DOI: 10.5137/1019-5149.JTN.14122-15.0
Received: 20.01.2015 / Accepted: 11.02.2015
Published Online: 11.07.2016

Case Report

Ancient Olfactory Schwannoma - Case Report and Literature Review

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ABSTRACT

Intracranial schwannomas are benign tumors that arise from Schwann cells. Since it is well known that optic and olfactory nerves do not have a Schwann cell sheath, schwannoma should not develop from these nerves.

We report a very unusual case of a 73-year-old female who presented with generalized seizures and had radiological features of an intracranial aneurysm. Additional imaging showed an extracerebral mass 2.5 x 2.0 cm in size, which most likely corresponded to a meningioma. It was resected in total. Subsequent histological analysis revealed that the tumor was in fact ancient schwannoma WHO grade I.

Only about 41 case of olfactory schwannoma have been reported in the literature. Olfactory groove schwannomas are extremely rare tumors, occurring less frequently than any other intracranial nerve schwannoma. As in this case, the schwannoma should be included in the differential diagnosis of the anterior cranial fossa tumor. Further research on the pathogenesis and the origin of olfactory groove schwannoma is needed.

KEYWORDS: Intracranial tumors, Neurinoma, Olfactory dysfunction, Olfactory groove, Subfrontal approach

INTRODUCTION

Intracranial schwannomas are benign tumors that arise from Schwann cells. Since it is well known that optic and olfactory nerves do not have a Schwann cell sheath, schwannoma should not develop from these nerves (17).

Schwannomas account for 8 - 10 % of all intracranial tumors and occur mostly in patients between 20 and 50 years of age. Patients affected by schwannomas may be asymptomatic or present with various symptoms depending on the location, size and nerve of origin, and may include motor and sensory dysfunction, intracranial hypertension, headache and seizures (20).

These tumors present on Magnetic Resonance Imaging (MRI) as well-circumscribed, encapsulated masses, low-to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The signal intensity on T2-weighted images may be either homogeneously hyperintense or it can show a characteristic high signal intensity in the periphery and low signal intensity in the central region of the lesion. This MR imaging finding corresponds with pathologic findings to fibrous tissue (with high collagen content) centrally and more myxoid tissue peripherally (20).

Histopathologically, it consists of compact hypercellular Antoni A areas and myxoid hypocellular Antoni B areas. Cells are narrow, elongated and wavy with tapered ends interspersed with collagen fibers – spindle cells. Nuclear palisading around fibrillary processes (Verocay bodies) are often seen in cellular areas. Large irregularly spaced vessels, usually with thickened hyalinized walls and thrombi, are most prominent in Antoni B areas. Tumor cells have ill-defined cytoplasm, dense chromatin. It often displays degenerative nuclear atypia, but

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rare mitotic figures. There are no axons except where nerve is attached.

Immunohistochemical staining helps differentiating from other tumors that can present with similar histological characteristic - fibroblastic meningiomas, tanacytic ependymomas, subependymomas and pilocytic astrocytomas. Schwannomas contain Leu-7 and S-100 protein.

Different course of treatment for schwannomas include surgical excision, observation and radiotherapy (10).

CASE REPORT

A 73-year-old female presented with the occasional generalized seizures over a two-year period and headaches over a two-month period. Neurological examination on admission was entirely normal, revealing preserved olfaction and no other abnormality. There were no symptoms of increased intracranial pressure or a focal neurological deficit. No family history of Neurofibromatosis type I (Von Recklinghausen disease) was present.

A Computerized Tomography (CT) scan was performed which showed a hyperdense mass in the right subfrontal region. The initial diagnosis was an intracranial aneurysm, so the patient was admitted for further examination. A Magnetic Resonance Imaging (MRI) scan with Magnetic Resonance Angiography was performed and showed an extracerebral mass 2.5x2.0 cm in size, which most likely corresponded to an intracranial meningioma, with compression on both anterior cerebral arteries (ACA) and ischemia in the vascularization area of the right anterior cerebral artery (ACA). No evidence of extension into the ethmoid sinus was seen (Figure 1A-F).

The decision was made to proceed with the resection of the tumor.

A right subfrontal craniotomy was performed. After the initial exposure of the tumor, which was solid, adherent to the olfactory nerve, fibrous and calcified and surrounded by glial tissue, the tumor was totally resected. Because of tight adherence to the olfactory nerve, part of it was resected with the tumor (Figure 1A-F).

During histopathological analysis, on gross examination there were multiple, yellow to pearly white soft tissue pieces measuring 2.5 x 2.0 cm altogether. Some of the soft tissue pieces showed areas of calcification. On microscopic examination, the tumor was hypocellular, although parts of it comprised of sweeping fascicles of slender elongated spindle cells with wavy serpentine nuclei and formation of Verocay bodies in some areas (Antoni A). These cells showed some pleomorphism and hyperchromatic nuclei, but mitotic figures were not evident. Occasional vessels displayed a periluminal hyaline ring. There were also areas of calcification. Immunohistochemical studies revealed that the tumor cells were positive for S-100 protein. The final diagnosis was schwannoma WHO grade I, with distinct degenerative changes, a so called “ancient schwannoma” (Figure 2A, B).

During the postoperative course, the patient did not have any seizures, but reported anosmia. Postoperative MRI confirmed total resection of the tumor. The patient was discharged on the 11th day after surgery with anosmia and no seizures. On the follow-up examination after six months she regained olfaction.

The patient has consented to the submission of the case report for submission to the journal.

DISCUSSION

The most common origin of intracranial schwannomas is the vestibular branch of the eighth cranial nerve.

The PubMed database was searched online (PubMed, http://pubmed.com). A search query using the terms olfactory schwannoma and subfrontal schwannoma in titles and/or abstracts revealed an additional 41 case (Table I). The mean age of the 41 patients, including our patient, was 31.85 years, which was significantly younger than our reported case. In addition, there were more males (65.85%) than females, the male/female ratio being 1.93:1. A generalized seizure was found only in one other case.

This particular type of schwannoma was first reported in conjunction with central neurofibromatosis – Morbus Von Recklinghausen (7,20), although there were no indications that our patient had this disease.

The origin of these lesions in the subfrontal region has always been enigmatic and many hypotheses have been put forth. The developmental theories suggest either transformation of mesenchymal pial cell into ectodermal Schwann cells or migration of the neural crest cells within the central nervous system (10). Also, dural branches of peripheral nerves which pass through the anterior cranial fossa may be nerves of origin for these tumors. Another theory suggests that reactive change after injury forms Schwann cells from multipotent mesenchymal cells (1).

Although literature shows that the majority of the reported cases were younger or middle aged males (Table I), our patient was elderly female, which makes this case even more interesting.

Histological type of the tumor in our case is above all fascinating. Because the tumor was discovered at late age, it can be concluded that it has been growing slowly, for a long period of time, and during its growth, many degenerative process took place, shaping our tumor into an ancient schwannoma. This histological type of schwannoma display pronounced degenerative changes in the form of cyst formation, calcification, hemorrhages and hyalinization. The tumor is usually infiltrated by a large number of foamy macrophages. Schwann cell nuclei can show atypical nuclei, which can be regarded as purely degenerative changes. The presence of hypercellularity and atypia my lead to the misdiagnosis of these lesions as sarcomas (melanomas, leiomyosarcomas, hemangiopericytomas). The absence of mitotic activity is the key feature to differentiate benign ancient schwannomas from malignant schwannomas. The presence of a capsule, evidence of prior hemorrhage, thick-walled

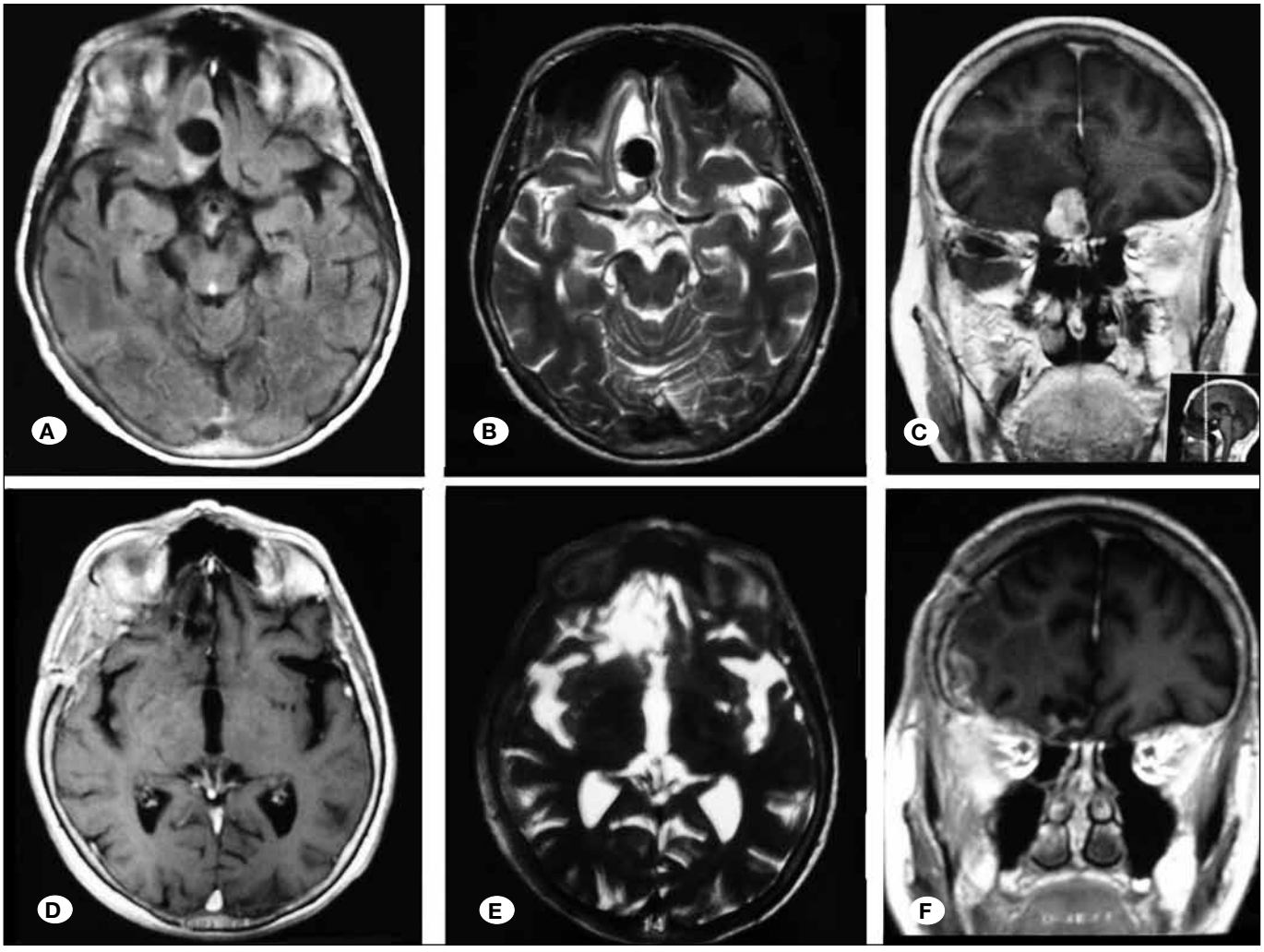

Figure 1: Magnetic Resonance Imaging (MRI) showing an extra-axial mass in the right frontal region near midline and elevating the olfactory bulb: **A)** Axial T1-weighted, **B)** axial T2-weighted and **C)** coronal T1-weighted preoperative MRI; **D)** Axial T1-weighted, **E)** axial T2-weighted and **F)** coronal T1-weighted postoperative MRI.

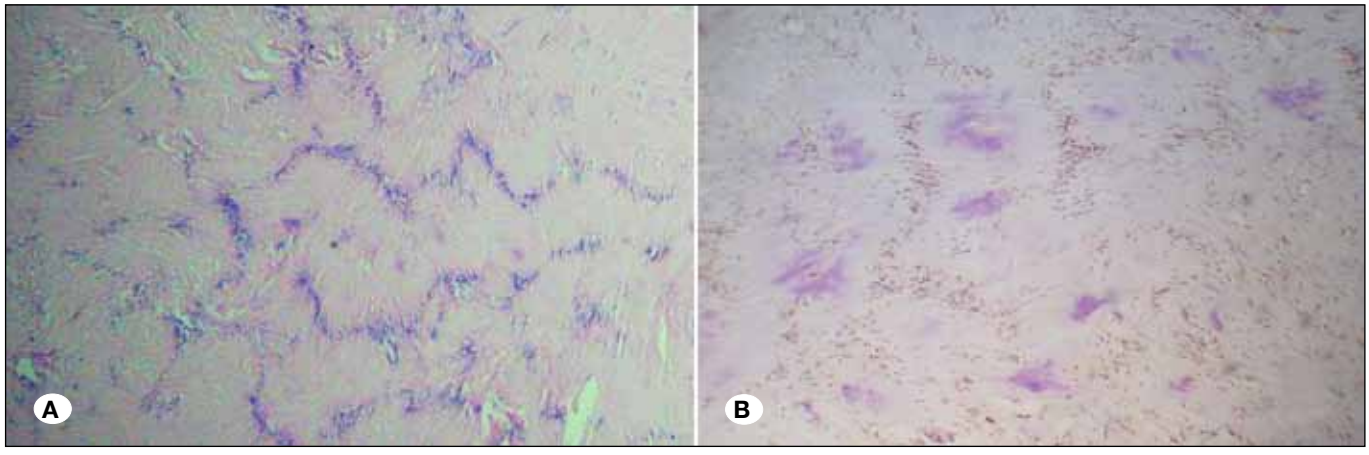

Figure 2: Microphotographs of the tumor: **A)** The tumor was hypocellular, although parts of it comprised of sweeping fascicles of slender elongated spindle cells with wavy serpentine nuclei and formation of Verocay bodies in some areas (Antoni A). These cells showed some pleomorphism and hyperchromatic nuclei, but mitotic figures were not evident. Occasional vessels displayed a periluminal hyaline ring. There were also areas of calcification; **B)** Immunohistochemical examination showed the tumor cells were positive for S-100 protein.

Table I: Summary of 41 Cases of Olfactory Schwannoma Found in the Literature

No.	Author	Age (years)	Gender	Clinical presentation	Anosmia	Radiological features	Olfactory tract
1	Strum et al. (33)	27	M	Headache	Yes	/	Not seen
2	Viale et al. (39)	22	M	Visual deficit	Yes	/	/
3	Ulrich et al. (37)	19	M	Seizures, visual deficit	Yes	/	/
4	Vassilouthis and Richardson (38)	17	M	Visual and cognitive deficit	Yes	Cystic, heterogeneous	/
5	Mauro et al. (19)	44	M	Visual and memory deficit	/	Cystic, heterogeneous	/
6	Sato et al. (31)	22	M	Seizures	Yes	Solid, homogenous	Thinned
7	Nagao et al. (23)	63	F	Memory impairment	No	Cystic heterogeneous	/
8	Husain et al. (14)	55	F	Headache	Yes	/	/
9	Harada et al. (12)	33	M	Headache	Yes	Solid, heterogeneous	Thinned
10	Sabel and Teepen (28)	17	M	Seizures	/	Solid, homogeneous	/
11	Huang et al. (13)	33	M	Headache, visual deficit	No	Solid, homogeneous	/
12	Boyd et al. (5)	29	F	Seizures, headache	Yes	Cystic, heterogeneous	Involved
13	Praharaj et al. (25)	45	M	Seizures, headache	/	Solid, homogeneous	Not seen
14	Timothy et al. (35)	33	F	Seizures	No	Solid, homogeneous	Not seen
15	Gelabert et al. (11)	19	M	Seizures	No	Solid, homogeneous	/
16	Tan et al. (34)	21	M	Seizures	No	Solid, homogeneous	/
17	Tsai et al. (36)	31	F	Seizures, headache	/	Solid, heterogeneous	/
18	Yang et al. (41)	55	M	Headache	No	Solid, homogeneous	/
19	Carron et al. (6)	59	F	Headache, rhinorrhea	No	Solid, homogeneous	Involved
20	Amador et al. (3)	24	F	Hypoesthesia, visual deficit	/	Cystic, heterogeneous	Not seen
21	de Souza et al. (9)	27	M	Headache	Yes	Cystic, homogeneous	/
22	Prasad et al. (27)	19	M	Seizures	Yes	Solid, heterogeneous	/
23	Yuen et al. (42)	33	F	Seizures	No	Solid homogeneous	Involved
25	Murakami et al. (22)	30	M	Headache	No	Solid, homogeneous	Thinned
26	Shenoy and Raja (32)	55	M	Seizures	No	Cystic, heterogeneous	Thinned
27	Sano et al. (30)	44	M	Seizures	No	Solid, heterogeneous	/
24	Komoribayashi et al. (16)	37	F	Seizures	Yes	Solid, homogeneous	Not seen
28	Yako et al. (40)	14	M	Headache	Yes	Solid, heterogeneous	Not seen
29	Ahmad et al. (2)	23	M	Seizures	No	Solid, heterogeneous	/
30	Prak et al. (26)	16	M	Headache, diplopia	No	Solid, heterogeneous	Thinned
31	Adachi et al. (1)	22	F	Seizures	No	Solid, heterogeneous	Involved
32	Saberi et al. (29)	35	F	Seizures, headache, diplopia	Yes	Cystic, heterogeneous	/
33	Daglioglu et al. (8)	21	M	Headache, aggressiveness	/	Cystic, heterogeneous	/
34	Kanaan et al. (15)	14	M	Headache, cognitive impairment	Yes	Cystic, heterogeneous	/

Table I: Cont.

No.	Author	Age (years)	Gender	Clinical presentation	Anosmia	Radiological features	Olfactory tract
35	Bezircioglu et al. (4)	33	F	Headache	Yes	Solid, heterogeneous	/
36	Mirone et al. (21)	38	M	Headache	Yes	Cystic, heterogeneous	/
37	Choi et al. (7)	39	M	Headache	Yes	Cystic, heterogeneous	Involved
38	Figueiredo et al. (10)	49	M	Headache	Yes	Cystic, heterogeneous	Thinned
39	Lin et al. (18)	32	M	Seizures	/	Solid, heterogeneous	/
40	Li et al. (17)	16	F	Seizures	No	Solid, heterogeneous	/
41	Ogino-Nishimura et al. (24)	41	F	Headaches	Yes	Cystic, heterogeneous	Involved

M: *Male,* ***F:*** *Female.*

vascular structures and areas representing degenerative changes also suggest a diagnosis of a benign lesion (21).

Treatment options for schwannomas include surgical excision, observation and radiotherapy, particularly Gamma knife therapy. Because the olfactory groove is an extremely rare location for these masses and is relatively difficult to biopsy, surgery is the optimal course of treatment. Complete removal of the tumor is necessary to prevent local recurrence.

CONCLUSION

Olfactory groove schwannomas are extremely rare tumors, occurring less frequently than any other intracranial nerve schwannoma. As in this case, the schwannoma should be included in the differential diagnosis of the anterior cranial fossa tumor. One of the objectives of this case report is to demonstrate that a benign tumor arising solely from the olfactory bulb can act as a primary epileptic focus and to illustrate the difficulty in making a preoperative diagnosis of pathological lesions in this area. Further research on the pathogenesis and the origin of olfactory groove schwannoma is needed. Complete removal is curative, but subtotal resection may be an acceptable option for the slow-growing nature of these tumors.

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