

Intracerebral Schwannoma—A Rare Cause of Epilepsy

S M Chng,*^{MBBS, FRCR}, C C T Lim,**^{FAMS, FRCR, M Med (Diag Radiol)}, W T Seow,***^{FAMS, MBBS, FRACS (Neurosurg)}

Abstract

Introduction: We report a case of schwannoma occurring in an intra-axial location causing seizures. **Clinical Picture:** A 13-year-old girl presented with a 4-month history of seizures. Preoperative computed tomography scan and magnetic resonance imaging revealed an enhancing right frontal lobe tumour with surrounding oedema. **Treatment:** The patient underwent craniotomy and a well-circumscribed tumour was completely resected. Histology with immunohistochemistry confirmed the diagnosis of an intracerebral schwannoma. **Outcome:** The patient made a complete recovery and was seizure-free at 1-year follow-up. **Conclusion:** Intracerebral schwannomas are rare, benign tumours. They may mimic astrocytomas or malignant tumours, and it is important to differentiate them from other neoplasms with less favourable outcomes. The neuroimaging features are, however, non-specific and definitive diagnosis can only be made on histology.

Ann Acad Med Singapore 2003; 32:547-9

Key words: Brain tumour, MR imaging, Schwannoma, Seizure

Introduction

Schwannomas are extra-axial neoplasms derived from the nerve sheath of peripheral or cranial nerves. They represent approximately 8% of all intracranial tumours, arising predominantly from the vestibular nerve.¹ Other less frequent locations include the 5th, 7th, 9th and 10th cranial nerves, sometimes associated with Von Recklinghausen's disease.² Intra-axial parenchymal schwannomas of the brain, not associated with cranial nerves, are very rare. We report a child with intracerebral schwannoma presenting with epilepsy and discuss the imaging features of this rare neoplasm. Although unusual, this benign tumour should be differentiated from other neoplasms with less favourable outcomes.

Case Report

A 13-year-old girl presented with a 4-month history of generalised tonic clonic seizures. She had no previous medical history. Clinical examination revealed no focal neurological deficit and no cutaneous signs. Computed tomograph (CT) scan of the brain revealed a 2-cm diameter, enhancing lesion in the right frontal lobe. On magnetic resonance (MR) imaging, the lesion was hyperintense on T2-weighted images (Fig. 1a) and isointense on T1-weighted

images (Fig. 1b) with a small central cystic area. There was no associated calcification or haemorrhage. Images in multiple planes clearly showed the tumour to be intra-axial, separated from the meningeal surfaces by normal brain parenchyma. Surrounding vasogenic oedema was prominent (Fig. 1c) and intense and uniform enhancement was noted after intravenous contrast injection (Fig. 1d). The patient underwent craniotomy and a discrete, multi-lobulated intra-axial tumour with well-defined planes was completely resected. Histology showed a 2.8 x 2.5 x 1.2-cm whitish tumour comprising intersecting compact fascicles of spindle cells with wavy nuclei. Final diagnosis of intracerebral schwannoma was made on immunohistochemistry, which revealed strong reactivity for S100 protein. The epithelial membrane antigen stain was negative. The patient made a complete recovery and is seizure-free at 1-year follow-up. Repeat imaging 2 months after resection showed no residual enhancing tumour. The white matter hyperintensity on T2-weighted images had also resolved.

Discussion

Schwannomas arising as intraparenchymal brain lesions without any connection to the peripheral nervous system are rare. Fewer than 50 cases have been reported in the

* Associate Consultant

** Consultant

Department of Neuroradiology

*** Senior Consultant

Department of Neurosurgery

National Neuroscience Institute

Address for Reprints: Dr Chng Soke Miang, Department of Neuroradiology, National Neuroscience Institute, 11 Jalan Tan Tock Seng, Singapore 308433.

Email: soke_miang_chng@ttsh.com.sg

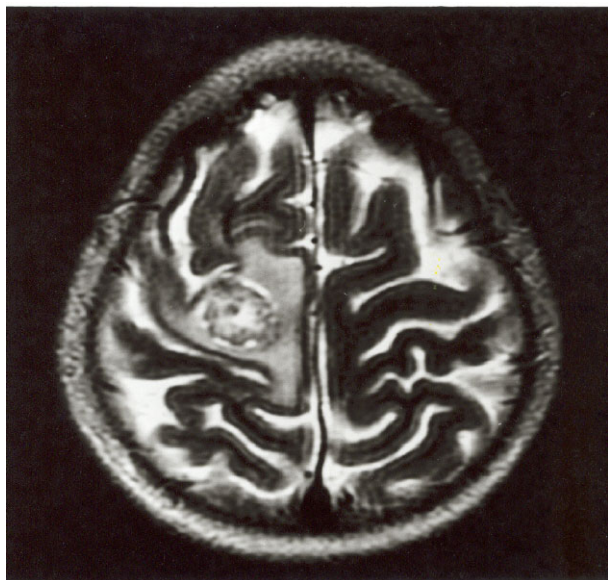


Fig. 1A. Axial T2-W MR image shows a tumour which is hyperintense to normal brain parenchyma.

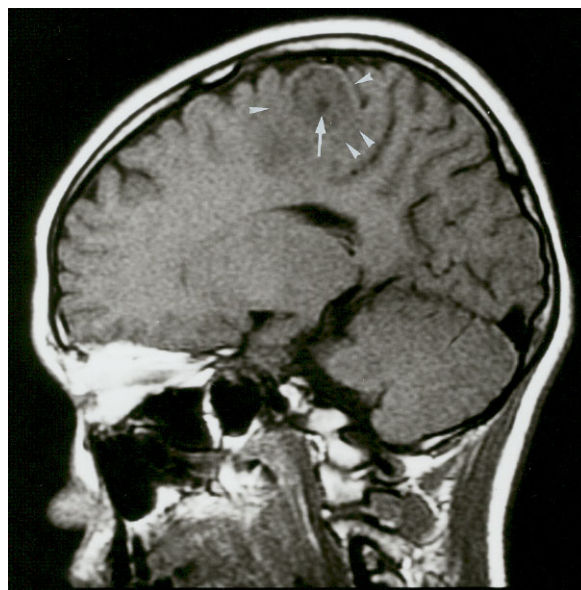


Fig. 1B. Sagittal T1-W MR image shows the intra-axial location of the tumour, which is circumferentially surrounded by brain parenchyma (arrowheads). The tumour is isointense with a small central cystic component (arrow).



Fig. 1C. Fluid-attenuated inversion recovery MR image obtained in the coronal plane shows prominent surrounding vasogenic oedema.

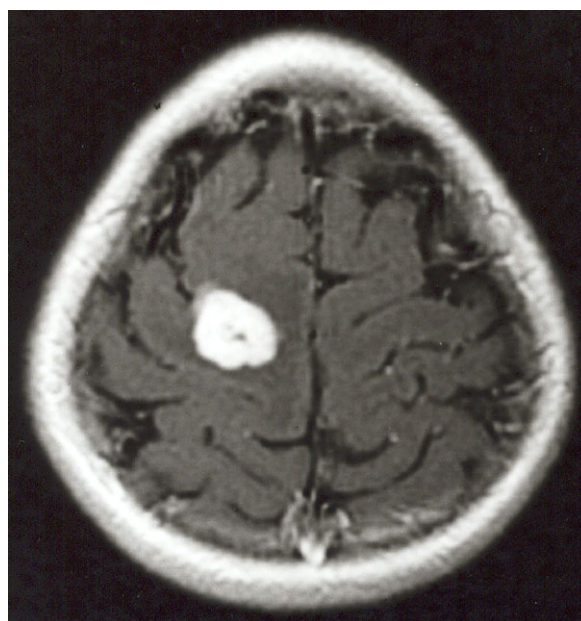


Fig. 1D. Enhanced axial T1-W MR image shows intense and uniform enhancement of the tumour.

literature.²⁻⁵ The majority of patients with intracerebral schwannomas are children or young adults below the age of 30, presenting with headache, seizures and focal neurological deficit.³

Most reports of this rare tumour have been case reports, the largest series being 9 patients reported by Casadei et al.⁴ On CT and MR imaging studies, all lesions were well-circumscribed and intra-axial in location, showing various patterns of contrast enhancement. Cyst formation or cystic

degeneration was found in 78% of the lesions.⁴ In a literature review of CT and MR imaging findings, Zagardo et al³ also found that cysts were documented pathologically or by imaging studies in 19 of 35 cases. Oedema and/or gliosis was found in 17 of 27 patients who had MR imaging or CT studies, and enhancement was found in 20 of 21 patients receiving intravenous contrast.³ Calcification and haemorrhage have also been described. Therefore, with the exception of its intracerebral location, the findings of

intraparenchymal schwannomas are similar to those described for peripheral schwannomas.^{1,6}

In our patient, a well-circumscribed mass with a small central cystic area was found in the frontal lobe, clearly intra-axial in location. T1-weighted sequences showed a tumour that was isointense with grey matter. It demonstrated intense contrast enhancement and was surrounded by cerebral oedema. These imaging features, although consistent with findings described in intracranial schwannomas, are not specific and overlap with many of the more common intracranial neoplasms.

Amongst children, intracranial tumours presenting with epilepsy tend to be low-grade neoplasms, especially gliomas.^{7,8} The most common glial tumour in a child is pilocytic astrocytoma. Frequently located in the cerebellar hemisphere, pilocytic astrocytomas may also be found in the optic nerve, diencephalon or cerebral hemispheres. On MR imaging, the tumour is typically well-circumscribed with macroscopic cyst formation and usually demonstrates intense contrast enhancement of the solid component. However, surrounding oedema is rare in pilocytic astrocytoma.⁹

The superficial intra-axial location of the tumour in our patient also led us to consider the neuronal-glial group of neoplasms in our differential diagnosis. This group of superficial or cortical neoplasms include ganglioglioma, pleomorphic xanthoastrocytoma and dysembryoplastic neuroepitheliomas.^{6,9-11} Dysembryoplastic neuroepitheliomas do not usually enhance; the frequent appearance of a cyst with solid nodule in pleomorphic xanthoastrocytoma and calcification in gangliogliomas are features that may aid differentiation in our patient. Finally, the combination of contrast enhancement and surrounding oedema did not allow us to exclude malignant tumours such as metastases, lymphoma or cerebral neuroblastoma.

Diagnosis of schwannoma is based on histological examination. Haematoxylin and eosin-stained sections reveal a neoplasm comprising compact, intersecting spindle cells with elongated, irregular and hyperchromatic nuclei, with indistinct cell borders: the characteristic Antoni Type A pattern seen in schwannomas. The tumour cells typically exhibit a strongly positive staining pattern for S100 protein on immunohistochemistry.

The origin of schwannomas in an intra-axial location is uncertain. Schwann cells are found in the peripheral and cranial nerves and they are generally absent from the central nervous system. Several theories have been proposed^{1-3,12} and the young age at onset suggests the possibility of a developmental basis for at least some

schwannomas. The majority of tumours follow a benign course and resection is curative.

Conclusion

Intracerebral schwannomas are a rare cause of epilepsy in children. The neuroimaging features are not specific, but cyst formation, peri-tumoural oedema and contrast enhancement are characteristic. The tumour may resemble pilocytic astrocytoma or malignant tumours. However, given its benign nature and favourable response to resection, it is important to differentiate it from other neoplasms.

Acknowledgements

We thank Dr Jennifer Teo from the Department of Pathology, Singapore General Hospital, for her assistance in the preparation of this manuscript.

REFERENCES

1. Russell D S, Rubenstein L J. Pathology of tumours of the nervous system. 5th ed. London: Edward Arnold, 1989:100-9.
2. Huang P P, Zagzag D, Benjamin V. Intracranial schwannoma presenting as a subfrontal tumor: case report. *Neurosurgery* 1997; 40:194-7.
3. Zagardo M T, Castellani R J, Rees J H, Rothman M I, Zoarski G H. Radiologic and pathologic findings of intracerebral schwannoma. *AJNR Am J Neuroradiol* 1998; 19:1290-3.
4. Casadei G P, Komori T, Scheithauer B W, Miller G M, Parisi J E, Kelly P J. Intracranial parenchymal schwannoma. A clinicopathological and neuroimaging study of nine cases. *J Neurosurg* 1993; 79:217-22.
5. Di Biasi C, Trasimeni G, Iannilli M, Poletti E, Gualdi G F. Intracerebral schwannoma: CT and MR findings. *AJNR Am J Neuroradiol* 1994; 15:1956-8.
6. Atlas S W. Magnetic resonance imaging of the brain and spine. 2nd ed. Philadelphia: Lippincott-Raven, 1996:361-2, 365-6, 463-6, 911, 1365-6.
7. Bruton C J. The neuropathology of temporal lobe epilepsy. Institute of Psychiatry Maudsley Monographs No. 31. New York: Oxford University Press, 1988.
8. Cascino G D. Structural brain imaging. Epilepsy: a comprehensive textbook. In: Engel J Jr, Pedley T A, editors. Philadelphia: Lippincott-Raven, 1997:940-1.
9. Lee Y Y, Van Tassel P, Bruner J M, Moser R P, Share J C. Juvenile pilocytic astrocytomas: CT and MR characteristics. *AJR Am J Roentgenol* 1989; 152:1263-70.
10. Lipper M H, Eberhard D A, Phillips C D, Vezina L G, Cail W S. Pleomorphic xanthoastrocytoma, a distinctive astroglial tumor: neuroradiologic and pathologic features. *AJNR Am J Neuroradiol* 1993; 14:1397-404.
11. Castillo M, Davis P C, Takei Y, Hoffman J C Jr. Intracranial ganglioglioma: MR, CT, and clinical findings in 18 patients. *AJNR Am J Neuroradiol* 1990; 11:109-14.
12. Feigin I, Ogata J. Schwann cells and peripheral myelin within human central nervous tissues: the mesenchymal character of Schwann cells. *J Neuropathol Exp Neurol* 1971; 30:603-12.