Case Report
Olfactory schwannoma mimicking esthesioneuroblastoma: case report and review of the literature

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Abstract: Olfactory groove schwannoma is a quite rare tumor. Here, we report a case of a 21-years-old man who had a chief complaint of headaches and blurred vision. The patient was preoperatively misdiagnosed as esthesioneuroblastoma but was finally diagnosed as schwannoma. After the tumor was totally removed, the patient’s symptoms were improved and no other neurological signs and symptoms appeared in 12 months after operation. We then performed a MEDLINE search of pertinent literature; stress the age, gender, chief complaint, presence or absence of hyposmia, radiological features of schwannoma in this unusual location. Age varied from 14 to 64 years (mean = 32.4). There were 17 male and 19 female patients. The most common clinical manifestation were headache (n = 22, 61.1%) and convulsion (n = 12, 33.3%). Hyposmia was present in 17 cases (47.2%), absent in 14 cases (38.9%), and unreported in 5 cases (13.9%). Tumor was solid in 19 patients (52.8%), cysticsolid in 11 patients (30.6%), and cystic in 5 patients (13.9%). Radiological appearance was unreported in 1 case (2.8%). Homogeneous enhancement was observed in 14 cases (38.9%), while 21 cases showed heterogeneous enhancement (58.3%), and 1 case was unreported (2.8%). Also 15 cases (41.70%) showed bone erosion and 7 cases (19.40%) showed calcification on computer tomography scan. The diagnosis and treatment of this rare tumor are also reviewed.

Keywords: Olfactory groove, schwannoma, esthesioneuroblastoma

Introduction
Schwannoma are usually benign tumors that arise from well-differentiated Schwann cells [1]. Tumors involving the olfactory groove of the anterior cranial base are extremely rare lesions, for the olfactory tract (part of the central nervous system) contains oligodendrocytes not schwann-cells [2]. So far, only 35 cases with schwannoma have been reported in English to the best of our knowledge [2-35]. Because of their rarity, the olfactory schwannoma can be misdiagnosed preoperatively as meningioma, dural-based metastasis or neuroblastoma. A young patient with olfactory schwannoma mimicking esthesioneuroblastoma is reported in present case report.

Case report
A 21-years-old young patient with a chief complaint of headaches and blurred vision for 6 days sought medical advice in our hospital. Before coming to our hospital, the patient was diagnosed as the case with esthesioneuroblastoma in local hospital and did not receive any treatment.

The results of his neurological and general examinations were normal except for decreased vision. The results of olfaction tests and visual field tests were within normal limits. The MRI revealed a 61 × 52 × 40 mm extra-axial tumor with cystic change in the left frontal region. The tumor was hypointense on T1-weighed images (Figure 1A, 1B) and hyperintense on T2-weighed images (Figure 1C), and showed heterogeneous enhancement extending into the ethmoid sinus (Figure 1D-F). And the results of Magnetic resonance spectroscopy (MRS) indicated that the level of N-acetylaspartate (NAA) decreased, while the levels of choline (Cho) and creatine (Cr) increased, along with an obvious lactate
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According to these findings, the patient was suspected of suffering from esthesioneuroblastoma, and a bifrontal craniotomy was performed.

At surgery, a cystic-solid mass of yellowish-grey appearance with rich blood supply was found in the extraxial space. The dura and the cribriform plate were invaded by the tumor. After reducing the blood supply of the mass, all tumor tissues were removed under the microscope. Reconstruction was performed with the use of artificial dura and fascia. The further course was uneventful and the postoperative imaging showed the preceding mass was totally removed.

Histologic examination revealed that the tumor was composed of spindle-shaped cells with elongated nuclei and fibrillary cytoplasm (Antoni A pattern), and loosely textured with microcystic component regions (Antoni B) (Figure 2A). Immunohistochemically, the tumor cells were positive for S100 (Figure 2B, 2C) and non-immunoreactive for Desmin, CAM5.2, EMA, GFAP, CK7, PanCK, Myogenin, CD99, NF. The tumor tissues also showed focal positively for CD34, and the Ki-67 index was 3%(+). These results were confirmed by department of pathology of University of California, Los Angeles. Thus the diagnosis of olfactory groove schwannoma was established. The patient’s symptoms were improved and no other neurological signs and symptoms occurred after operation. The 12-months follow-up indicated that the patient did not have any clinical or neuroradiological evidences of recurrence (Figure 2D-F).

**Literature search and analysis**

PubMed search for all cases of olfactory groove schwannoma was conducted up to June 2016, and 36 Cases (including the current case) were analyzed for basic demographic features including age, gender, chief complaint, presence or absence of hyposmia, radiological features (Table 1). SPSS13.0 software was used to analyze the data. The used method was the compare means to calculate the average values. The rates of different indexes were acquired

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*Figure 1.* (A) Axial and (B) sagittal T1-weighted MRI reveals a 61*52*40 mm hypointense mass in the olfactory groove with a well-defined margin and minimal perifocal edema. (C) Axial T2-weighted MRI shows a mixed hyperintense mass. (D) Axial, (E) Sagittal and (F) Coronal enhanced T1-weighted MRI shows a heterogeneously hyperintense mass with multiple cystic change (White arrow) in the olfactory groove growing into the ethmoid sinus.
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Schwannoma of the olfactory groove is extremely rare, for the olfactory nerve contains oligodendrocytes not Schwann-cells. Up to now, only 35 cases have been reported in English. There are several hypotheses which may explain the probable origin of olfactory groove schwannoma. The developmental theories hold that these lesions primarily arise from aberrant Schwann cells, such as transformation of mesenchymal pial cells into ectodermal Schwann cells, and migration or displacement of neural crest elements [25, 28, 32]. The non-developmental theories postulate that olfactory schwannoma arise from Schwann cells which normally present on adjacent structures. For example, Adachi performed an extensive review of the literature and postulated that these tumors originate from the filaolfactoria which acquire Schwann cells 0.5 mm beyond the olfactory bulb, the embryonic terminal nerve, and the nerve plexus of dural vessels [21]. In addition, the anterior ethmoidal nerve and the ramus meningeus of the trigeminal nerve have both been proposed as the source of an olfac-

Figure 2. Histologic examination revealed that the tumor was composed of spindle-shaped cells, and loosely textured with microcystic component regions (A) (hematoxylin and eosin, 100×). Immunohistochemically, the tumor cells showed the Ki-67 index was 3% (B) and stained positively for S100 (C) Axial, Sagittal, and Coronal T1-weighted gadolinium-enhanced MRI image shows there is no evidence of tumor exists at 12-months follow-up (D-F) (White arrow).

Discussion

Intracranial Schwannoma accounts for 6-8% of all intracranial tumors and arises from the cranial nerves that contain Schwann cells [33]. Using the method of descriptive statistics. Age varied from 14 to 64 years old (mean = 32.4). There were 17 male and 19 female patients. The most common clinical manifestation were headache (n = 22, 61.1%) and convulsion (n = 12, 33.3%). Hyposmia was present in 17 cases (47.2%), absent in 14 cases (38.9%), and unreported in 5 cases (13.9%). Tumor was solid in 19 patients (52.8%), cystic-solid in 11 patients (30.6%) and cystic in 5 patients (13.9%). Radiological appearance was unreported in 1 case (2.8%). Homogeneous enhancement was observed in 14 cases (38.9%), while 21 cases showed heterogeneous enhancement (58.3%), and 1 case was unreported (2.8%). There were 15 cases (41.70%) showed bone erosion and 7 cases (19.40%) showed calcification on computer tomography scan.
TABLE 1. Summary of previously reported cases of olfactory schwannoma

<table>
<thead>
<tr>
<th>Study</th>
<th>Age/Sex</th>
<th>Chief complaint</th>
<th>Olfaction</th>
<th>Calcification</th>
<th>Enhanced</th>
<th>Aspect</th>
<th>Bone erosion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vassilouthis 1980 [4]</td>
<td>17/F</td>
<td>Difficulty in maintaining concentration, forgetfulness, headache, dizziness and amaurosis</td>
<td>Hyposmia</td>
<td>No</td>
<td>Well</td>
<td>Cystic</td>
<td>Yes</td>
</tr>
<tr>
<td>Harada 1992 [5]</td>
<td>33/M</td>
<td>Headache</td>
<td>Hyposmia</td>
<td>No</td>
<td>Hetero</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Sabel 1995 [6]</td>
<td>17/M</td>
<td>Convulsion</td>
<td>UR</td>
<td>No</td>
<td>Well</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Huang 1997 [7]</td>
<td>33/M</td>
<td>Headache, leathargy, decreased eyevision</td>
<td>Normal</td>
<td>UR</td>
<td>Well</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Boyd 1997 [8]</td>
<td>29/F</td>
<td>Headache, convulsion</td>
<td>Hyposmia</td>
<td>No</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Yes</td>
</tr>
<tr>
<td>Timothy 1999 [9]</td>
<td>33/F</td>
<td>Convulsion</td>
<td>Normal</td>
<td>UR</td>
<td>Well</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Praharaj 1999 [10]</td>
<td>45/M</td>
<td>Headache, convulsion</td>
<td>UR</td>
<td>No</td>
<td>Well</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Tsai 2001 [12]</td>
<td>31/F</td>
<td>Headache, convulsion</td>
<td>UR</td>
<td>No</td>
<td>Hetero</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Carron 2002 [13]</td>
<td>59/F</td>
<td>Headache</td>
<td>Normal</td>
<td>No</td>
<td>Well</td>
<td>Solid</td>
<td>Yes</td>
</tr>
<tr>
<td>Amador 2002 [14]</td>
<td>24/F</td>
<td>Hypoesthesia on the left side of face, impaired vision</td>
<td>UR</td>
<td>No</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Yes</td>
</tr>
<tr>
<td>Yuen 2004 [15]</td>
<td>33/F</td>
<td>Convulsion</td>
<td>Normal</td>
<td>UR</td>
<td>Well</td>
<td>Solid</td>
<td>Yes</td>
</tr>
<tr>
<td>Murakami 2004 [16]</td>
<td>30/M</td>
<td>Headache</td>
<td>Normal</td>
<td>No</td>
<td>Well</td>
<td>Solid</td>
<td>Yes</td>
</tr>
<tr>
<td>Shenoy 2004 [17]</td>
<td>55/M</td>
<td>Convulsion</td>
<td>Normal</td>
<td>No</td>
<td>Hetero</td>
<td>Cystic</td>
<td>UR</td>
</tr>
<tr>
<td>Sano 2004 [18]</td>
<td>44/M</td>
<td>Headache</td>
<td>Normal</td>
<td>No</td>
<td>Hetero</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Prasad 2004 [19]</td>
<td>19/M</td>
<td>Convulsion</td>
<td>Anosmia</td>
<td>UR</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
<tr>
<td>Yako 2005 [20]</td>
<td>14/M</td>
<td>Headache, vomiting</td>
<td>Anosmia</td>
<td>Yes</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
<tr>
<td>Adachi 2007 [21]</td>
<td>22/F</td>
<td>Convulsion</td>
<td>Normal</td>
<td>Yes</td>
<td>Hetero</td>
<td>Solid</td>
<td>UR</td>
</tr>
<tr>
<td>Bezircioğlu 2008 [22]</td>
<td>33/F</td>
<td>Headache</td>
<td>Anosmia</td>
<td>UR</td>
<td>Hetero</td>
<td>Solid</td>
<td>Yes</td>
</tr>
<tr>
<td>Daglioglu 2008 [23]</td>
<td>21/M</td>
<td>Headache, aggressive behavior</td>
<td>UR</td>
<td>UR</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Yes</td>
</tr>
<tr>
<td>Kanaan 2008 [24]</td>
<td>14/M</td>
<td>Headache, declining school performance and weight loss</td>
<td>Hyposmia</td>
<td>UR</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
<tr>
<td>Saberi 2008 [25]</td>
<td>35/F</td>
<td>Convulsion, diplopia and headache</td>
<td>Left hyposmia</td>
<td>Yes</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
<tr>
<td>Choi 2009 [26]</td>
<td>39/F</td>
<td>Headache</td>
<td>Anosmia</td>
<td>Yes</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
<tr>
<td>Figueiredo 2009 [27]</td>
<td>49/M</td>
<td>Headache and anosmia</td>
<td>Anosmia</td>
<td>No</td>
<td>Hetero</td>
<td>Cystic</td>
<td>No</td>
</tr>
<tr>
<td>Mirone G 2009 [28]</td>
<td>38/M</td>
<td>Headache, amnesia</td>
<td>Left hyposmia</td>
<td>No</td>
<td>Hetero I</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
<tr>
<td>Li YP 2012 [29]</td>
<td>16/F</td>
<td>Convulsion</td>
<td>Normal</td>
<td>Yes</td>
<td>Hetero</td>
<td>Solid</td>
<td>Yes</td>
</tr>
<tr>
<td>Salunke P 2014 [31]</td>
<td>24/F</td>
<td>Abnormal sensations on the right half of face</td>
<td>Normal</td>
<td>Yes</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
<tr>
<td>Quick J 2015 [2]</td>
<td>64/F</td>
<td>Headache</td>
<td>Left anosmia</td>
<td>UR</td>
<td>Well</td>
<td>Solid</td>
<td>No</td>
</tr>
<tr>
<td>Quick J 2015 [2]</td>
<td>45/F</td>
<td>Hyposmia</td>
<td>Hyposmia</td>
<td>UR</td>
<td>Well</td>
<td>Solid</td>
<td>No</td>
</tr>
<tr>
<td>Nascimento 2015 [32]</td>
<td>39/M</td>
<td>Nasal obstruction, anosmia</td>
<td>Anosmia</td>
<td>No</td>
<td>Hetero</td>
<td>Cystic</td>
<td>No</td>
</tr>
<tr>
<td>Kim DY 2015 [33]</td>
<td>49/F</td>
<td>Headache, nausea, vomiting</td>
<td>Normal</td>
<td>No</td>
<td>Well</td>
<td>Solid</td>
<td>No</td>
</tr>
<tr>
<td>Pereira MC 2016 [34]</td>
<td>25/F</td>
<td>Headache, epistaxis</td>
<td>Normal</td>
<td>No</td>
<td>Well</td>
<td>Solid</td>
<td>Yes</td>
</tr>
<tr>
<td>Manto A 2016 [35]</td>
<td>39/F</td>
<td>Headache, anosmia</td>
<td>Right anosmia</td>
<td>Yes</td>
<td>Well</td>
<td>Solid</td>
<td>Yes</td>
</tr>
<tr>
<td>Current case 2016</td>
<td>21/M</td>
<td>Headache, blurred vision</td>
<td>Normal</td>
<td>No</td>
<td>Hetero</td>
<td>Cystic</td>
<td>Solid</td>
</tr>
</tbody>
</table>

UR = unreported.

Olfactory schwannomas [28, 31, 32, 36]. The differential diagnosis of tumors involving in olfactory groove include meningiomas, olfactory esthesioneuroblastoma, adenocystic carcinoma, metastatic disease and olfactory ensheathing cells [2, 28, 37]. Olfactory groove schwannoma can have similar neuroradiological features as olfactory groove meningiomas, including extraxial location, calcification, contrast enhancement and perifocal edema [28]. However, young age at presentation (mean = 32.4), the presence of cystic component (in 16 of 36 cases), the absence of dural tail sign and low vascularity may help us to make differential diagnosis between schwannoma and meningioma before surgery [29]. In addition, most meningiomas are iso-to hypointense on T2WI, whereas schwannoma tend to be more hyperintense [34]. Moreover, schwannoma may often be isointense to gray matter [23].

Esthesioneuroblastoma and ethmoid carcinomas should be carefully differentiated especially for younger patients because those lesi-
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ons require radical craniofacial resection [34]. Bone erosion over the cribriform plate is more commonly noted in schwannoma, while lesions like esthesioneuroblastoma and ethmoid carcinoma could invade paranasal sinuses and cause marked bony destruction [23]. More importantly, there is a substantial risk of misdiagnosing an esthesioneuroblastoma from schwannoma on frozen sections [20]. This will complicate the decision making at the operation, so as the following treatment plan. Therefore, immunohistochemistry is essential for the differential diagnosis between schwannoma and esthesioneuroblastoma. The present case was suspected as having got the esthesioneuroblastoma before surgery, and was finally diagnosed as schwannoma. No special treatment was given after surgery and he remained no clinical evidence of recurrence. Other malignant tumors such as lymphoma and hemangiopericytoma are unusual entities which should also be included in the differential diagnosis [34, 35].

Recently, some authors argue that some of the published cases, were not schwannoma but olfactory ensheathing cell tumors (OECs) instead [2, 35]. OECs derive from olfactory placodes whereas schwann cells originate from the neural crests. These tumors are of very similar appearance in clinical, imaging and histological characteristics [2, 35, 37]. The only way to distinguish these two tumors rely on the immunohistochemical staining, for schwann cells show positive staining for Leu 7 (CD 57) while OECs show negative staining for Leu 7 (CD 57) [37]. Therefore, real olfactory schwannoma must be even rarer, because in some cases Leu7 or CD 57 examinations had not been performed.

Operation seems the only method for the treatment of olfactory schwannoma. Due to the benign nature of this tumor, complete resection of the tumor is the best treatment modality, and adjunctive therapy is unnecessary. The classic surgical approach to the anterior cranial fossa is the bifrontal craniotomy; however, Pereira MC used a unilateral extended endonasal endoscopic approach to remove the tumor completely [34]. This new approach has the advantage of fewer operative complications because of its “minimal access” nature, however, better skull base reconstruction should be performed to prevent postoperative cerebrospinal fluid fistula [34, 38]. In conclusion, olfactory schwannoma is a rare tumor, which is often underreported or misdiagnosed. Its origin is still largely uncertain and need further investigations. These cases should alert the clinicians to keep in mind that olfactory schwannoma is in the differential diagnosis of extra-axial lesions of the anterior cranial fossa, especially in young patients.

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Disclosure of conflict of interest

None.

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