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Case Report

Supratentorial extraparenchymal schwannoma mimicking parasagittal meningioma: A rare case report

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Abstract

Background: Intracranial schwannomas not related to cranial nerves are very rare. Young age, no known history of neurofibromatosis, and seizure as initial symptom have been reported to be associated with intraparenchymal schwannoma.

Case Description: We report a case of supratentorial parasagittal schwannoma in the right frontal region presenting with seizure episode in a 70-year-old man. Computed tomography and magnetic resonance imaging showed a right frontal solid, enhancing extra-axial lesion based on anterior and middle third junction of superior sagittal sinus. The preoperative diagnosis was right parasagittal meningioma, however, the microscopic examination of the mass showed the characteristic pattern of cellular Antony A pattern. Immunohistocemically, the tumor stained positive for S-100 protein but negatively for epithelial membrane antigen and glial fibrillary acidic protein. These findings are consistent with schwannoma. Cysts, calcification, and peritumoral edema are common in intracerebral schwannoma, which were not seen in our case.

Conclusion: On the basis of clinical presentation and radiological appearances, schwannoma in unusual sites can easily be mistaken for meningiomas; immunochemistry plays an important role in differentiating them. Till date, to the best of our knowledge, this is the second reported case of schwannoma mimicking meningioma in parasagittal location.

Key Words: Immunohistochemistry, parasagittal meningioma, supratentorial schwannoma

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INTRODUCTION

Schwannomas represent approximately 8% of all intracranial tumors predominantly arising from the vestibular portion of the 8th cranial nerve and, less commonly with descending order of frequency, from 5th, 9th, 10th, and 7th cranial nerves.[6] While schwannomas arising from other cranial nerves are rare and usually associated with von Recklinghausen's

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disease, intracranial schwannomas not arising from cranial nerves are extremely rare.[7] We present a case of supratentorial frontal schwannoma mimicking a parasagittal meningioma in a 70-year-old man. This is only second case of supratentorial schwannoma mimicking meningioma in parasagittal location reported in the literature.

CASE REPORT

A 70-year-old man presented after a generalized tonic–clonic seizure. Residual left leg weakness was noted after the seizure. Neurological examination confirmed left lower limb weakness with grade 3/5 power. He had no other neurological deficits. Magnetic resonance imaging (MRI) revealed a large well-defined dural-based extra-axial lesion that was hypointense on T1 weighted and iso-hyperintense on T2-weighted images along the right frontal convexity causing mass effect and contralateral midline shift. Post-gadolinium images showed homogenous enhancement of the lesion based on the lateral superior sagittal sinus wall at the junction of anterior and middle third and on adjacent falx [Figures 1 and 2]. However, no thrombosis of superior sagittal sinus (SSS) was evident. A clinical diagnosis of right frontal parasagittal meningioma was made.

During surgery, a well-defined mildly vascular, firm, grayish, extra-axial tumor measuring 8 cm × 5 cm was found. In contrast to meningioma, note was made of more soft and suckable tumor, more dirty grayish color, and plenty of thin fibrous septae radiating towards the flimsy capsule dividing the tumor into infinite number of small compartments which could be easily emptied with suction. It was attached to the right inferolateral wall of the sinus and was growing between the two leaves of superior-lateral angle of sinus as a very thin sheet. There was no attachment of tumor capsule to the convexity dura. Gross total resection of the tumor was achieved (Simpson grade 2) which was confirmed by immediate postoperative computed tomography (CT) scan [Figure 3]. Postoperative course was uneventful, and the patient was discharged from the hospital on postoperative day 4.

Microscopically, on hematoxylin and eosin staining, spindle cells arranged in fascicles forming verrocay bodies at places were seen [Figure 4]. Immunochemistry demonstrated that the tumor cells stained strongly positive with S-100 protein [Figure 5] and negative with epithelial membrane antigen (EMA) and glial fibrillary acidic protein (GFAP). These findings were compatible with schwannoma.

The power of the left lower limb improved to normal after the operation. At follow-up 1 year later the patient remained well with no evidence of recurrence.

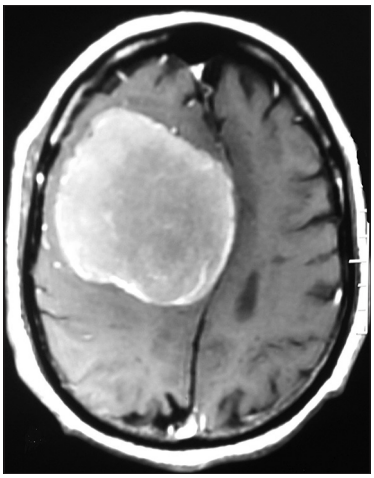

Figure 1: MRI Brain with contrast (axial view) - Homogenous enhancement of the lesion based on the lateral superior sagittal sinus wall at junction of anterior and middle third, and on adjacent falx

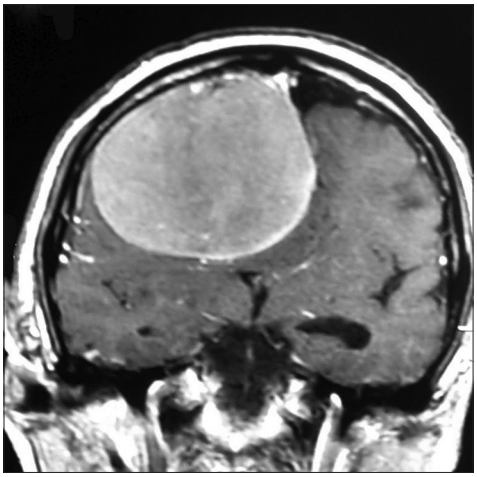

Figure 2: MRI Brain with contrast (coronal view) - Homogenous enhancement of the lesion based on the lateral superior sagittal sinus wall at junction of anterior and middle third, and on adjacent falx

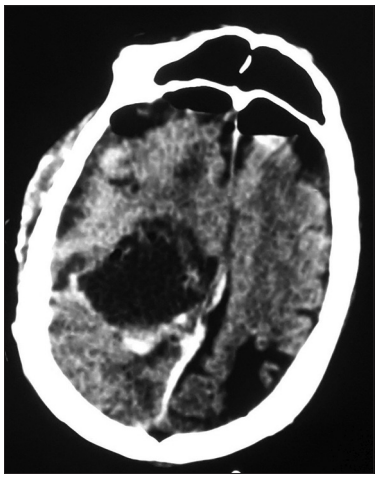

Figure 3: Post –operative CTscan brain- Gross total resection of the tumor was achieved (Simpson grade 2)

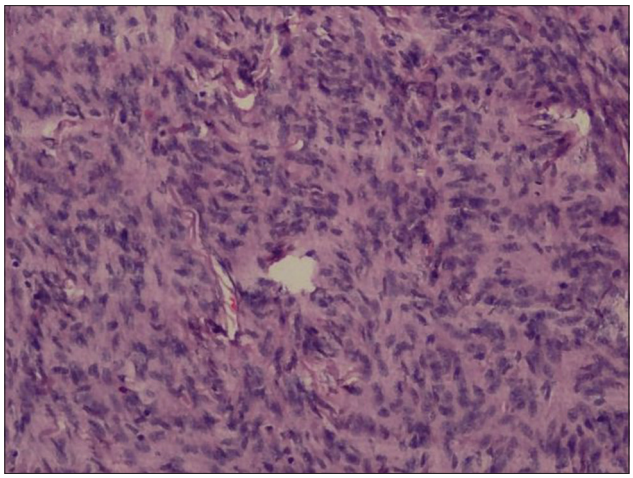

Figure 4: On haematoxylin and eosin staining, spindle cells arranged in fascicles forming verrocay bodies at places were seen

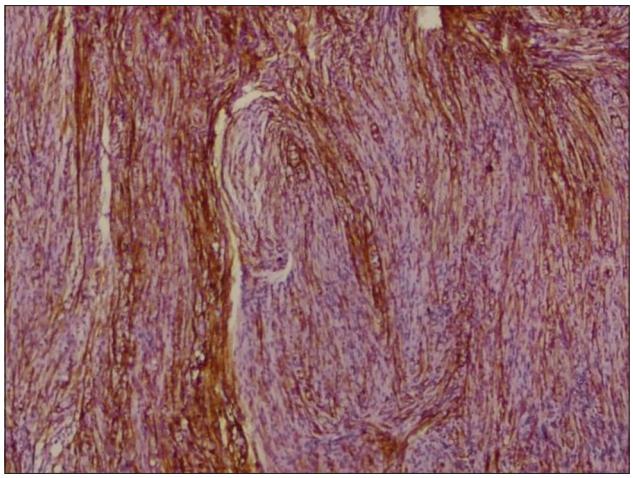

Figure 5: Immunochemistry demonstrated that the tumor cells stained strongly positive with S-100 protein

DISCUSSION

Intracranial schwannomas not related to cranial nerves are very rare and represent less than 1%. As reported elsewhere, approximately 70 cases have been reported so far.[17] Unlike vestibular schwannomas, the intraparenchymal schwannomas are slightly more common in males, though some authors such as Sharma *et al*. have reported significant male to female ratio (M:F = 3:1). Furthermore, majority of cases have been reported in children and young adults, with the average age being less than 21 years. Vestibular schwannomas, however, are more common in the 5th decade unless associated with neurofibromatosis.[2,17] Our was a 70-year-old man.

The location and size of the intracranial schwannoma decide the varied clinical manifestations. Most present with headache, seizures, and focal deficits. Our patient presented with a history of seizures. The typical MRI characteristics of meningioma consist of isointensity to slight hypointensity on T1-weighted sequence and isointensity to slight hyperintensity on T2 sequence. After contrast administration, meningiomas typically demonstrate avid, homogenous enhancement.[15] Some commonly reported radiographic characteristics of intraparenchymal schwannoma are intratumoral calcification (25%), cyst formation (60%), mild to moderate peritumoral edema (50%), and varying degree of gadolinium enhancement of the solid tumor component.[12,17] However some do not consider this true and find it impossible to distinguish them from other tumors such as pilocytic astrocytoma, ganglioglioma, pleomorphic xanthoastrocytoma, and dysembryoplaastic neuroepithelial tumor based on radiography alone. In addition, some authors have reported to have mistaken large intraparenchymal schwannomas for meningioma or high-grade glioma.[1,10] In our case, radiologically it was not possible to differentiate schwannoma from meningioma.

Microscopically, tissue analysis reveals areas of nuclear palisading, characteristic of schwannoma and dense, cellular tumor alternating with loosely textured myxoid tumor is present in equal portions, consistent with Antoni type A and Antoni type B tissue.[3] This appearance may be mimicked by meningioma.[8,9] In addition, schwannomas occasionally have a conspicuously whorled pattern resembling meningioma.[3] The diagnosis of schwannoma presenting in the unusual location in our patient would not have been possible on routine hematoxylin and eosin stains alone. Immunohistochemistry is a simple and accurate technique in this regard. S-100 protein is a diagnostic marker for schwannoma. EMA immunostaining is a characteristic feature of meningioma.[3,11,16] 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 and schwannomas may be EMA positive.[11] Another marker Leu-7 has been detected in 80% of schwannomas whereas it is notably absent in meningiomas.[13] If the results of immunohistochemical staining are equivocal, then electron microscopy is recommended which is a relatively expensive technique accessible only in well-established laboratories.

The histogenesis of intracranial schwannomas not arising from cranial nerves is still unclear as schwann cells are not normally present in the cerebral parenchyma. Many theories have been proposed to explain the possible mechanism underlying the histogenesis and origin of these rare tumors. There are two common theories. One suggests a developmental origin according to which 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.[6] Nondevelopmental theories base their assumption on the fact that schwann cells have been detected around arteries in the intracranial

perivascular nerve plexuses in the subarachnoid space and the brain parenchyma.[4] Our case was typically arising in close association with sinus wall and appeared to grow between the two leaves of superior-lateral angle of sinus as a very thin sheet.

Young age, no known history of neurofibromatosis, and seizure as initial symptom have been reportedly found associated with intraparenchymal schwannoma.[5] Only one earlier case of intracerebral schwannoma mimicking meningioma in parasagittal location has been reported by Ma *et al*. in a 24-year-old female.[14] The tumor in their case revealed a cystic and heterogeneously enhanced giant mass in the right frontal lobe mimicking parasagittal meningioma. Our patient is the second such case to present with a right frontal parasagittal mass which was presumed to be meningioma, however, final histopathological diagnosis was of schwannoma. In our case, we had a 70-year-old patient with a large supratentorial mass with no cystic mass and no peritumoral edema, along with an atypical presentation of earlier reported supratentorial schwannoma. Thus demographic, clinical, and radiological findings are insufficient to arrive at an accurate diagnosis. We emphasize the importance of obtaining a surgical specimen to make proper diagnosis.

The treatment protocol of intracerebral schwannomas is total excision; however, it depends on the location of tumors. Complete relief of clinical symptoms and signs is mostly achieved after total or radical surgical removal.[17] We also achieved gross total resection of tumor in our case.

In summary, on the basis of clinical presentation and radiological appearances, schwannoma in unusual sites can easily be mistaken for meningiomas. Light microscopy may also reveal similar findings. Immunohistochemical techniques with a battery of antibodies offer a greater diagnostic specificity. Our case demonstrates only the second rare case of a supratentorial schwannoma mimicking a meningioma in parasagittal location.

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Conflicts of interest
There are no conflicts of interest.

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