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CASE REPORT

Solitary olfactory schwannoma without olfactory dysfunction: a new case report and literature review

Y. P. Li · S. Jiang · P. Z. Zhou · Y. B. Ni

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Abstract Schwannomas are rare and seldom extend into the anterior cranial fossa. Herein, we report a case of schwannoma arising from the olfactory groove in a 16-year-old girl who presented with generalized seizures without olfactory dysfunction or other neurologic deficits. Computerized tomography (CT) scan showed a large mass with abundant calcification located in the olfactory groove, which was confirmed as a schwannoma by histology and totally resected via basal subfrontal approach. The presentation, imaging findings and histogenesis of the tumor are discussed along with a review of the pertinent literature.

Keywords Olfactory groove · Subfrontal · Schwannoma · Olfactory dysfunction

Y. P. Li · S. Jiang (✉) · P. Z. Zhou
Department of Neurosurgery, West China Hospital, Sichuan University, Chengdu 610041, Sichuan, People's Republic of China
e-mail: Leeurop@gmail.com

Y. P. Li
e-mail: andybile@gmail.com

P. Z. Zhou
e-mail: 150983737@qq.com

Y. B. Ni
Department of Pathology, West China Hospital, Sichuan University, Chengdu 610041, Sichuan, People's Republic of China
e-mail: 759230307@qq.com

Introduction

Intracranial schwannomas arise predominantly from the vestibular branch of the eighth cranial nerve and rarely from the olfactory groove. Olfactory groove schwannomas are rare and likely to be misdiagnosed as olfactory groove meningiomas, dural-based metastasis or neuroblastomas. Only about 30 cases of olfactory groove meningioma have been reported in the world literature, and most of these cases were associated with olfactory dysfunction, while only 13 cases were reported to have no olfactory dysfunction. In this article, we discuss a rare case of schwannoma arising from the olfactory groove in a 16-year-old girl who presented with generalized seizures without olfactory dysfunction.

History

A 16-year-old girl presented with episodes of generalized seizures in the past 15 years, and the attacks became more frequent in recent 2 years prior to admission in a local hospital, where a large subfrontal mass was noticed on computerized tomography (CT) scan (Fig. 1a). The patient was transferred to our hospital for further evaluation and treatment. Neither significant abnormality was elicited from the general physical examination on admission nor was any focal neurologic deficit detected on neurologic examination.

CT scan demonstrated a 5 × 4 × 3 cm mass located in the right frontobasal region adjacent to the anterior falx, accompanied with minimal perifocal edema (Fig. 1a). The mass was heterogeneously intensed with a large amount of calcification around the rim. T2-weighted magnetic resonance images revealed a heterogeneously hyperintensed

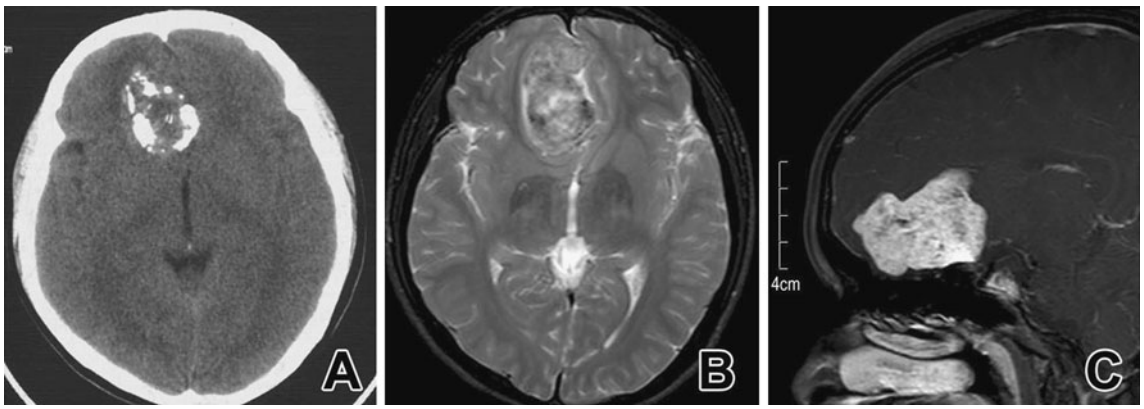

Fig. 1 **a** CT scan shows a large mass located in the right frontobasal region adjacent to the anterior falx. The mass is heterogeneously intense, with a large amount of calcification. **b** Axial T2-weighted MRI shows a huge subfrontal heterogeneously hyperintense mass. **c** Sagittal T1-weighted MRI with gadolinium shows strong contrast enhancement of the lesion

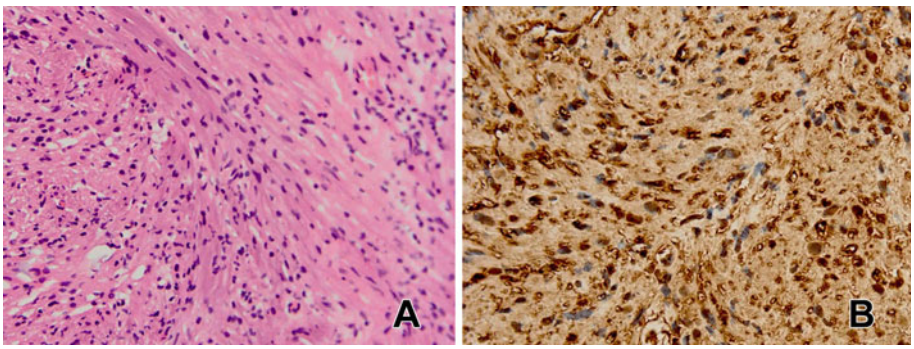

Fig. 2 **a** Photomicrography shows the characteristic features of schwannoma composed of benign spindle-shaped cells, with elongated nuclei and fibrillary cytoplasm (Antoni A pattern), and less cellular, loosely textured tumor areas (Antoni B) (H&E 200×). **b** Immunohistochemistry shows that the tumor cells are positive for S-100 protein (400×)

mass in the right frontal region near the midline, causing elevation of the right frontal lobe. The mass was heterogeneously enhanced after IV administration of gadolinium (Fig. 1b, c). No evidence of dural tail or extension into the ethmoid sinus was seen.

Surgical procedures

Right frontal craniotomy was performed. A 5-cm extra axial tumor was found at the base of the anterior cranial fossa and totally resected. The tumor was solid, moderately vascular and firmly adhered to the right olfactory groove with abundant calcification. The right olfactory tract was not identifiable. After total resection of the tumor, the cribriform plate of ethmoid bone was seen erosive. The gross appearance of the tumor looked like an olfactory groove meningioma.

Histopathology

Histologic examination revealed that the resected tissue was composed of spindle-shaped cells, with elongated nuclei and fibrillary cytoplasm (Antoni A pattern), and less cellular and loosely textured tumor areas (Antoni B). Immunohistochemistry showed that the tumor cells were positive for S-100 protein, confirming the diagnosis of schwannoma (Fig. 2).

The postoperative course was uneventful. Postoperative imaging confirmed gross-total resection of the tumor (Fig. 3). The patient was discharged 7 days after surgery without new neurologic deficits, and follow-up visits demonstrated normal neurologic function.

Literature review and analysis

The PubMed database was searched online (Pubmed, http://pubmed.com/) in the English language. Search query using the terms olfactory schwannoma and subfrontal schwannoma in titles and/or abstracts revealed additional 34 cases (Table 1). It was found that the age and sex distributions of the 35 patients including ours with subfrontal schwannomas were different from those of patients with ordinary intracranial schwannomas. The mean age of the 35 patients was 32.7 ± 14.0 years, which was younger than that of patients with schwannoma in other common

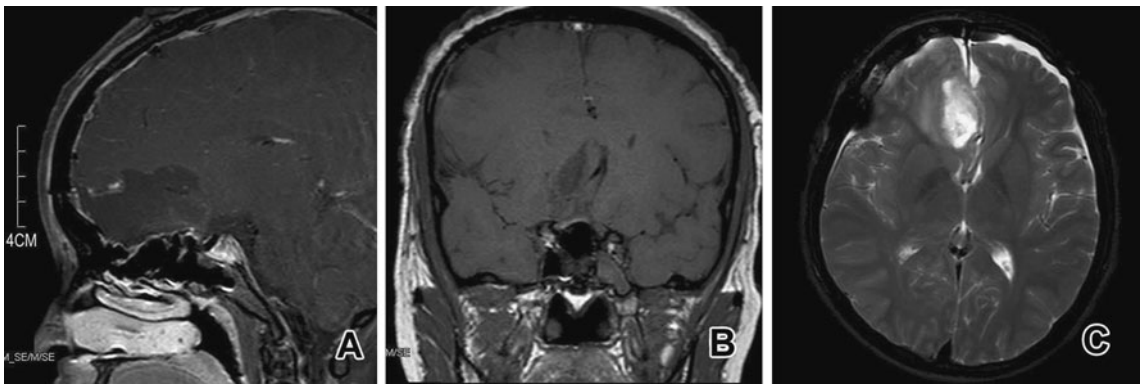

Fig. 3 Postoperative sagittal and coronal enhanced T1WI MRI and T2WI MRI shows complete removal of the tumor

sites. In addition, there were more males (57.1%) than females, the male/female ratio being 1.14:1 versus 1:1.5–2 for ordinary intracranial schwannomas.

Discussion

Schwannomas are benign tumors derived from Schwann cells. Theoretically, the olfactory nerve, as part of the central nervous system, does not contain any Schwann cells and cannot develop schwannomas. Thus, the origin of olfactory schwannomas is enigmatic. Various developmental and non-developmental hypotheses [1] have emerged in an attempt to explain the possible origin of olfactory schwannomas. The developmental theories include transformation of mesenchymal pial cells into ectodermal Schwann cells [2], and migration or displacement of neural crest within the substance of the central nervous system [3–5]. On the other hand, the non-developmental theories argue that olfactory schwannomas arise from Schwann cells normally present on adjacent structures such as anterior ethmoidal nerves innervating the anterior cranial fossa and the olfactory groove, the meningeal branch of the trigeminal nerve [6], the filia olfactoria which develop a Schwann cell layer about 0.5 mm beyond the olfactory bulbs, adrenergic nerve fibers innervating cerebral arterioles [7], a kind of 'ensheathing cell' of the olfactory nerve that expresses phenotypic features of both astrocyte and Schwann cell [2], and terminal nerve (cranial nerve 0) [8]. In addition, post-trauma reactive changes including formation of schwann cells from multipotential mesenchymal cells in patients with pathological changes like multiple sclerosis or infarction have also been described [9, 10].

Olfactory groove meningioma should be highly suspected in differential diagnosis of an extra-axial anterior cranial fossa mass involving the cribriform plate. However, subfrontal schwannoma can have similar neuroradiological features to olfactory groove meningioma, including extra-axial location, calcification, contrast enhancement and perifocal edema, thus making pre-operative differentiation of the two conditions difficult. Clinically, young age at presentation, the presence of bone scalloping on CT, the absence of dural tail sign and low vascularity may help us to make differential diagnosis between schwannoma and meningioma before surgery. In addition, bone erosion in olfactory schwannomas is usually destructive (17 of 35), while meningioma tends to induce hyperostosis (Table 1). Esthesioneuroblastoma and carcinoma of the paranasal sinus tend to be more aggressive, though they should be excluded from the differential diagnosis. Immunohistochemically, schwannomas always showed strong positive for S-100 and negative for EMA.

Olfactory groove schwannomas are classified into two main types by Adachi et al. [1]: schwannomas arising from the olfactory site such as the olfactory groove or cribriform plate, and those arising from non-olfactory sites. Accordingly, our case should belong to the first type as evidenced by the intraoperative findings. Recently, some researchers doubted whether these olfactory groove schwannomas are truly schwannomas. Yasuda et al. [11] individualized the first case of olfactory ensheathing cell tumor (OECT) based on immunohistochemical findings and suspected OECs as one of the origins of olfactory schwannoma. Embryologically, OECs derive from olfactory placodes, whereas Schwann cells originate from the neural crests. Despite the different origin of their cells, olfactory groove schwannoma and OECT have similar clinical, imaging, and histologic features, and can only be distinguished by immunohistochemical staining.

Conclusion

Olfactory groove schwannomas are extremely rare tumors, occurring less frequently than any other intracranial nerve

Table 1 Summary of 35 cases of olfactory groove schwannoma

No.	Author	Year	Age (years)	Sex	Main initial symptom	Olfaction	Calcification	Enhanced	Aspect	Bone erosion	Attachment	Detection of intact olfactory nerve(s)
1	Our case	2010	16	F	Convulsion	Normal	Yes	Hetero	Solid	Yes	OG	Not detected
2	Mirone et al. [12]	2009	38	M	Headache	Left hyposmia	Unknown	Hetero	Cystic-solid	Yes	OG	Not detected
3	Martine-Soto et al. [13]	2009	54	M	Headache, dysphasia	Normal	No	Hetero	Solid	Yes	CP	Not detected
4	Figueiredo et al. [14]	2009	49	M	Headache	Anosmia	Unknown	Hetero	Cystic-solid	Yes	Unknown	Both were involved and adherent to skull base dura
5	Choi et al. [15]	2009	39	F	Headache	Anosmia	Yes	Hetero	Cystic-solid	Yes	CP	Relateted to the tumor
6	Saberi et al. [2]	2008	35	F	Convulsion, diplopia and headache	Left hyposmia	Yes	Hetero	Cystic-solid	Yes	Unknown	Involved in tumor
7	Kanaan et al. [16]	2008	14	M	Headache, decling school performance and weight loss	Hyposima	Unknown	Hetero	Cystic-solid	Yes	Ethmoid sinus	Unknown
8	Daglioglu et al. [17]	2008	21	M	Headache, aggressive behavior	Unknown	Unknown	Hetero	Cystic	Yes	Right OG	Adhere to the tumor
9	Bezircioglu et al. [18]	2008	33	F	Headache	Anosmia	Unknown	Hetero	Solid	Yes	Unknown	Unknown
10	Adachi et al. [1]	2007	22	F	Convulsion	Normal	Yes	Partial	Solid	Unknown	CP	Thinned
11	Yako et al. [19]	2005	14	M	Headache, vomiting	Anosmia	Yes	Hetero	Cystic-solid	No	Left OG	On the right olfactory nerve was detected
12	Komoribayashi et al. [20]	2005	37	F	Convulsion	Anosmia	No	Total	Solid	No	Skull base dura	Not detected
13	Prasad et al. [21]	2004	19	M	Convulsion	Anosmia	Unknown	Hetero	Cystic-solid	Unknown	Left OG	Unknown
14	Sano et al. [22]	2004	44	M	Headache	Normal	No	Hetero	Solid	Unknown	Dura of the skull base	Unknown
15	Shenoy et al. [23]	2004	55	M	Convulsion	Normal	No	Hetero	Cystic	Unknown	Lateral to the CP	Thinned
16	Murakami et al. [24]	2004	50	M	Headache	Normal	No	Total	Solid	Yes	CP	Thinned
17	Yuen et al. [25]	2004	33	F	Convulsion	Normal	Unknown	Total	Solid	Yes	CP	Involved in tumor
18	de-Souza et al. [26]	2003	27	M	Headache	Anosmia	No	Total	Multicystic	Unknown	Unknown	Unknown
19	Amador et al. [6]	2002	24	F	Hypoesthesia on the left side of face, impaired vision	Unknown	No	Hetero	Cystic	Yes	OG	Not detected
20	Carron et al. [27]	2002	59	F	Headache	Normal	No	Total	Solid	Yes	Left OG	Adhere to the tumor
21	Tsai et al. [28]	2001	31	F	Headache, convulsion	Unknown	No	Hetero	Solid	Unknown	OG	Unknown

Table 1 continued

No.	Author	Year	Age (years)	Sex	Main initial symptom	Olfaction	Calcification	Enhanced	Aspect	Bone erosion	Attachment	Detection of intact olfactory nerve(s)
22	Tan et al. [29]	2001	21	M	Convulsion	Normal	Unknown	Well	Solid	Yes	Lateral to the CP	Unknown
23	Gelabert et al. [30]	2000	19	M	Convulsion	Normal	Unknown	Unknown	Unknown	Unknown	CP	Unknown
24	Praharaj et al. [31]	1999	45	M	Headache, convulsion	Unknown	No	Well	Solid	Unknown	CP	Not detected
25	Timothy et al. [32]	1999	33	F	Convulsion	Normal	Unknown	Well	Solid	Unknown	Crista galli	Not detected
26	Boyd et al. [33]	1997	29	F	Headache, convulsion	Hyposima	No	Hetero	Cystic	Yes	CP	Surrounded tumor
27	Huang et al. [10]	1997	33	M	Headache, leathargy, LOC decreased eye vision	Normal	Unknown	Well	Solid	Unknown	CP	Unknown
28	Sabel et al. [34]	1995	17	M	Convulsion	Unknown	No	Well	Solid	Unknown	Skull base dura	Not thinned
29	Bando et al. [35]	1992	55	F	Hyposmia, defect of visual field	Hyposima	Yes	Hetero	Unknown	Yes	Not detected	Not detected
30	Harada et al. [36]	1992	33	M	Headache	Hyposima	No	Hetero	Solid	Unknown	OG	Thinned
31	Nagao et al. [37]	1991	63	F	Recent memory disturbed	Normal	No	Hetero	Cystic	No	Lateral to the CP	Unknown
32	Sato et al. [38]	1985	22	M	Convulsion	Anosmia	No	Well	Solid	No	OG	Thinned
33	Vassilouthis et al. [39]	1980	17	M	Difficulty in maintaing concerntration, forgetfullness, headache, dizziness and amaurosis	Hyposima	No	Well	Cystic	Yes	Falx	Unknown
34	Ulrich et al. [40]	1978	19	M	Epilesy, partially blind, anosmia, diminished corneal reflex, hypesthenia	Anosmia	Unknown	Unknown	Unknown	Unknown	Skull base	Not detected
35	Harano et al. [41]	1974	26	F	Convulsion	Normal	Unknown	Unknown	Cystic	Unknown	Skull base	Tumor distant from olfactory tract

CP cribriform plate, *OG* olfactory groove

schwannoma. Complete removal is curative, but subtotal resection may be an acceptable option for the slow-growing nature of these tumors. Schwannoma should be kept in mind in differential diagnosis of anterior cranial fossa neoplasms, especially in young males. Further research on the pathogenesis and the origin of olfactory groove schwannoma is needed.

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