

Atypical Intracerebral Schwannoma Mimicking Glial Tumor: Case Report

Glial Tümörü Taklit Eden Atipik İntrakranial Şivannom: Olgu Sunumu

ABSTRACT

A rare case of a solitary schwannoma arising within parenchyma of the brain is reported. A 37-year old right-handed man presented with a 3-month history of progressive headaches, lethargy and vomiting. The initial diagnosis being considered was primary intracranial tumor, including high-grade astrocytoma, metastasis or lymphoma. Histopathological examination revealed schwannoma. Cysts, calcification and mild to moderate peritumoral edema are common in intracerebral schwannomas. However, our case is atypical and has no cystic component, calcification or vascularization. The pathogenesis and neuroradiological findings of intraparenchymal schwannomas are discussed and we review the related literature.

KEYWORDS: Cerebral tumor, Focal convulsive seizure, Schwannoma

ÖZ

Parankim içerisinde köken alıp, çok nadir görülen bir soliter şivannom olgusu sunuldu. 37 yaşında erkek hasta 3 aylık progresif artan baş ağrısı, sersemlik ve kusma nedeniyle başvurdu. İlk tanı yüksek dereceli astrositom, metastaz veya lenfoma şeklindeydi. Histopatolojik inceleme sonucunda şivannom tanısı kondu. İntraserebral şivannomlar kistik, kalsifiye ve hafiften orta dereceye kadar etrafı ödemli bir şekilde görülür. Halbuki bizim olgumuz atipik formda olup kist, kalsifikasyon ve vasküler komponent içermiyordu. İntraparankimal şivannomanın patogenezi ve nöroradyolojik bulguları tartışıldı ve ilgili literatür gözden geçirildi.

ANAHTAR SÖZCÜKLER: Serebral tumor, Fokal konvülsif nöbet, Şivannom

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INTRODUCTION

Schwannomas represent approximately 8% of all intracranial tumors, predominantly arising from the vestibular portion of the VIIIth cranial nerve and less commonly, with descending order of frequency, from the Vth, IXth, Xth, and VIIth cranial nerves (10). While, schwannomas arising from other cranial nerves are rare and usually associated with von Reclinghausen's disease, intracranial schwannomas not arising from cranial nerves are extremely rare (5). The pathogenesis of intracerebral schwannoma and the diagnostic tools for differentiating other mass lesions from schwannomas are discussed.

CASE REPORT

A 37-year old right-handed man presented with a 3-month history of progressive headaches, lethargy and vomiting. On June 20, 2003, he had suffered from a focal convulsive seizure in his upper left arm for which he was referred to our hospital for further evaluation. The results of the physical examination performed at the time of admission were unremarkable. No cutaneous stigmata of neurofibromatosis Type 1 was observed. A fundoscopic examination revealed papilledema. A neurological examination revealed no significant findings. Computerized tomography (CT) scans revealed a large, uniformly high-attenuation lesion in the right frontal lobe, surrounded by severe brain edema. Magnetic resonance images revealed a large, heterogeneously enhancing intra-axial lesion with solid component inside of the deep right frontal lobe (Figure 1). There was severe peritumoral edema in flair scans (Figure 2). The initial diagnosis being considered was primary intracranial tumor, including high grade astrocytoma, metastasis or lymphoma. The patient underwent right fronto-temporal craniotomy using neuronavigation (VectorVision; BrainLab, Heimstetten, Germany), with gross total resection of the tumor. Histopathological examination revealed schwannoma. On microscopic examination, the tumor was composed of compact interlacing fascicles of spindle cells, characterized by elongated, hyperchromatic, and irregular nuclei and occasional intranuclear inclusions, with scattered foci of nuclear palisading (Figure 3). The postoperative course was uneventful, and the patient was discharged home on fourth post-operative day without neurological deficits. An MR images study performed one year later after surgery showed no evidence of residual tumor (Figure 4). The patient remains asymptomatic 3 years later on follow-up.

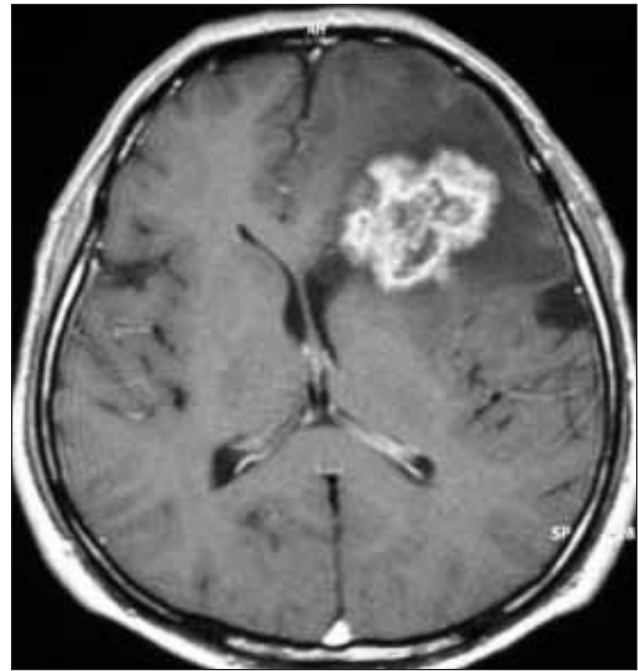


Figure 1. Magnetic resonance images revealed a large, heterogeneously enhancing intra-axial lesion with a solid component inside of the deep right frontal lobe.

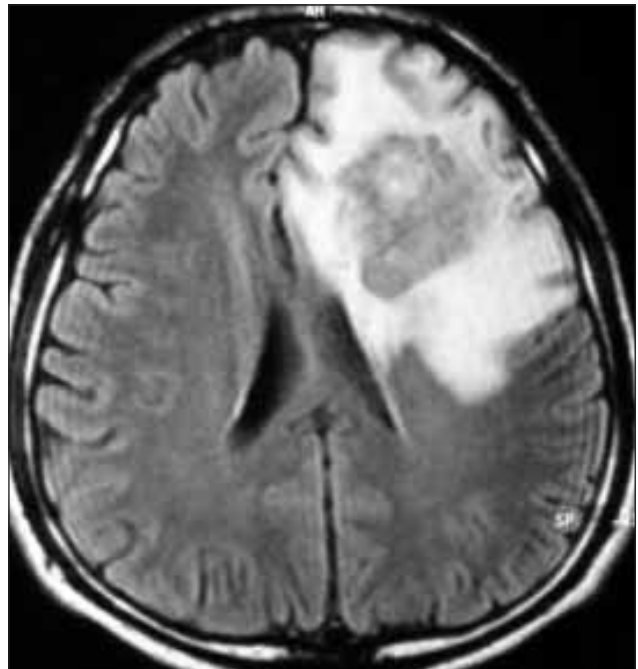


Figure 2. There was severe peritumoral edema in flair scans.

DISCUSSION

It is an extremely rare event for a schwannoma to occur within the parenchyma of the brain (3, 5, 6). These tumors commonly arise from the nerve sheaths of peripheral and cranial nerves (10). As schwann cells are normally not present in the

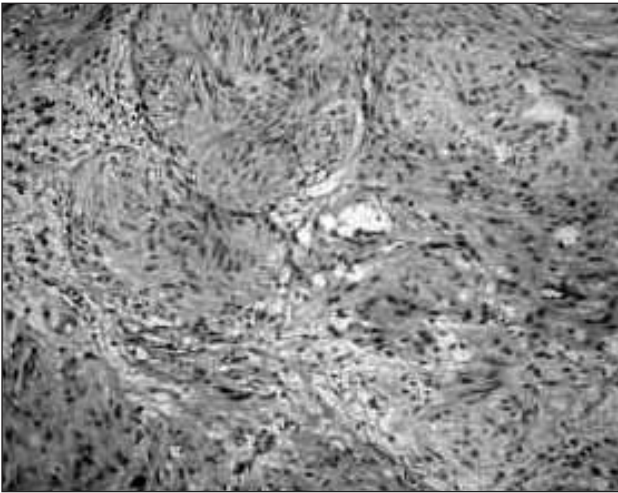


Figure 3. Antoni type A area demonstrating compact interlacing fascicles of spindle cells, characterized by elongated, hyperchromatic, and irregular nuclei, with scattered foci of nuclear palisading (hematoxylin-eosin).

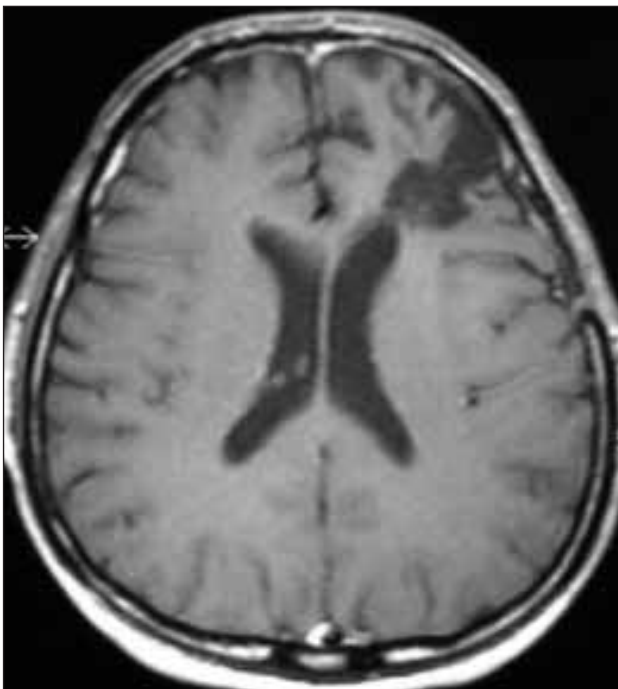


Figure 4. MR images showing no evidence of residual tumor.

cerebral parenchyma, it is difficult to explain the origin of intracranial parenchymal schwannomas. Schwann cells have been detected around arteries in the subarachnoid space and within the periphery of the brain along the perivascular nerve plexus (9). Frim et al. (4) suggest that they either originate from the multipotent mesenchymal cells or displaced neural crest cells, which form the foci of schwann cells termed as schwannosis within the CNS parenchyma.

The histogenesis of these tumors is still not settled, and several theories have been proposed for their intracerebral occurrence. These theories can broadly be considered under two groups as 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 cells, or from displaced neural crest cells that form the foci of schwann cells (10). Non-developmental theories base their assumption on the fact that schwann cells are present within the perivascular nerve plexi and large arteries in the subarachnoid spaces (9), although the existence of these structures deep in the brain parenchyma is doubted (2). However, schwann cells are clearly present in the adrenergic nerve fibres innervating the cerebral arterioles. These nerve plexi are common in tela choroidea, which may explain their predilection for periventricular location (1).

The differential diagnosis of an intracerebral schwannoma includes several other neoplasms that may occur in children and young adults. These include pilocytic astrocytoma, pleomorphic xanthoastrocytoma, ganglioglioma, meningioma and glioblastoma multiforme. The specimens should be examined using immunohistochemistry to make a correct diagnosis (8). Microscopic differential diagnosis includes fibroblastic meningioma and solitary fibrous tumor (Antoni type A predominant schwannoma), and pilocytic/pilomyxoid astrocytoma (Antoni type B predominant schwannoma). It is sometimes very difficult to differentiate between these neoplasms based on the histological examination alone, as there is an overlap in the histological features. Thus, either immunohistochemical or electron microscopic examination is crucial for the differential diagnosis (11).

The neuroradiological characteristics of intracerebral schwannoma are thought to be high frequency of calcifications, cystic formations, peritumoral edema, and angiographical vascularization (7). On CT, the tumors present as isodense or hypodense intra-parenchymal lesions in the frontal or temporal lobes, with a predilection for the periventricular white matter. They show uniform contrast enhancement and often have a cystic component. MRI characteristics are variable, with T1 isointensity and hypointensity and mixed T2 isointensity and hyperintensity (7). Cysts, calcifications and mild to moderate peritumoral

edema are common with intracerebral schwannomas. However, our case is atypical and has no cystic component, calcification or vascularization.

CONCLUSION

Intracerebral schwannoma is a rare, benign neoplasm. It is usually located superficially or adjacent to a ventricle. These tumors cannot be preoperatively differentiated from other parenchymal tumors. Surgical excision is curative and the long-term prognosis is good. There are no reported recurrences after gross total resection.

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