

Frontal intraparenchymal schwannoma: a case report

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Abstract

The Intraparenchymal schwannomas are very rare tumors with only 52 reported cases. We describe a 38 years old man with eight months history of poorly controlled seizures despite of medical therapy. On brain CT scan a 1.5 x 1.5 cm hypodense enhancing lesion was seen in right posterior-inferior frontal lobe. On brain MRI the lesion was hypointense in T1 and hyperintense in T2 with homogenous enhancement after contrast injection. He underwent localized craniotomy and total excision of the tumor. The tumor was completely covered by brain cortex and well demarcated with fleshy consistency and grayish color. After histopathologic examination and Immunohistochemical staining, diagnosis of the schwannoma was established. During 6 months period of follow up no seizure was reported by the patient.

Keywords

schwannoma, intraparenchymal, frontal lobe

Introduction

The Schwannomas accounts for 5% to 8% of all primary brain tumors [10]. The vestibular portion of eighth cranial nerve is the most affected location, accounting for 80% to 90% of all intracranial schwannomas. Intraparenchymal schwannomas are very rare tumors with only 52 reported cases in the literature review [1,3,6-9]. In 1966 the first case reported by Gibson in a 6 year old boy with seizures [18]. Although vestibular schwannoma are less common in children than in adults, it appears that intraparenchymal schwannomas mostly occur in younger population. When intraparenchymal schwannomas involve a hemispheric location, the differential diagnosis must include pilocytic astrocytomas, pleomorphic xanthoastrocytomas,

gangliogliomas, hemangioblastomas and meningiomas. The first two are more common in pediatric population, and the later three in adult population [10]. Many lesions exhibit a cystic component with areas of variable enhancement, however, imaging characteristics for individual tumors must be considered [10]. Therefore intraparenchymal schwannoma should be considered in the differential diagnosis, despite of its rarity, of any solid or solid/ cystic enhancing cerebral lesions.

Case report

A 38 years old man with eight months of poorly controlled seizure referred to our clinic. On clinical examination no neurological deficit was found. Brain CT scan revealed a hypodense lesion in the posterior-inferior area of right frontal lobe with homogenous enhancement af-

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ter contrast injection (Fig.1).On brain MRI the lesion was hypointense in T1 and hyperintense in T2. Homogenous enhancement after contrast injection was seen in the lesion (Fig.2).He underwent elective localized craniotomy. After opening the dura no tumor was seen but with a small corticectomy the tumor was found. The tumor was completely intraaxial and has fleshy consistency with well demarcated margin.Total excision of the tumor was done. Histopathological study revealed compact fascicle of fusiform cells with elongated nuclei and indistinct cytoplasmic borders arranged in palisading pattern (Antoni A) admixed with haphazardly oriented loosely arranged cells in a myxoid matrix (Antoni B) (Fig.3). In immunohistochemical staining the reticulin stain and S-100 were positive, whereas the glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA) were negative. Slides were reviewed by two pathologists in two centers and

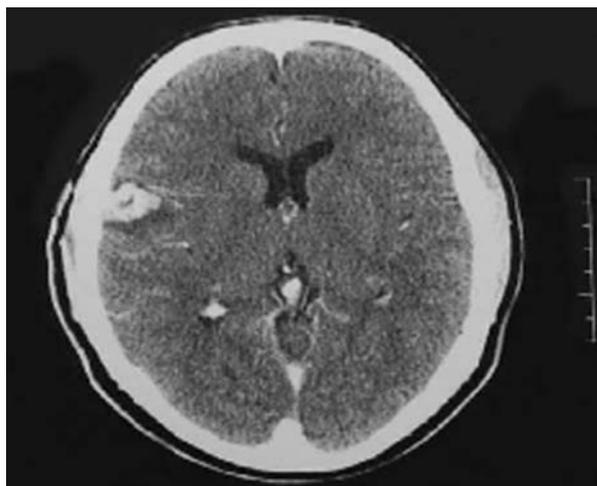


Fig. 1. Axial brain CT Scan after contrast injection.

the diagnosis of intraparenchymal schwannoma was established (Fig. 4).

Discussion

Although the intraparenchymal schwannomas are rare tumors and 52 related cases were

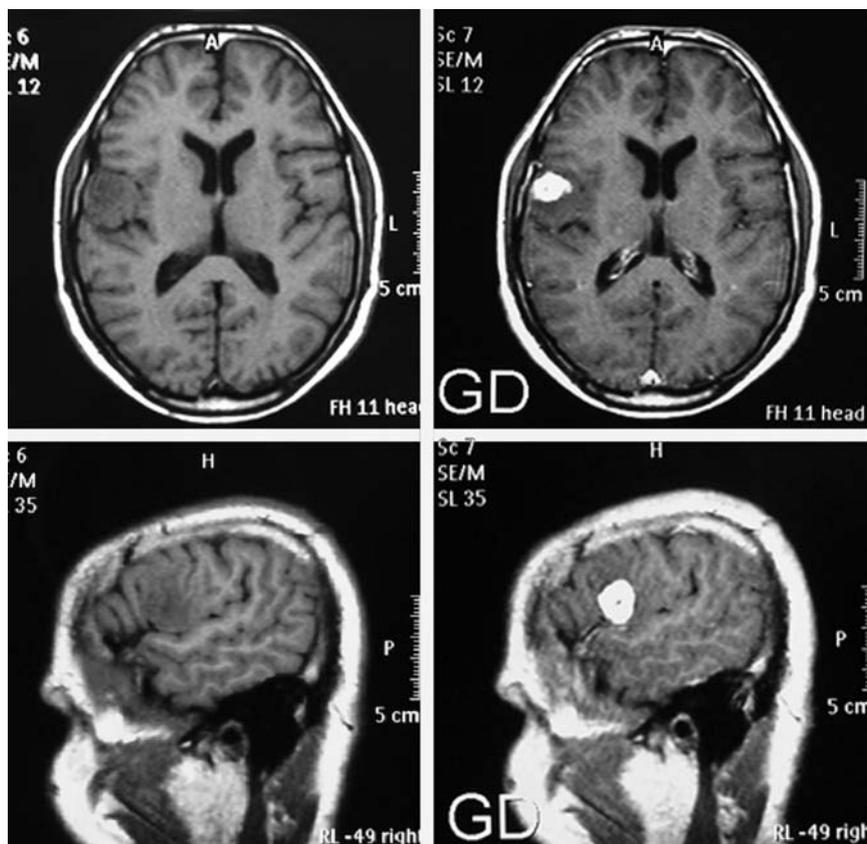


Fig. 2. Axial and sagittal view of brain MRI before and after contrast injection.

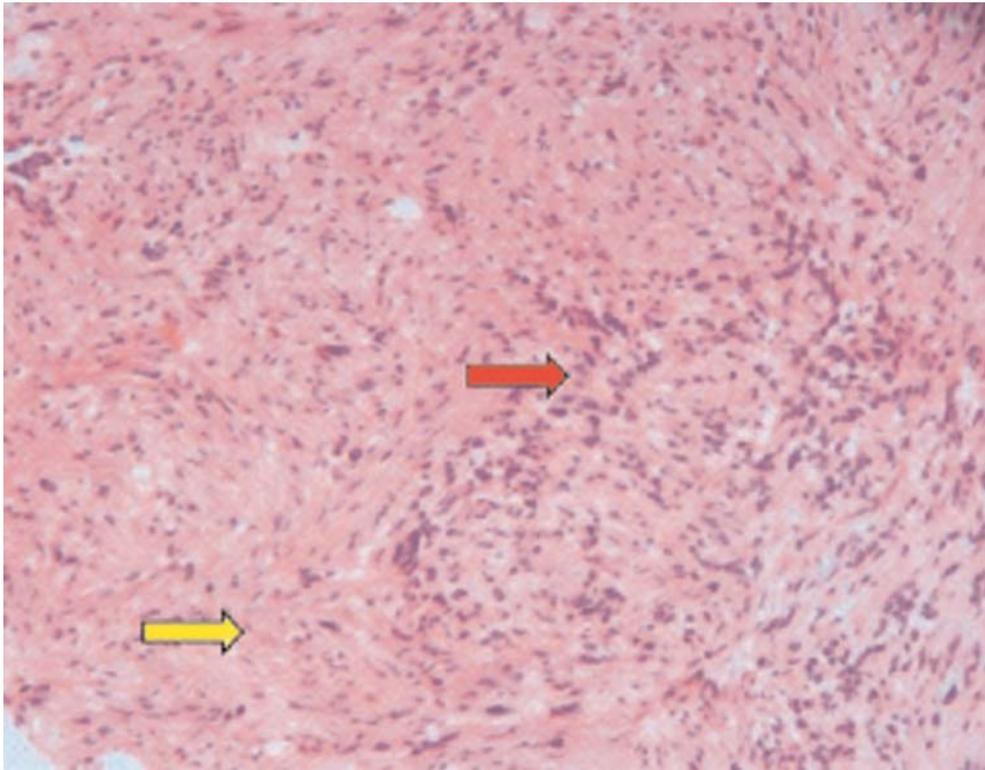


Fig. 3. H&E staining x20, Antoni A (red arrow) and Antoni B (yellow arrow).

reported in the literature review since 1966 [10,17]. Supratentorial lesions are more common than infratentorial lesions as in our case, and association with the ventricular system was

also reported with various imaging characteristics [3]. Of 20 patients 10 underwent MRI with contrast, as in our case, and enhancement of the tumor was homogenous. Peritumoral edema,

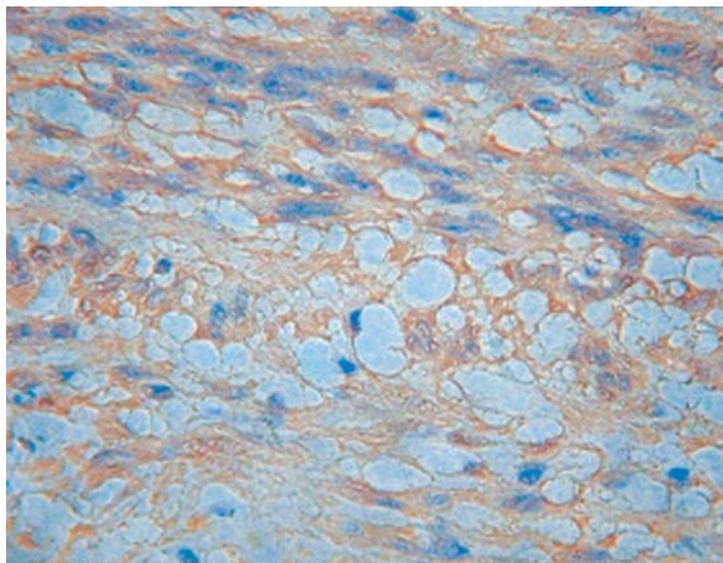


Fig. 4. IHC stainingx20. Tumor cells are exhibited nuclei strongly positive for S-100 protein.

represented by T2-weighted hyperintensity, occurred in 22 out of 27 patients. The influence of vascular endothelial growth factor was proposed as a mechanism underlying the formation of edema around meningiomas, but its presence was not confirmed in intracerebral schwannomas [3]. In a review of 35 cases, Zargardo et al [3] found no gender predilection, unlike schwannomas of eighth cranial nerve, which has a female predominance. Furthermore, intracerebral schwannomas occur in a younger population: 70% of patients present before the age of 30 years and almost half (13 out of 32) are younger than 15 years old [12]. Vestibular schwannomas are most common in the 5 and 6 decade, unless they are associated with neurofibromatosis type 2, in which case the mean age at presentation is 20 years [13].

The origin of intracerebral schwannomas remains unclear. Although similarities between mesodermal pial cells and neuroectodermal Schwann cells were reported [18,14], the potential for pial cells to undergo conversion to Schwann cells is theoretical. This theory was based on morphological studies performed by Russell and Rubinstein. The most likely site of origin is Schwann cells of the perivascular nerve plexus in the subarachnoid space [4]. This hypothesis would explain the frequent presentation of these lesions along the convexity or in deep ventricular locations, although lesions might also be expected to occur adjacent to the circle of Willis. Cerebral vascular innervation studies suggest a differential response to stimulation among vessels of different diameters [11], with hypothesis that smaller vessels in the cortical and periventricular regions have a propensity to developing schwannomas than the larger basal vessels lack. Another theory presented by Feigin and Ogata [5] and by Hirano et al [19] relates to the observation of myelin in cerebral infarction and multiple sclerosis plaques. Feigin and Ogata [5] also proposed that mesenchymal multipotential cells differentiate into Schwann cells. This proposal con-

tributes to the developmental origin theory and helps account for their occurrence in children. Finally, the concept of schwannosis was originally espoused by Gibson et al. [15] in the first description of an intracerebral schwannoma. Schwannosis describes the existence of aberrantly located Schwann cells with neoplastic potential that must be considered as the source of this tumor. Erongul et al [9] proposed trigeminal fibers innervating the dura as a source of Schwann cells for tumors located adjacent to the dura. Huang et al. [7] suggested differentiating subfrontal schwannomas from other intraparenchymal schwannomas. In their review of 16 cases of strictly subfrontal lesions, none of the tumors were intraaxial. These patients also presented at an older age. In contrast 26 out of 31 other intracerebral, nonvestibular schwannomas were intraaxial and presented at a mean age of 21. They hypothesized a congenital or developmental source for the intraparenchymal lesion, thus accounting for the age difference.

In all but one report, such as our case intracerebral schwannomas were described as "well-demarcated. Bruni et al. [16] reported an adult with a left frontal parasagittal lesion that exhibited parenchymal invasion. Because these tumors are so rare, the implications of this finding are unclear. Although the histologic appearance of schwannomas with biphasic areas of Antoni A and B cells is characteristic, the immunohistochemistry tend to be also useful in differentiating intracerebral schwannomas. On histochemical analysis, these tumors exhibit strong staining for reticulin and diffuse nuclear and cytoplasmic positivity for S-100 protein. These lesions are negative for synaptophysin, neurofilaments, and cytokeratins features that help distinguishing them from tumors of neural origin. Although discrete areas of IHC staining may be positive for GFAP, but they are grossly negative, and unlike meningiomas, negative for EMA. The presence of basement membrane on electron microscopy helps distinguish schwann-

nomas from meningiomas. Although Verocay bodies are less common in intracerebral lesions, they were reported in some studies.

Conclusion

Supratentorial intracerebral schwannomas are rare tumors [1,3,6-9,17]. The majority of these occurred in children and young adults [2]. The most common signs and symptoms include headache, seizure, and focal neurological deficits. Superficial or deep periventricular locations are believed to be characteristic. The differential diagnoses are tuberculoma, meningioma, pilocytic astrocytoma, pleomorphic xanthoastrocytoma and ganglioglioma [6]. Total surgical resection is the mainstay of treatment for clinical improvement. Although the margins appeared grossly clean at the end of tumor resection, a recurrence is still possible. Routine follow-up imaging at 6 months intervals will be obtained for the first 2 years. Subsequently, imaging will be obtained with decreasing frequency for at least 10 years.

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