

Intracranial cystic (ancient) schwannoma of the temporal lobe: A Rare Occurrence

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Citation

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Abstract

We report a rare case of intracranial cystic schwannoma in the temporal fossa. A 42-year-old female admitted to our hospital with a 18-year history of deviation of right angle of mouth. CT scan revealed a large cystic mass in the temporal lobe. Magnetic resonance imaging (MRI) revealed a slightly enhanced, partly solid, partly cystic mass in the right temporal lobe. Preoperative diagnosis was glioma and then total resection was performed. Microscopic examination of the tumor showed the characteristic biphasic pattern with cellular Antoni A and hypocellular Antoni B areas. Immunohistochemically, the tumor was positive for S-100. From these findings, the tumor was diagnosed as an ancient schwannoma. Intracranial schwannomas, which are not associated with cranial nerves, are rare. Origin of these tumors and differential diagnosis are discussed.

INTRODUCTION

Schwannomas are benign tumors accounting for approximately 8% of all intracranial lesions. Intracranial schwannomas not arising from cranial nerves are extremely rare¹. The presence of a cyst together with the tumor appears to be characteristic of such intraparenchymal schwannomas of the brain. Here we report one such rare occurrence in a 42-year old female.

CASE REPORT

A 42-year old female presented with deviation in the right angle of mouth for the last 18 years. She also complained of headache in the frontoparietal region since 4 years before, which increased in severity over the last year. On examination there was right facial palsy of LMN type. CT Scan showed a large, 5.8x5.5 cm, well-defined cystic mass with a thin peripheral rim of calcification, situated in the right temperoparietal lobe (fig1). Possibility of degenerating hydatid cyst was suggested. MRI showed a heterogenously enhancing mass in the temporal lobe with curvilinear calcification. Possibility of ganglioglioma was then suggested (fig2). Temporal craniotomy was performed and a large 8x8 cm sized cyst was found, adherent to the surrounding brain parenchyma. Total excision of the cystic mass was done.

On gross examination there were multiple, grey white to pearly white soft tissue pieces measuring altogether 5 cc.

Some of the soft tissue pieces showed areas of calcification. On microscopic examination, there were hypercellular and hypocellular areas. The hypercellular areas comprised of sweeping fascicles of slender elongated spindle cells with wavy serpentine nuclei and formation of Verocay bodies in some areas (Antoni A) (fig3). These cells showed some pleomorphism and nuclear atypia, but mitotic figures were not evident. Other areas were hypocellular with myxoid background and occasional foamy macrophages (Antoni B) (fig4). Occasional vessels displayed a periluminal hyaline ring. Reticulin stain showed a rich pericellular reticulin staining in Antoni B areas. Immunohistochemically the tumor cells were positive for S-100 protein. The tumor was then diagnosed as ancient schwannoma of the temporal lobe.

Figure 1

Figure 1

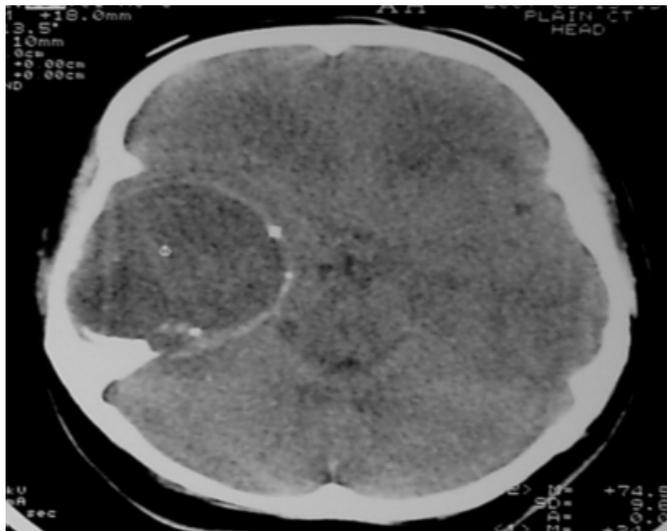


Figure 2

Figure 2

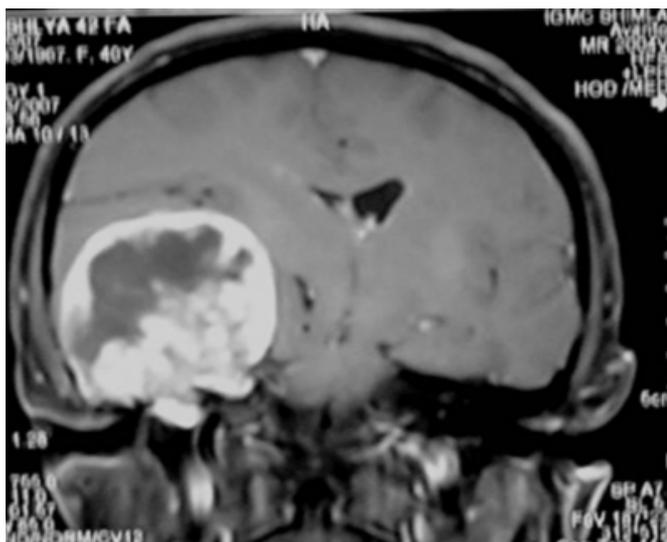


Figure 3

Figure 3

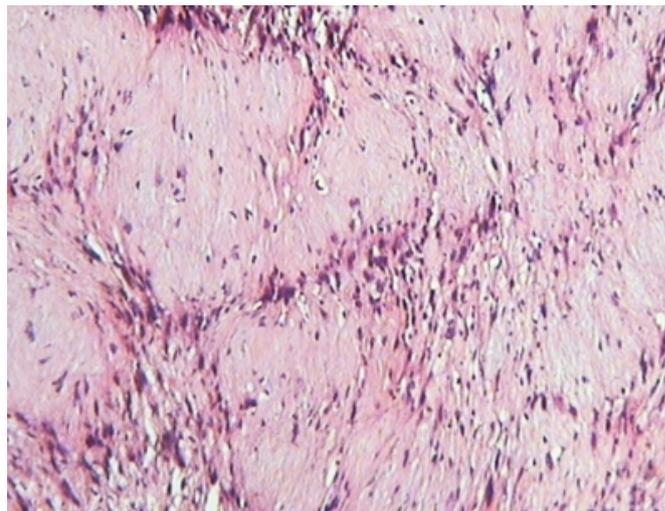
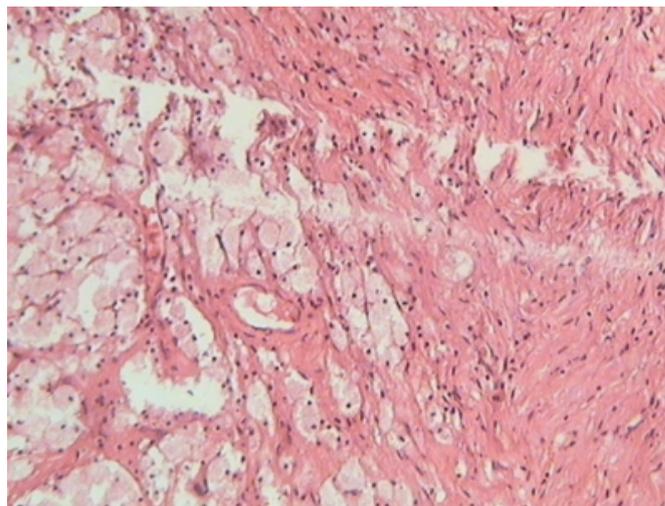


Figure 4

Figure 4



DISCUSSION

Intracranial schwannomas not arising from the facial, trigeminal, or vestibular nerves are extremely rare, in non-neurofibromatosis patients. Such schwannomas account for less than 1% of surgically treated schwannomas of the central and peripheral nervous systems². Furthermore, intracerebral schwannomas are even rarer lesions; only 37 well-documented cases have been reported in world literature³. Gibson et al⁴ were the first ones to report intracerebral intraparenchymal schwannoma in 1966.

Schwannomas commonly arise from the nerve sheaths of peripheral and cranial nerves. Thus, since the central nervous system is devoid of the Schwann cells present in nerves, it is unclear the pathogenesis of intracerebral schwannomas.

Several theories have been proposed for their intracerebral occurrence. These theories can broadly be considered in two groups, the developmental and non-developmental. According to the developmental theory, aberrant Schwann cells in the brain parenchyma may occur due to the transformation of the mesenchymal pial cells⁵, or from displaced neural crest cells that form foci of Schwann cells ('schwannosis')⁶. The relatively young age at presentation also suggests a developmental etiology. Non-developmental theories base their assumption on the fact that Schwann cells are present within the perivascular nerve plexuses and large arteries in the subarachnoid spaces⁷, although the existence of these structures deep in the brain parenchyma is doubted⁸. However, Schwann cells are present in the adrenergic nerve fibers innervating the cerebral arterioles⁹. These nerve plexi are common in tela choroidea, which may explain their predilection for periventricular location¹⁰.

Intraparenchymal schwannomas are detected either in the first two decades, when they present with an indolent, slow-growing course, or in the elderly, when their symptoms evolve rapidly³. Males are affected more often and present with headache and seizures. Most of the tumors are located in the supratentorial compartment. The presence of a cyst together with the tumor appears to be characteristic of intraparenchymal schwannoma of the brain¹¹.

Histopathologically, the detection of Antoni A and Antoni B structures, Verocay bodies, infiltration by foamy macrophages and vascular hyalinization usually suffice for recognition of schwannomas. However these tumors need to be differentiated from fibroblastic meningiomas (with prominent nuclear palisading), tanacytic ependymomas, subependymomas, and pilocytic astrocytomas. Schwannoma can be distinguished from astrocytoma and ependymoma by its abundant parenchymal reticulin. Fibroblastic meningiomas are more difficult to distinguish from schwannoma when they lack the characteristic features of meningioma like meningeal whorls and psammoma bodies. Furthermore, Antoni A and Antoni B growth patterns resemble fibroblastic meningiomas. Verocay bodies are more distinctive than Antoni patterns, but they are not seen in all schwannomas. Schwannomas contain Leu-7 and S-100 protein. In the rare meningiomas that express S100 protein, electron microscopy can be of great help, since schwannomas have continuous basement membranes along the exterior surface of their cells.

Ancient schwannomas are usually large tumors of long

duration that display pronounced degenerative changes in the form of cyst formation, calcification, hemorrhage and hyalinization. The tumor is usually infiltrated by large number of foamy macrophages. Schwann cell nuclei can show nuclear atypia, which can be regarded as purely degenerative change. The presence of hypercellularity and atypia may lead to the misdiagnosis of these lesions as sarcomas, which includes melanomas, leiomyosarcomas, hemangiopericytomas, etc. The absence of mitotic activity is the key feature to differentiate benign ancient schwannomas from malignant schwannomas. The presence of a capsule, evidence of prior hemorrhage, thick-walled vascular structures and areas representing degenerative changes also suggest a diagnosis of a benign lesion.

Surgery remains the main therapeutic modality and due to the benign nature of the tumor, complete excision is associated with cure and the long-term outcome after excision is generally good. The patient was discharged from our hospital with no neurological deficits.

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