Case Report

A usual tumor at an unusual site: Report of two cases of schwannoma at nonindigenous sites

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Abstract

Schwannoma is a benign, slow-growing encapsulated tumor arising from schwann cells of the nerve sheath and believed to originate from the embryonic neural crest cells. They are common tumors of the head-and-neck region and extremities arising from the nerve sheaths of cranial and spinal nerves. We present two cases of schwannoma occurring at uncommon sites. The first case is a solitary intra-cranial schwannoma presenting in the frontal region which was radiologically diagnosed as meningioma. And, the second case presented as a polypoidal lesion in the right nasal cavity.

Keywords: Intracranial, meningioma, nasal septum, schwannoma

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INTRODUCTION

Schwannoma is a benign, slow-growing encapsulated tumor arising from schwann cells of the nerve sheaths of motor and sensory peripheral nerves and believed to originate from the embryonic neural crest cells.[1,2] These tumors were first described by Verocay in 1910[3] and he named them neurinomas and subsequently these were renamed as neurilemmomas in 1935 by Stout.[4] Schwannomas commonly arise in the head-and-neck region constituting around 25%–40% of the cases. They account for 8% of all the primary brain tumors, and out of this, 80%–90% of the tumors are situated in the cerebellopontine angle.[5] They usually have a single site of origin but around 10% of the tumors are found to have multiple locations of origin. Intracerebral schwannomas are a rare entity and were first reported in 1966 by Gibson *et al.*[5,6]

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CASE REPORTS

Case 1

A 39-year-old male patient presented with difficulty in speech of 1-year duration associated with frontal headache for 2 months. Past history did not reveal any significant illness. Systemic examination was unremarkable. Magnetic resonance imaging (MRI) of the brain [Figure 1a and b] revealed a large well-defined heterogeneously enhancing altered signal intensity extra-axial lesion with a broad base toward the dura in the frontal region, arising from the crista galli and adjacent dura, with significant surrounding mass effect and displacement of the left optic nerve suggesting a diagnosis of meningioma. Bifrontal craniotomy with excision of the lesion was done. The tumor was firmly adherent to the anterior skull base, and no attachment to any cranial nerve was noted.

The specimen consisted of a pale white globular tissue mass measuring 5 cm × 4 cm × 1.5 cm. The outer surface

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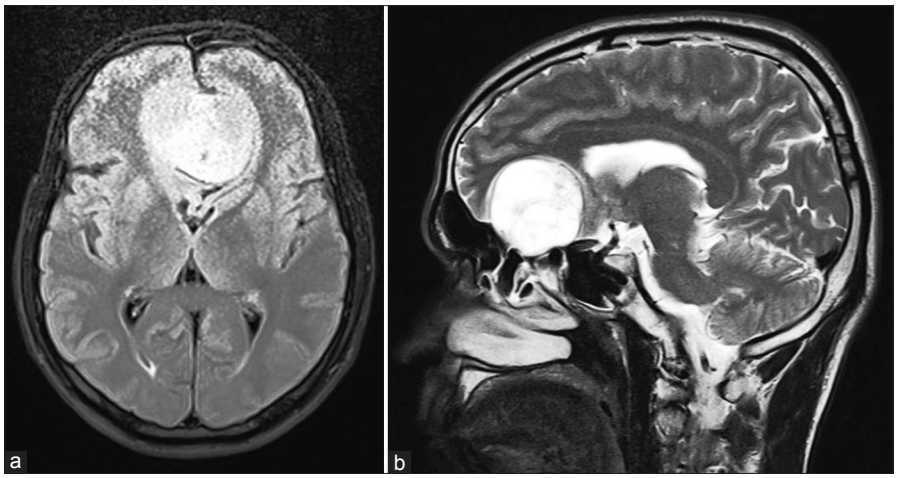

Figure 1: (a and b) Magnetic resonance imaging of brain – well-defined heterogeneously enhancing altered signal intensity extra-axial lesion with a broad base toward the dura in the frontal region, (a) axial view; (b) sagittal view

was partially encapsulated and had cystic and solid consistency. The cut surface revealed a well-circumscribed lesion with solid pale white and myxoid areas seen along with cystic areas which showed papillary excrescences and hemorrhage. Microscopically, sections showed predominantly hypocellular, myxoid Antoni B areas and few hypercellular Antoni A areas. Cells were narrow, elongated, and wavy with tapering ends. Focal nuclear palisading around the fibrillary process (Verocay bodies) was seen in the Antoni A areas [Figure 2a and b]. Also seen were many congested hyalinized blood vessels. It was reported as schwannoma of the anterior cranial fossa.

Case 2

An 81-year-old female patient presented with mass in the right nostril of 1-year duration. A plain and contrast computed tomography (CT) scan revealed a right nasal polyp and chronic right maxillary sinusitis [Figure 3a]. Functional endoscopic sinus surgery was done which showed a septal hemorrhagic mass. The contents from the right maxillary sinus and nasal cavity were separately sent for histological examination. The gross specimen consisted of fragmented tissue bits, one of which was solid and well-circumscribed and measured 0.8 cm × 0.5 cm × 0.3 cm. Microscopic examination revealed elongated spindle cells with Antoni A and Antoni B areas. Palisading of nuclei and Verocay bodies was noted [Figure 3b]. A diagnosis of septal schwannoma was made. The surface epithelium showed ulceration and granulation tissues.

DISCUSSION

Schwannomas can occur at any site where peripheral and cranial nerves are present.[1]

Intracranial schwannomas are most commonly associated with vestibular nerve followed by trigeminal nerve and rarely with other intracranial nerves.[2] Intracranial tumors that are not associated with cranial nerves account for <1% of the surgically treated schwannomas of central and peripheral nervous system, and they are called ectopic schwannomas.[6] They are rare and can be divided into intra- (intraparenchymal) and extra-axial tumors.[5] Studies have shown that there is a close relation in the occurrence of schwannomas in individuals with neurofibromatosis type 2 (NF2), but intracerebral schwannomas are more commonly associated with NF1.[5] These tumors are commonly seen to arise in children or young adults and there is no sex predilection.[5] The most common location of these tumors has been the supratentorial brain parenchyma, with a preference for the periventricular regions.[5] Seizures have been observed as the most common presenting symptom.[6] Other symptoms include raised intracranial pressure and focal neurological deficits.[5] In our case, the patient presented with difficulty in speech and headache.

Based on the clinical and radiological appearances, intracranial schwannomas in unusual sites may be mistaken for other tumors such as meningiomas or gliomas. CT and MRI examinations are not pathognomonic of intracranial, intraparenchymal, and intraventricular schwannomas.[6] On imaging studies, schwannomas may show calcifications, cysts, and peritumoral edema. Noncystic tumors have been identified most commonly in the frontal lobe.[5] In MRI studies, these tumors are relatively isointense to gray matter on T2-weighted images.[5]

Histopathologically, the most common pattern in schwannomas arising in uncommon sites resembles that of a conventional peripheral schwannoma. The differential diagnosis in intracranial schwannomas includes fibroblastic

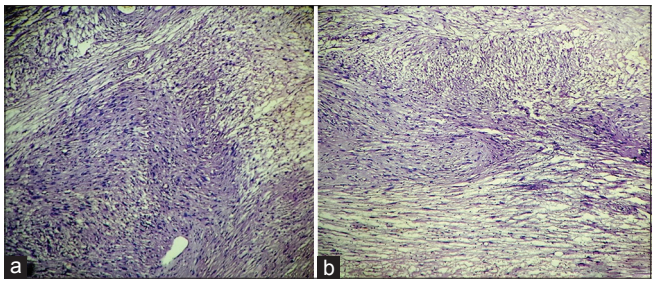

Figure 2: (a and b) Histopathological microphotographs – (H and E, ×100) showing both cellular Antoni A areas comprising interlacing bundles of spindle cells and hypocellular, myxoid Antoni B areas

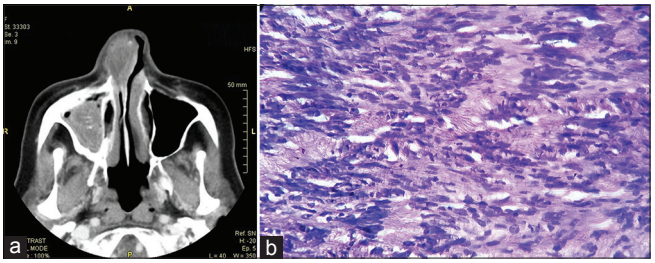

Figure 3: (a) (Computed tomography) scan - a right nasal polyp and chronic right maxillary sinusitis. (b) Microscopic examination - Elongated spindle cells with Antoni A and Antoni B areas. Palisading of nuclei and verocay bodies (H and E, ×400)

meningioma and solitary fibrous tumor in Antoni type A predominant schwannomas and pilocytic/pilomyxoid astrocytoma in Antoni type B predominant schwannoma.[6] The tumor cells may show nuclear pleomorphism and hyperchromasia. Differentiating a cellular schwannoma and malignant peripheral nerve sheath tumor (MPNST) can be challenging. Unlike cellular schwannomas, MPNST shows a more infiltrative border and palisading necrosis.[5] However, the use of immunohistochemistry provides greater diagnostic specificity. The S-100 protein is the diagnostic immunohistochemical marker for schwannoma.[6] Lack of staining for epithelial membrane antigen can help to rule out meningioma.

In our first case, the tumor was located in the anterior cranial fossa displacing the left optic nerve, but an exact site of origin could not be made out intraoperatively as the tumor was extra-axial and not attached to any cranial nerves. As schwann cells are not indigenous to the cerebral parenchyma, various hypotheses have been proposed regarding the origin of intracranial tumors which are not associated with cranial nerves. These can be classified under two groups: developmental and nondevelopmental.[6,7] According to the developmental theories, the mesenchymal pial cells transform into ectodermal schwann cells or the tumor develops from the aberrant neural crest cells within the central nervous system.[8,9] Another study has hypothesized that these cells arise from multipotential mesenchymal cells.[10] Nondevelopmental theories postulate that these tumors arise from schwann cells that are indigenously present in the adjacent structures, for example, perivascular nerve plexus and meningeal branch of the trigeminal or anterior ethmoidal nerves innervating the anterior cranial fossa or fila olfactoria.[5,11-13]

The incidence of schwannomas at sinonasal area is 4% and most commonly seen at ethmoidal sinuses followed by maxillary sinuses, nasal cavity, and sphenoid sinuses. The incidence of nasal septal schwannomas is extremely rare and about twenty cases have been reported in literature.[14] The nasal schwannomas are thought to arise from ophthalmic and maxillary branches of the trigeminal nerve and autonomic ganglia. Due to their occurrence within the confines of the nasal cavity, they tend to present early and are usually smaller in size. Symptoms are nonspecific such as nasal obstruction, rhinorrhea, or recurrent epistaxis.[4] MRI and CT findings in a nasal schwannoma may suggest a differential diagnosis of nasal polyp, lobular capillary hemangioma, MPNST, myxoma, fibromyxoma, and sarcoma. The septal origin of the tumor is usually confirmed intraoperatively and may not be obvious on radiological imaging. In our patient, MRI was suggestive of a nasal polyp and anterior rhinoscopy showed an hemorrhagic mass in the nasal septum.

The treatment of choice is surgical excision and the possibility of recurrence is very remote. Both our patients have been free of tumor at 1 year and 4 months of follow-up, respectively.

CONCLUSION

Schwannomas are rare, benign, slow-growing neoplasms and their occurrence in unusual sites can be misleading both clinically and radiologically. The microscopic appearance along with immunohistochemistry in problematic cases can help in accurate diagnosis. Surgical excision is curative and prognosis is good.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Canbay S, Hasturk AE, Markoc F, Caglar S. Schwannoma of the conus medullaris: A rare case. Chin J Cancer 2011;30:867-70.
2. Kachhara R, Raje P, Pauranik A. Schwannoma originating in lateral recess of the fourth ventricle. Asian J Neurosurg 2012;7:151-3.
3. Saha R, Bhattacharya A, Deb J, Saha G, Panda D. Schwannomas arising at unusual locations – A report of 4 cases. Int J Biomed Adv Res 2014;5:313-8.
4. Mitra B, Debnath S, Paul B, Pal M, Banerjee TJ, Saha TN. Schwannomas of the nasal septum – A rare case report with review of literature. Egypt J Ear Nose Throat Allied Sci 2012;13:121-5.
5. Scott WW, Koral K, Margraf LR, Klesse L, Sacco DJ, Weprin BE, *et al*. Intracerebral schwannomas: A rare disease with varying natural history. J Neurosurg Pediatr 2013;12:6-12.
6. Luo W, Ren X, Chen S, Liu H, Sui D, Lin S, *et al*. Intracranial intraparenchymal and intraventricular schwannomas: Report of 18 cases. Clin Neurol Neurosurg 2013;115:1052-7.
7. Tan TC, Ho LC, Chiu HM, Leung SC. Subfrontal schwannoma masquerading as meningioma. Singapore Med J 2001;42:275-7.
8. Menkü A, Oktem IS, Kontaş O, Akdemir H. Atypical intracerebral schwannoma mimicking glial tumor: Case report. Turk Neurosurg 2009;19:82-5.
9. Russell DS, Rubinstein LJ, editors. Pathology of Tumours of the Nervous System. 5th ed. Baltimore: Wilkins & Wilkins; 1989. p. 537-60.
10. Tsuiki H, Kuratsu J, Ishimaru Y, Nakahara T, Kishida K, Takamura M, *et al*. Intracranial intraparenchymal schwannoma: Report of three cases. Acta Neurochir (Wien) 1997;139:756-60.
11. Feigin I, Ogata J. Schwann cells and peripheral myelin within human central nervous tissues: The mesenchymal character of Schwann cells. J Neuropathol Exp Neurol 1971;30:603-12.
12. Riggs HE, Clary WU. A case of intramedullary sheath cell tumor of the spinal cord; consideration of vascular nerves as a source of origin. J Neuropathol Exp Neurol 1957;16:332-6.
13. Nelson E, Rennels M. Innervation of intracranial arteries. Brain 1970;93:475-90.
14. Karatas A, Cebi IT, Salviz M, Kocak M, Selcuk T. Schwannoma of the nasal septum. Egypt J Ear Nose Throat Allied Sci 2016;17:185-8.