannome intra-cerebral: a propos diun case.** *Neurochirurgie* 1988;34:123-127
18. Ladouceur D, Bergeron D, Lamarche J, Lamontagne L. **Cystic schwannoma of the brainstem.** *J Can Sci Neurol* 1989;16:357-360
19. Bennazza A, Houtteville J, Chapon F, Khouri S, Hubert P. **Schwannome intra-cerebelleux: apropos d'un cas.** *Revue de la litterature.* *Neurochirurgie* 1989;35:246-250
20. Redekop G, Elisevich K, Gilbert J. **Fourth ventricular schwannoma.** *J Neurosurg* 1990;73:777-781
21. Ezura M, Ikeda H, Ogawa A, Yoshimoto T. **Intracerebral schwannoma: case report.** *Neurosurgery* 1992;30:97-100
22. Tran-Dinh H, Soo Y, O'Neil P, Chaseling R. **Cystic cerebellar schwannoma: case report.** *Neurosurgery* 1991;29:296-300

23. DiBiasi C, Trasimeni G, Iannilli M, Polettini E, Gualdi G. **Intracerebral schwannoma: CT and MR findings.** *AJNR Am J Neuroradiol* 1994;15:1956-1958
24. Casadei G, Komori T, Scheithauer B, Miller G, Parisi J, Kelly P. **Intracranial parenchymal schwannoma: a clinicopathological and neuroimaging study of nine cases.** *J Neurosurg* 1993;79:217-222
25. Russell DS, Rubenstein LJ. *Pathology of Tumours of the Nervous System.* 5th ed. London: Edward Arnold; 1989: 100-109
26. Allcut DA, Hoffman HJ, Isla A, Becker LE, Humphreys RP. **Acoustic schwannomas in children.** *Neurosurgery* 1991;29:14-18
27. Thomson J, Klinken L, Tos M. **Calcified acoustic neurinoma.** *J Laryngol Otol* 1984;98:727-732
28. Tali E, Yuh W, Nguyen H, Feng G, Koci T, Jinkins J, Robinson R, Hasso A. **Cystic acoustic schwannomas: MR characteristics.** *AJNR Am J Neuroradiol* 1993;14:124-127
29. Kalkanis SN, Carroll RS, Zhang J, Zamani AA, Black PM. **Correlation of vascular endothelial growth factor messenger RNA expression with peritumoral vasogenic cerebral edema in meningiomas.** *J Neurosurg* 1996;85:1095-1101
30. Feigen I, Popoff N. **Neuropathological changes late in cerebral edema: the relationship to trauma, hypertensive disease and Binswanger's encephalopathy.** *J Neuropathol Exp Neurol* 1963;22: 500-511
31. Nelson E, Rennels M. **Innervation of the intracranial arteries.** *Brain* 1970;93:475
32. Feigen I, Ogata J. **Schwann cells and peripheral myelin in central nervous tissues: the mesenchymal character of Schwann cells.** *J Neuropathol Exp Neurol* 1971;30:603-612
33. Stefando SZ, Vuzeuski VD, Moos AR, et al. **Intracerebral malignant schwannoma.** *Acta Neuropathol* 1986;71:321-325
34. Lee YY, Van Tassel P, Bruner JM, Moser RP, Share JC. **Juvenile pilocytic astrocytomas: CT and MR characteristics.** *AJNR Am J Neuroradiol* 1989;10:363-370
35. Lipper MH, Eberhard DA, Phillips CD, Vezina LG, Cail WS. **Pleomorphic xanthoastrocytoma, a distinctive astroglial tumor: neuroradiologic and pathologic features.** *AJNR Am J Neuroradiol* 1993;14:1397-1404
36. Otsubo H, Hoffman H, Humphrey R, et al. **Detection and management of gangliogliomas in children.** *Surg Neurol* 1992;38:371-378
37. Castillo M, Davis P, Takei Y, Hoffman JC Jr. **Intracranial ganglioglioma: MR, CT, and clinical findings in 18 patients.** *AJNR Am J Neuroradiol* 1990;11:109-114
38. Hashimoto M, Fujimoto K, Shinoda S, Masuzawa T. **Magnetic resonance imaging of ganglion cell tumors.** *Neuroradiology* 1993; 35:181-184