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Subfrontal Schwannoma Masquerading as Meningioma

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ABSTRACT

Intracranial schwannomas not associated with cranial nerves are rare and seldom encountered in the subfrontal region. We report a case of subfrontal schwannoma in a 21-year-old man who presented with seizures. Radiological features resembled an olfactory groove meningioma. The histological diagnosis of schwannoma was confirmed by immunohistochemical staining with S-100 and electron microscopy. We advocate the use of immunohistochemistry and electron microscopy as adjuncts to conventional light microscopy in differentiating schwannomas from meningiomas. Surgery remains the main therapeutic modality and complete excision is associated with cure.

Keywords: Immunohistochemistry, schwannoma, subfrontal, S-100 protein

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INTRODUCTION

Schwannomas represent 8% of all primary intracranial tumours, mostly arising from the VIII cranial nerve[1]. Intracranial schwannomas without association with cranial nerves account for less than 1% of surgically treated schwannomas of the central and peripheral nervous system[2]. A search through the literature revealed 59 cases of intracranial schwannomas not arising from cranial nerves, of which only 17 cases (29%) were found in the subfrontal region[2-18]. We report a case of subfrontal schwannoma masquerading as a meningioma. The pathogenesis of subfrontal schwannoma and the diagnostic tools for differentiating meningiomas from schwannomas are discussed.

CASE REPORT

A 21-year-old Chinese man presented after a generalised, tonic-clonic seizure. Residual right leg weakness was noted after the seizure. Neurological examination confirmed right lower limb weakness with grade 3/5 power. He had no other neurological deficits. Gadolinium enhanced magnetic resonance scans revealed a large, extra-axial tumour with solid and cystic components in the anterior cranial fossa which enhanced heterogeneously (Fig. 1a). A clinical diagnosis of an olfactory groove meningioma was made.

At surgery, a firm yellowish extra-axial tumour measuring 5 cm in diameter was found upon retraction of the frontal lobes. The tumour was attached to a 1 cm area at the skull base, lateral to the right cribriform plate. Dura and bone were absent at the site of

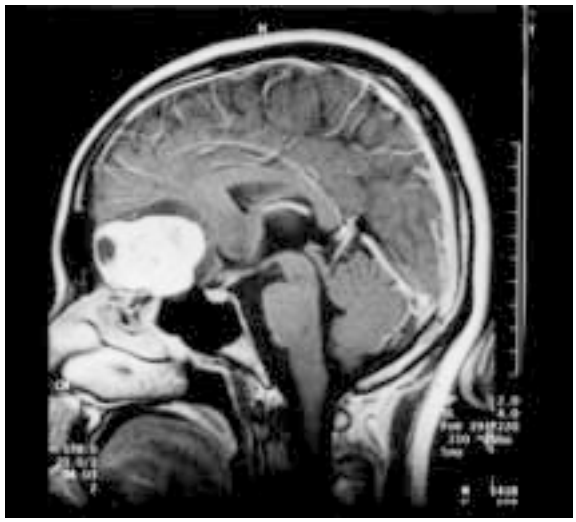

Fig. 1a Sagittal T1-weighted MRI with gadolinium enhancement revealed a large heterogeneous extra-axial tumour with cystic component located subfrontally in the anterior cranial fossa.

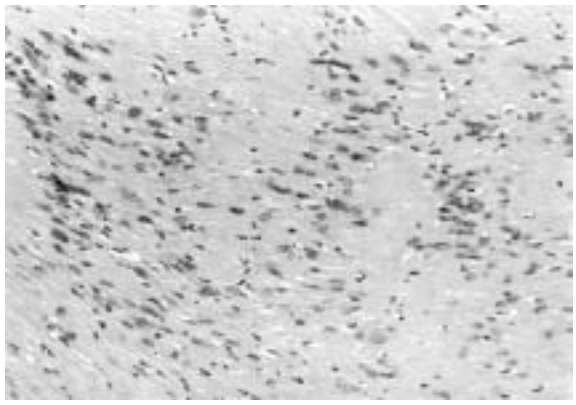

Fig. 1b Photomicrograph showing the typical Antoni A pattern of schwannoma with palisading nuclei. (H & E, X 200)

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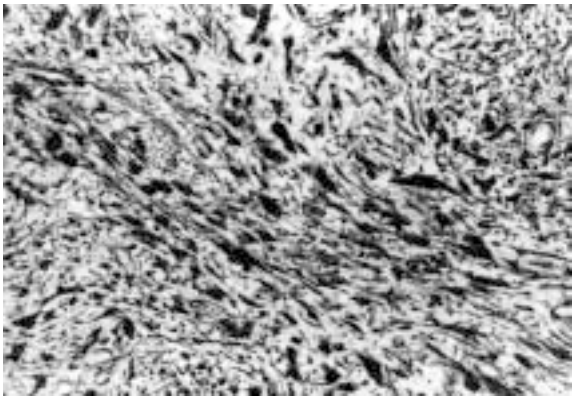

Fig. 1c Photomicrograph with tumour cells showing the nuclei staining strongly positive for S-100 protein. (X 200)

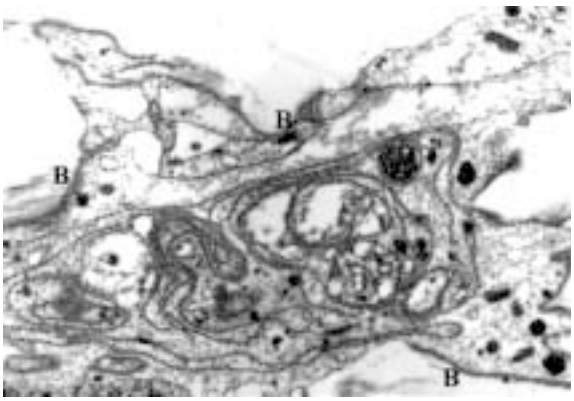

Fig. 1d Electron micrograph showing intermingled cell processes and the presence of a basement membrane (labelled as B) surrounding the cell membrane. (X 14 600)

tumour attachment. Gross total tumour excision was achieved. The skull base defect was repaired with autologous muscle, fascia and histoacry1 glue.

Microscopically, the tumour cells formed patterns of compact fascicular Antoni A areas with palisading nuclei (Fig. 1b) and reticular Antoni B areas. Immunohistochemistry demonstrated that the tumour cells stained strongly positive with S-100 protein (Fig. 1c) and negative with epithelial membrane antigen (EMA). Immunostaining of the tumour tissue with Leu-7 revealed focal positivity. Electron microscopy showed the presence of a basement membrane (Fig. 1d). These findings were compatible with a schwannoma.

The power of the right lower limb improved to normal after the operation. At follow-up one year later the patient remained well with no evidence of recurrence.

DISCUSSION

Intracranial schwannomas not associated with cranial nerves are rare and can be divided into intra-axial (intraparenchymal) and extra-axial tumours. The postulated origins of these tumours include developmental (congenital) and nondevelopmental derivations.

More than 80% of intra-axial schwannomas occurred in patients younger than the age of 30 and were therefore thought to be developmental in nature[19,20]. According to proposed developmental theories, the tumour cells may arise from displaced neural crest cells[1,21], pial mesodermal cells[1] or multipotent mesenchymal cells[22].

Proponents of nondevelopmental theories advocated that schwannomas arose from Schwann cells present on adjacent structures, such as perivascular nerve plexuses or branches of the trigeminal nerve[1,2,12,15]. We believe the tumour in our patient was nondevelopmental in aetiology and most likely originated from the anterior ethmoidal branch of the trigeminal nerve. This would explain the absence of dura under the tumour base as well as its location in the subfrontal region. All the subfrontal tumours reported in the literature were extra-axial, which were consistent with a nondevelopmental origin[2].

Subfrontal schwannomas are slow growing, allowing them to reach a considerable size before diagnosis. The commonest presentation is headache[5,7,18] followed by seizures[5,12,15], visual disturbances[2,4,7,15], nasal stuffiness and epistaxis[4,5,7,18].

Subfrontal schwannomas have similar radiological features as meningiomas such as extra axial location, contrast enhancement, perifocal oedema, and bone erosion[12,15]. The presence of cysts on radiological imaging of our patient is more suggestive of schwannomas[19,23], as cysts are found in only 2-4% of meningiomas[24].

The microscopic appearance of schwannoma with Antoni A and B areas has been well defined[1]. Antoni A areas are identified by a compact texture composed of interwoven bundles of long spindle cells. These cells have oval central nuclei containing variable amounts of chromatin and inconspicuous nucleoli. The cytoplasm is pale with a stringy appearance. The nuclei may be aligned in rows separated by clear hyaline bands forming a palisading pattern. The Antoni B areas in schwannomas are distinguished by the loose texture and polymorphism of tumour cells embedded in a fine honeycomb eosinophilic matrix. This appearance may be mimicked by meningioma[25,26]. In addition, schwannomas occasionally have a conspicuously whorled pattern resembling meningiomas[1,27]. The diagnosis of schwannoma presenting in this unusual location in our patient would have been difficult to make based upon routine haematoxylin and eosin stains alone. Immunohistochemistry is a simple, accurate, and cost-effective technique used in the diagnosis of brain tumours. S-100 protein is a diagnostic immunohistochemical marker for schwannoma[1,28,29].

Epithelial membrane antigen (EMA) immunostaining is a characteristic feature of meningiomas[1,28,29]. The strong staining reaction with S-100, and lack of immunoreactivity with EMA confirmed the diagnosis of schwannoma in this patient. However, up to 15% of meningiomas are S-100 positive, and some schwannomas may be EMA positive[29]. Leu-7 is an immunohistochemical stain that has been detected in 80% of schwannomas while it is notably absent in meningiomas[30]. Louw et al therefore recommended that tumours staining for S-100 but not with Leu-7 should be studied under electron microscopy[25].

The ultrastructural features of schwannoma on electron microscopy include typical interdigitating cytoplasmic processes covered by a basement membrane. These characteristics are absent in meningiomas. As electron microscopy is a relatively expensive technique accessible only in well-established laboratories, its application should be reserved for those rare cases in which the immunohistochemical results are equivocal.

In summary, schwannomas in unusual sites can be easily mistaken for meningiomas on the basis of clinical presentation and radiological appearances. They may mimic each other on routine light microscopy. The judicious application of immunohistochemical techniques with a battery of antibodies then offers a greater diagnostic specificity. In problematic scenarios where the immunohistochemical findings are indeterminate, we recommend the use of electron microscopy for definitive histopathological diagnosis.

REFERENCES

1. Russell DS, Rubinstein LJ, editors. Pathology of tumours of the nervous system. 5th ed. Baltimore: Wilkins & Wilkins, 1989; 537-60.
2. Huang PP, Zagzag D, Benjamin V. Intracranial schwannoma presenting as a subfrontal tumor: Case report. Neurosurgery 1997; 40:194-7.
3. Bando K, Obayashi M, Tsuneharu F. A case of subfrontal schwannoma. No Shinkei Geka 1992; 20:1189-94.
4. Bavetta S, Mcfall MR, Afshar F, Hutchinson I. Schwannoma of the anterior cranial fossa and paranasal sinuses. Br J Neurosurg 1993; 8:697-700.
5. Boyd JH, Dalsao TA Jr, Bee CS, Smith KR Jr, Martin DS. Subfrontal schwannoma with intracranial mucocele: a case report. Am J Otolaryngol 1997; 18:72-5.
6. Enion DS, Jenkins A, Miles JB, Diengdoh JV. Intracranial extension of a naso-ethmoid schwannoma. J Larynol Otol 1991; 105:578-81.
7. Gatscher S, Love S, Coakham HB. Giant nasal schwannoma with intracranial extension. J Neurosurg 1998; 89:161.
8. Harada T, Kawauchi M, Watanabe M, Kyoshima K, Kobayashi S. Subfrontal schwannoma - case report. Neurol Med Chir (Tokyo) 1992; 32:957-60.
9. Harano H, Hori S, Kamata K, Shinohara T, Toyama K. A case report of subfrontal schwannoma. No Shinkei Geka 1974; 2:643-7.
10. Mauro A, Sciolla R, Sicuro L, Ponzio R. Solitary neurinoma of the anterior cranial fossa. J Neurosurg Sci 1983; 27:45-9.
11. Nagao S, Aoki T, Kondo S, Gi H, Matsunaga M, Fujita Y. Subfrontal schwannoma: a case report. No Shinkei Geka 1991; 19:47-51.
12. Sabel LHW, Teepen JLJM. The enigmatic origin of olfactory schwannoma. Clin Neurol Neurosurg 1995; 97:187-91.
13. Sato S, Toya S, Nakamura T, Ohtani M, Imanishi T, Kodaki K, Nakamura Y. Subfrontal schwannoma: report of a case report. No Shinkei Geka 1985; 13:883-7.
14. Spiller WG, Handrickson WF. A report of two cases of multiple sarcomatosis of the central nervous system and of one case of intramedullary primary sarcoma of the spinal cord. Am J Med Sci 1903; 126:10-32.
15. Ulrich J, Levy A, Pfister C. Schwannoma of the olfactory groove. Acta Neurochir (Wien) 1978; 40:315-21.
16. Vassilouthis J, Richardson AE. Subfrontal schwannoma: Report of a case. Acta Neurochir (Wien) 1980; 53:259-66.
17. Viale ES, Pau A, Turtas S. Olfactory groove neurinomas. J Neurosurg Sci 1973; 17:193-6.
18. Zovickian J, Barba D, Alksne JF. Intranasal schwannoma with extension into the intracranial compartment. Neurosurgery 1986; 19:813-5.
19. 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; 79:217-22.
20. Frim DM, Ogilvy CS, Vonsttal JP, Chapman PH. Is intracerebral schwannoma a developmental tumor of children and young adults? Pediatr Neurosurg 1992; 18:190-4.
21. Ramamurthi B, Anguli VC, Iyer CGS. A case of intramedullary neurinoma. J Neurol Neurosurg Psychiat 1958; 21:92-4.
22. Feigin I, Ogata J. Schwann cells and peripheral myelin within human central nervous tissues. The mesenchymal nature of Schwann cells. J Neuropathol Exp Neurol 1971; 30:603-12.
23. Ezura M, Ikeda H, Ogawa A, Yoshimoto T. Intracerebral schwannoma: Case report. Neurosurgery 1992; 30:97-100.
24. Kulah A, Ilcayto R, Fiskeci C. Cystic meningiomas. Acta Neurochir (Wien) 1991; 111:108- 13.
25. Louw D, Sutherland G, Halliday W, Kaufmann J. Meningiomas mimicking cerebral schwannoma. J Neurosurg 1990; 73:715-9.
26. Sobel RA, Michaud J. Microcystic meningioma of the falx cerebri with numerous palisaded structures: an unusual histological pattern mimicking schwannoma. Acta Neuropathol 1985; 68:256-8.
27. Kepes JJ. Presidential address: the histopathology of meningiomas. A reflection of origins and expected behaviour? J Neuropathol Exp Neurol 1986; 45:95-107.
28. Schnitt SJ, Vogel H. Diagnostic value of immunoperoxidase staining for epithelial membrane antigen. Am J Surg Pathol 1986; 10:640-9.
29. Winek RR, Scheithauer BW, Wick MR. Meningioma, meningeal hemangiopericytoma (angioblastic meningioma), peripheral hemangiopericytoma, and acoustic schwannoma. A comparative immunohistochemical study. Am J Surg Pathol 1989; 13:251-61.
30. Perentes E, Rubinstein LJ. Immunohistochemical recognition of human nerve sheath tumours by anti-Leu 7 monoclonal antibody. Acta Neuropathol 1985; 68:319-24.