Subfrontal Schwannoma Mimicking Neuroblastoma: Case Report

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ABSTRACT

Computed tomography (CT), performed in a healthy 28-year-old man after minor head injury, detected a frontal base tumor. Neurological examination revealed left hyposmia. On magnetic resonance imaging scans, there was a heterogeneously enhanced tumor located in the left paramedian frontal base with extension into the left ethmoid sinus. Angiography showed a hypervascular mass in the left anterior cranial fossa; it was mainly fed by the left ethmoidal artery. Positron emission tomography scanning showed moderate accumulation of 11-methylmethionine and low accumulation of 18-fluorodeoxyglucose (FDG) at the tumor site. Bone image CT disclosed compressive, nondestructive deformation of the left frontal base. The preoperative diagnosis was olfactory neuroblastoma or meningioma. The tumor was totally resected via bifrontal craniotomy. The tumor was histologically diagnosed as typical schwannoma; it was positive for S-100 protein. We report a rare subfrontal schwannoma with extension into the nasal cavity that mimicked neuroblastoma. Low FDG accumulation and compressive deformation of the anterior skull base may help in the differential diagnosis of these tumors.

KEYWORDS: Subfrontal schwannoma, olfactory nerve, neuroblastoma, skull base

Schwannomas arise from the nerve sheaths of peripheral and cranial nerves. They account for 6 to 8% of all intracranial tumors.[1,2] They commonly arise from the vestibular nerve and less commonly from the fifth, ninth, and tenth cranial nerves. Schwannomas of the olfactory groove or subfrontal region are rare; 49 cases have been reported to date.[3–26] Because of their rarity, these tumors can be misdiagnosed preoperatively as meningioma or olfactory neuroblastoma. We report a rare anterior cranial fossa schwannoma with extension into the ethmoid sinus and highlight factors that can contribute to the preoperative differential diagnosis of these tumors.

CASE REPORT

This 28-year-old man underwent computed tomography (CT) after a minor head injury. It revealed a tumor at the anterior skull base. His neurological and general examinations were normal except for left hyposmia. The results of intravenous olfaction tests were within normal limits.

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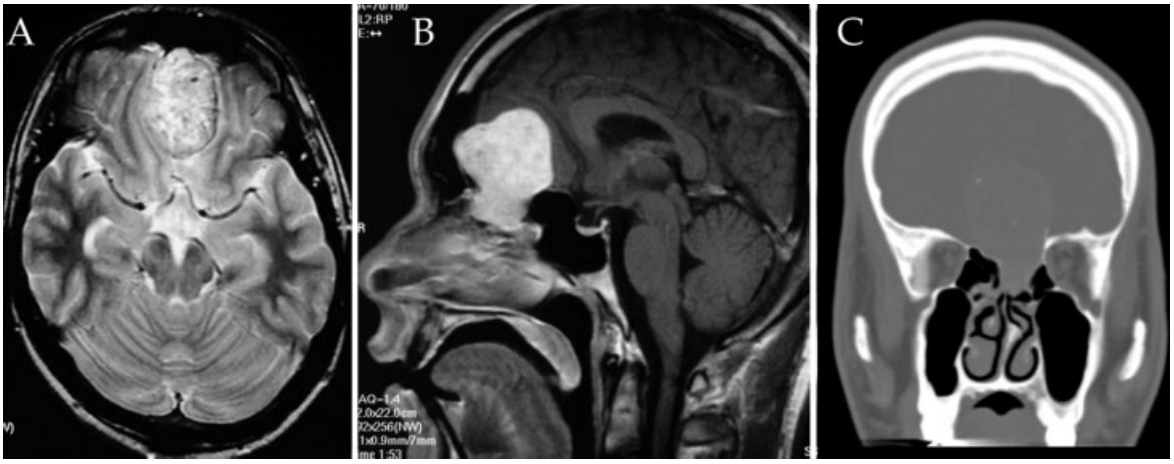

Figure 1 (A) Axial T2-weighted magnetic resonance imaging (MRI) revealing a subfrontal heterogeneously hyperintense mass. (B) Sagittal MRI with gadolinium demonstrating an enhanced subfrontal mass with extension to the ethmoid sinus. (C) Coronal computed tomography showing erosion of the left cribriform plate.

T2-weighted magnetic resonance images (MRI) revealed a subfrontal tumor; it was hyperintense and iso-mixed intense to the white matter (Fig. 1A). It extended into the left ethmoid sinus and was hypointense on T1-weighted images and heterogeneously enhanced (Fig. 1B). Coronal bone CT showed thinning of the left cribriform plate and medial endofrontal fovea with marked compression in the direction of the nasal cavity (Fig. 1C). A left carotid angiogram disclosed a hypervascular mass in the base of the anterior cranial fossa mainly; it was fed by the left anterior ethmoidal artery and displaced the anterior cerebral artery upward (Fig. 2). Positron emission tomography (PET) scanning with 18-fluorodeoxyglucose (FDG) and 11-methylmethionine was performed to rule out metastatic disease and carcinoma of the ethmoid sinus (Fig. 3). Methionine PET revealed moderate accumulation of the tracer compared with surrounding cerebral tissue; the tumor was depicted as a low-accumulation area on FDG PET. The preoperative diagnosis included neuroblastoma and olfactory groove meningioma.

We performed bifrontal craniotomy. Upon opening the dura mater, on the left frontal lobe we found an extra-axial mass partially attached to the dura of the left frontal base. It was elastic, hard, and hypervascular. After cauterization of the large feeding artery arising from the cribriform plate, we performed internal decompression. The tumor was separated from arachnoid tissue covering the frontal lobe. The dura mater of the frontal base was thin and almost absent at the lowermost part of the tumor. The bone of the frontal base was depressed toward the nasal cavity; however, the bone cortex was preserved. The left olfactory nerve was thinned and stretched medially but anatomically preserved. The tumor did not attach to the falx. It was totally resected. The defect in the left frontal base was filled with abdominal fat and covered with a periosteal flap secured with fibrin glue. The craniotomy was closed and dressed in standard fashion. Postoperative MRI confirmed total resection of the tumor (Fig. 4).

Histological examination disclosed proliferation of spindle cells with columnar nuclei exhibiting a fascicular pattern and focal nuclear palisading (Fig. 5). Loose myxoid stroma and hyalinized vessels were also noted. Some areas of fibrosis with calcification and vascular proliferation were noted. There was no necrosis or cellular atypia. Immunohistochemical staining revealed tumor-cell positivity for S-100 and CD57.

The patient's hyposmia improved postoperatively, and he was discharged without any neurological deficit at 19 days after the operation.

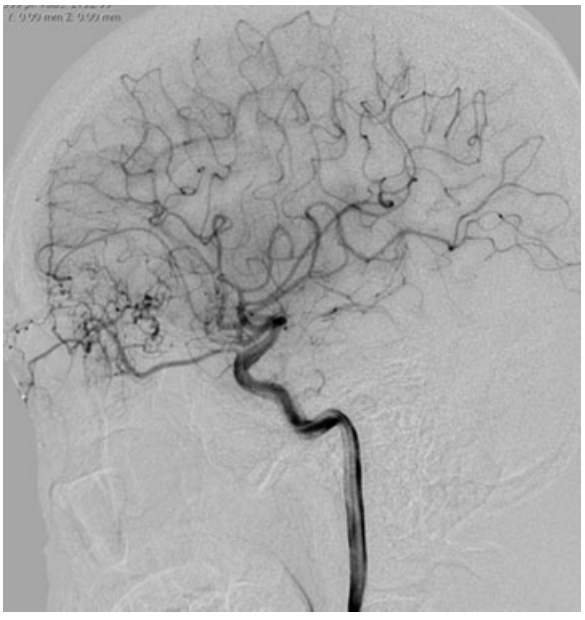

Figure 2 Digital subtraction angiogram with left internal carotid artery injection showing the hypertrophic ophthalmic artery feeding the subfrontal mass.

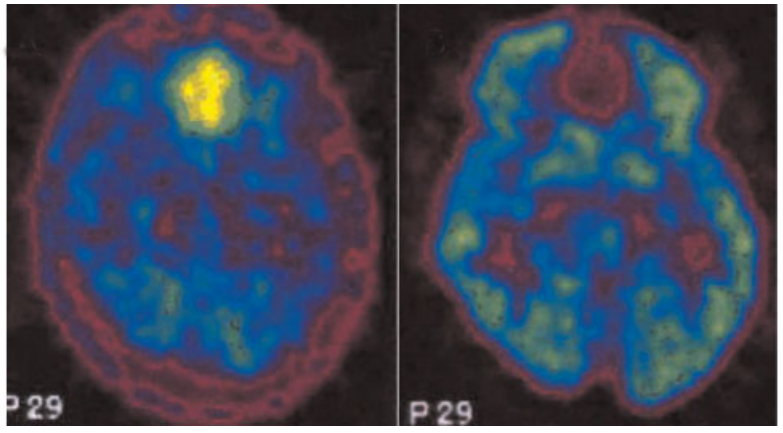

Figure 3 (A) Methionine positron emission tomography (PET) imaging showing a moderate-attenuation lesion in the frontal lobe. (B) The tumor was not identified on fluorodeoxyglucose PET.

DISCUSSION

Schwannomas are benign, slowly growing nerve sheath tumors that usually arise from peripheral nerves containing Schwann cells, including distal portions of the cranial nerve. The tumor in our patient was located in the subfrontal subdural space adjacent to the olfactory tract. As the olfactory nerve does not have a Schwann cell layer, theoretically, schwannoma cannot arise at this nerve.[18,19] Hypotheses on the possible origin of subfrontal schwannoma focus on developmental and nondevelopmental origins.[15,19] The developmental hypothesis proposes that they arise from aberrant Schwann cells. Others suggested that these tumors originate from either multipotent mesenchymal cells or displaced neural crest cells that form foci of Schwann cells, termed *schwannosis*, within the central nervous system parenchyma.[27,28] The

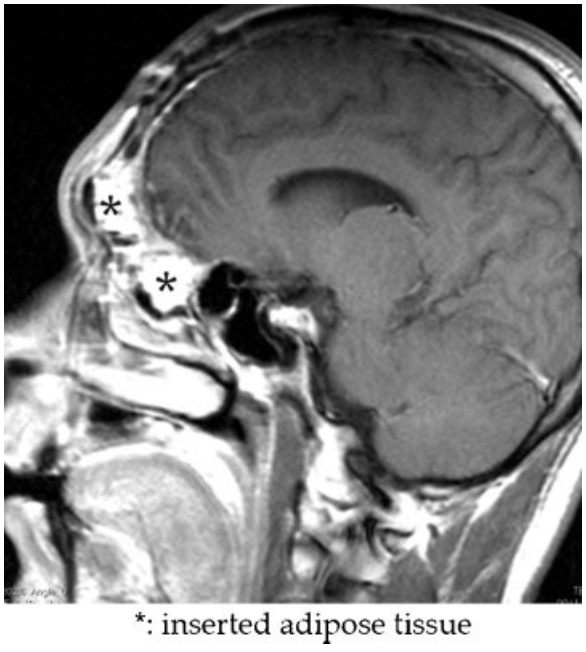

Figure 4 Postoperative magnetic resonance imaging with gadolinium demonstrating total resection of the tumor and a small hemorrhagic scar at the tumor site.

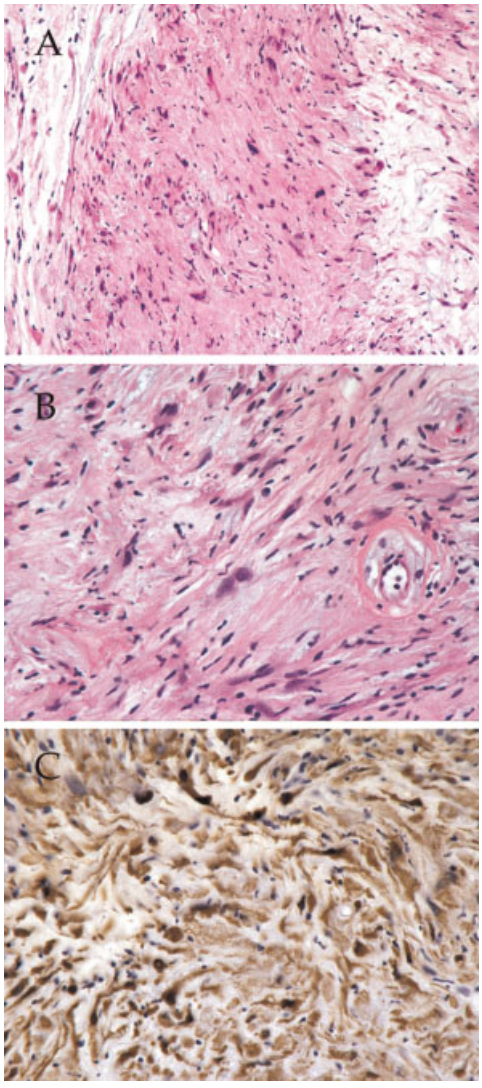

Figure 5 Histopathologic examination of the surgical specimen showed that the tumor consisted of alternating areas of compact, elongated cells (Antoni type A) and less cellularized areas (Antoni type B) (hematoxylin and eosin; A × 50, B × 100). On immunohistochemical staining, the tumor cells were positive for S-100 protein (C) (×100).

nondevelopmental hypothesis proposes that Schwann cells in perivascular nerve plexi surrounding cerebral arterioles and large arteries in the subarachnoid space develop into schwannomas.[29] Schwann cells are also seen in association with meningeal branches of the trigeminal and anterior ethmoidal nerves innervating the anterior cranial fossa and olfactory groove. In our case, the tumor was attached to the dura of the frontal base and grew upward and toward the ethmoid sinus without destroying the bone cortex. Therefore, we concluded that its most likely origin was the meningeal branch of the trigeminal nerve on the dura mater of the frontal base.

Including our patient, there are 49 reported cases of subfrontal or olfactory groove schwannomas.[3–26] Of these, three occurred in patients with neurofibromatosis.[19] The patients' age ranged from 14 to 63 years (average 33 years) with a 2:1 male predominance. These tumors are different from meningiomas arising in the same area in that meningiomas are seen in older age groups and meningiomas are different from schwannomas arising at other sites where there is a female predominance.[19] Patients usually present with headache, seizures, anosmia, frontal lobe dysfunction, and signs and symptoms related to elevated intracranial pressure.[16,19]

As shown in Table 1, of the 49 documented schwannomas, 16 (33%) extended into the nasal cavity.[6–8,10,12,13,19,21,23,24,26] They included four intranasal schwannomas that extended into the anterior cranial fossa.[6–8,10] Radiographic features of the frontal base were bony erosion ($n = 6$) and bony destruction ($n = 2$). With the exception of tumors preoperatively diagnosed by biopsy, they were preoperatively misdiagnosed as neuroblastoma, sinonasal malignancy, infectious disease, and meningioma. Our preoperative misdiagnosis of olfactory neuroblastoma or meningioma was based on radiological findings of extension into the nasal cavity and hypervascularity.

The differential diagnosis of tumors involving the extra-axial anterior cranial fossa and cribriform plate with extension to the ethmoid sinus should include meningioma, schwannoma, olfactory neuroblastoma, and metastatic disease.[16,17,19,24] Among these differential diagnoses, neuroblastoma outranked schwannoma based on imaging features and the sex and age distribution of neuroblastoma.[16] However, the correct diagnosis of schwannoma is essential because neuroblastomas require more aggressive craniofacial resection.[16,30] Neuroblastoma and ethmoid carcinoma can invade the paranasal sinuses and cause marked bony destruction.[16,20,31,32] On the other hand, schwannomas are slow-growing tumors characterized by expansion and thinning of the confining bone.[6,19] In our case, intraoperative findings were sufficient for a diagnosis of benign tumor because the lesion manifested a well-demarcated margin and there was no bone destruction. The absence of bone destruction may represent a strong radiological clue for the differentiation between olfactory neuroblastoma and other benign diseases.

Yu et al reported an esthesioneuroblastoma that revealed strong 18-FDG uptake on PET scans and, as in our case, Sakamoto et al found that schwannoma could not be identified on the same modality.[33,34] Ours is the first methionine PET study of a subfrontal schwannoma; by this modality, it was revealed as an area with moderate tracer accumulation, indicating that PET yields useful

Table 1 Summary of Reported Cases of Subfrontal or Olfactory Schwannoma with Extension to Nasal Cavity

No.	First Author/Year	Age/Sex	The Bone of the Frontal Base	Preoperative Diagnosis	Remarks
1	Von Strum/1968[3]	27/M	ND	ND	
2	Mauro/1983[35]	44/M	Bony erosion	ND	
3	Zovickian/1986[6]	40/M	ND	Preoperative biopsy	Nasal schwannoma
4	Nagao/1991[36]	63/F	ND	ND	
5	Enion/1991[7]	28/M	ND	ND	Nasal schwannoma
6	Bando/1992[37]	55/F	ND	ND	
7	Harada/1992[38]	33/M	ND	Meningioma	
8	Bavetta/1993[8]	41/M	Bony destruction	Preoperative biopsy	Nasal schwannoma
9	Gatsher/1998[39]	50/F	ND	ND	Nasal schwannoma
10	Carron/2002[12]	59/F	Bony erosion	Neuroblastoma/sinonasal malignancy	
11	de Souza/2002[13]	27/M	ND	Infectious disease	
12	Bezircioglu/2008[40]	33/F	Bony destruction	Preoperative biopsy	
13	Kanaan/2008[23]	14/M	Bony erosion	Preoperative biopsy	Endonasal resection
14	Mirone/2009[26]	38/M	Bony erosion	ND	
15	Choi/2009[24]	39/F	Bony erosion	ND	
16	Present case	28/M	Bony erosion	Neuroblastoma/meningioma	Hypervascular

ND, not described.

information on the proliferative nature of tumors that is essential for the differential diagnosis of subfrontal tumors.

CONCLUSION

We reported a rare case of subfrontal schwannoma with extension into the nasal cavity. We posit that its origin was the meningeal branch of the trigeminal nerve in the frontal base. Subfrontal schwannoma should be included in the differential diagnosis of tumors of the anterior cranial fossa with extension into the nasal cavity.

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