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

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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

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 $5 \times 4 \times 3$ cm mass located in the right frontobasal region adjacent to the anterior falx, accompanied with minimal perifocal edema (Fig. 1a). The mass was heterogeneously intensified with a large amount of calcification around the rim. T2-weighted magnetic resonance images revealed a heterogeneously hyperintensified...
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 locations.
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
| No. | Author                  | Year (years) | Age | 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       | Related to the tumor               |
| 6   | Saberi et al. [2]       | 2008         | 35  | M   | Convulsion, diplopia and headache             | Left hyposmia | Yes           | Hetero   | Cystic-solid | Yes      | Unknown  | Involved in tumor                  |
| 7   | Kanaan et al. [16]      | 2008         | 14  | M   | Headache, declining 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                            |
| No. | Author                          | Year (years) | Age  | 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 CP | Unknown                                           |
| 23  | Gelabert et al. [30]            | 2000         | 19   | M   | Convulsion                                    | Normal    | Unknown      | Unknown Unknown | Unknown CP   | CP         | Unknown                                           |
| 24  | Praharaj et al. [31]            | 1999         | 45   | M   | Headache, convulsion                          | Unknown   | No           | Well Solid      | Unknown CP   | Not detected | Unknown                                           |
| 25  | Timothy et al. [32]             | 1999         | 33   | F   | Convulsion                                    | Normal    | Unknown      | Well Solid      | Unknown CP   | 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   | CP         | Unknown                                           |
| 28  | Sabel et al. [34]               | 1995         | 17   | M   | Convulsion                                    | Unknown   | No           | Well Solid      | Unknown Skull base dura | Not detected | 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 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 maintaining concentration, forgetfulness, headache, dizziness and amaurosis | Hyposima  | No           | Well Cystic     | Yes          | Falx        | Unknown                                           |
| 34  | Ulrich et al. [40]              | 1978         | 19   | M   | Epilepsy, 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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