Schwannoma with tentorial attachment in the cerebellopontine angle mimicking a meningioma

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Abstract

Intracranial schwannoma not associated with the cranial nerves is rare. It is also an intriguing neoplasm since the Schwann cell is not native to the central nervous system. To date only four cases of intracranial schwannoma arising from the tentorium have been reported. We present a 49-year-old woman who harboured a schwannoma with a tentorial attachment in the right cerebellopontine angle and describe the relevant clinical, radiological and pathological findings. In addition, we briefly review the main hypotheses for the origin of this neoplasm and highlight its resemblance to meningioma and inclusion as a differential diagnosis.

Keywords: Schwannoma; Tentorium; Dura; Cerebellopontine angle; Meningioma

1. Case report

A 49-year-old woman with no stigmata of neurofibromatosis presented with a 2-week history of right hemicranial headache and neck pain, vomiting and unsteadiness of gait with drift to the right. Examination revealed a right facial paresis and paraesthesia, which was accompanied by slightly depressed corneal sensation and reflex.

1.1. Radiology

A CT brain scan showed a contrast-enhancing meningeal-based $4 \times 2.3 \times 2.6$-cm bi-lobed soft tissue mass, with central low density in the right cerebellopontine angle with local mass effect on the brainstem but no hydrocephalus. The internal auditory meatus was not widened, and there were no peri-tumoral oedema or adjacent bone hyperostosis.

An MRI brain scan demonstrated a lobulated heterogeneous enhancing cystic lesion with meningeal enhancement along the inferior aspect of the tentorium and anteriorly along the sphenoid wall in the right cerebellopontine region indenting the brainstem (Fig. 1). The T2-weighted posterior fossa images showed no involvement of the right internal auditory meatus, nor the 7th or 8th cranial nerves (Fig. 2).

1.2. Operation

A right retrosigmoid suboccipital craniotomy using frameless stereotaxy was performed. At operation the lesion was adherent to the tentorial and petrosal dura, but dissected away easily from the parenchyma and complete macroscopic resection was achieved. The cranial nerve complexes V, VII, VIII and IX–XI were all identified and preserved, and the mass was not found to be associated with these cranial nerves. Frozen section demonstrated a spindle cell tumour with some nuclear enlargement and hyperchromasia and a provisional diagnosis of meningioma was made. The patient made an uneventful recovery and was discharged on post-operative day 8. At 6-weeks follow-up, she was progressing well and except for residual paraesthesia involving the right mandible, she was symptom-free and neurologically intact.

1.3. Pathology

Light microscopy revealed a spindle cell tumour with a loosely textured pattern and foam cells and mast cells. There was also nuclear hyperchromasia and pleomorphorism seen, but no meningothelial whorls (Fig. 3). Immunohistochemistry for epithelial membrane antigen, cytokeratins AE1/AE3, CK8/18, and glial fibrillary acidic protein were negative, and positive for S100 (Fig. 4). Tissue was also processed for electron microscopy which demonstrated neoplastic spindle cells with long thin intertwining processes with basal lamina surrounded by collagen fibres consistent with Schwannian differentiation.

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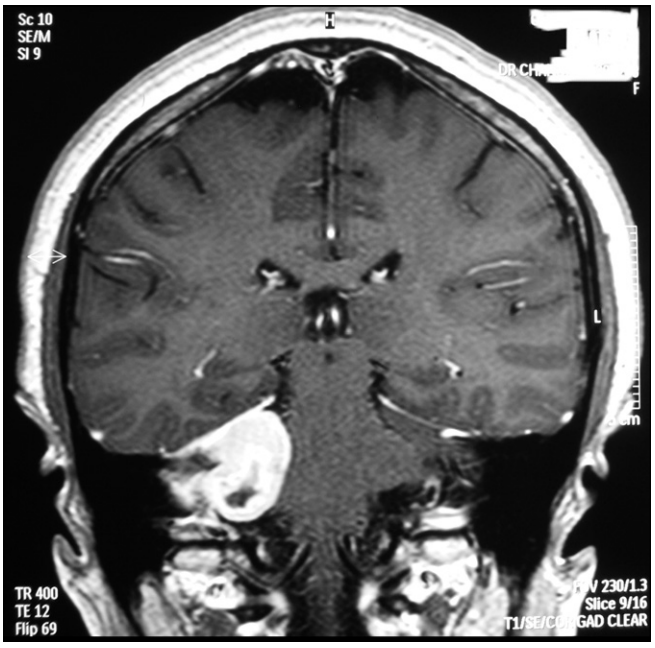

Fig. 1. Coronal T1-weighted MRI of the brain with gadolinum demonstrating a lobulated heterogeneous enhancing cystic mass with meningeal enhancement along the inferior aspect of the tentorium.

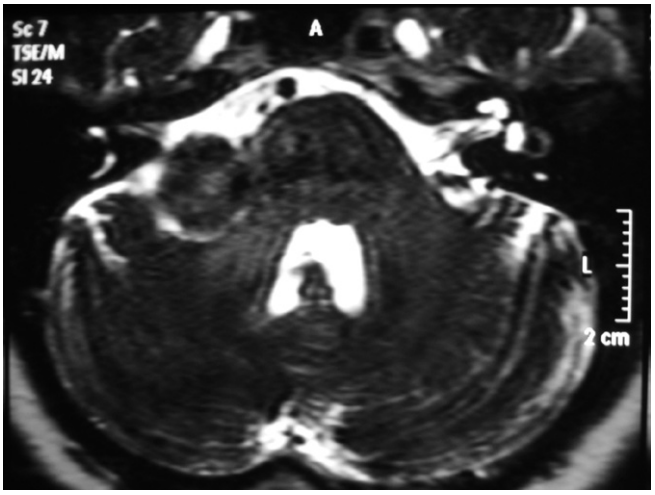

Fig. 2. Axial T2-weighted MRI of the posterior fossa showing no involvement of the right internal auditory meatus.

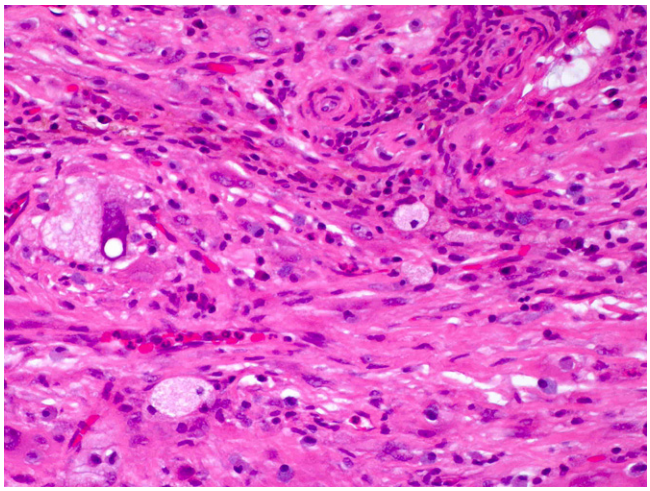

Fig. 3. Light microscopy with haematoxylin and eosin stain showing a spindle cell tumour in a loosely textured pattern (×20 magnification).

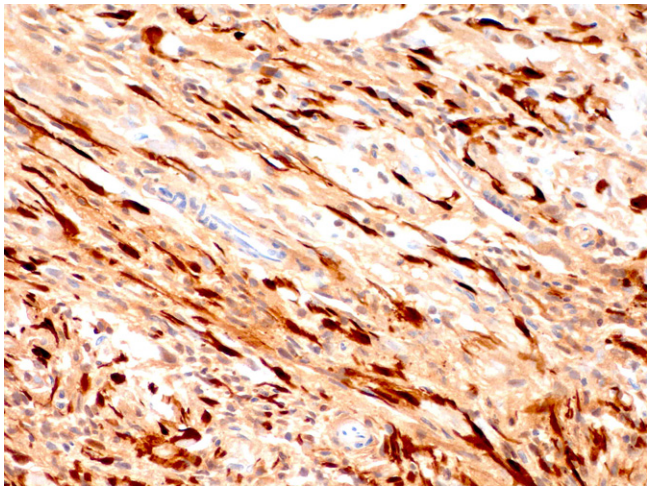

Fig. 4. Immunohistochemistry stain with positive staining for S100 (×20 magnification).

2. Discussion

Intracranial schwannomas account for about 6.8–8.0% of all primary brain tumours, of which 80–90% are found in relation to the vestibulocochlear nerve. Other cranial nerves such as the trigeminal, facial, and hypoglossal nerves may also be involved.[1] Indeed, intracranial schwannoma unrelated to cranial nerves is unusual as the central nervous system is devoid of Schwann cells. In 1966 Gibson et al. published what appears to be the first recorded case of an intracerebral Schwann cell tumour in the temporal lobe of a 6 year-old boy.[2] Since that time, fewer than 50 similar cases have been reported. A thorough search of English language literature has revealed most cases arose as intra-axial lesions in the supratentorial compartment, and only 13 were attached to the dura (Table 1),[3–15] of which four were in the posterior fossa attached to the tentorium (Table 2).[10–13] In addition, we report an extracanalicular schwannoma with tentorial attachment occurring in the right cerebellopontine angle of a 49-year-old woman.

The incidence of vestibular schwannoma increases with age and peaks between the fourth and sixth decades; there is also a female preponderance of 1.5–2.1.[1] These are in contrast to the recent reviews of intracerebral schwannoma, which have attributed different clinical characteristics. Reports have suggested a relatively younger age at presentation and a male preponderance. Casadei et al. reported a mean age of 23.5 years with 79.3% of patients less than 29 years and a slight male preponderance of 1.6.[16] Most lesions are benign and involve the supratentorial