Oncology and Translational Medicine
DOI 10.1007/s10330-018-0316-6

December 2018, Vol. 4, No. 6, P259–P262

CASE REPORT

Olfactory schwannoma: a case report

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Abstract

Objective Intracranial schwannomas are frequently located in the VIII cranial nerve, followed by the V and VII cranial nerves, but are quite rare in the olfactory cranial nerves. Here, we report a case of an olfactory schwannoma and review the literature to improve the diagnosis and treatment of olfactory schwannoma.

Methods We report a case of olfactory schwannoma in a 51-year-old Chinese man who experienced dizziness and no other symptoms.

Results Magnetic resonance imaging (MRI) showed a neoplastic mass located on the anterior cranial base to the right of the midline and near the cribriform plate and sphenoidal plane. The lesion travelled through the cribriform plate into the nasal cavity. This mass was initially thought to be an olfactory groove meningioma. We performed a craniotomy for surgical excision, and the tumor was completely resected, and the skull base was reconstructed at the same time. There were no complications during surgery, and the patient recovered well. The histopathological diagnosis was a schwannoma.

Conclusion Olfactory schwannomas are extremely rare and similar to olfactory ensheating cell tumors, and the immunohistochemical staining of leukocyte antigen 7 (Leu7/CD57) can be used to identify them. Although the standard treatment of olfactory schwannoma remains unclear, in all reports, most patients can have excellent prognosis after an effective surgical resection.

Received: 9 November 2018
Revised: 10 December 2018
Accepted: 21 December 2018

Key words: olfactory schwannoma; anterior skull base schwannoma; schwannoma; diagnosis; treatment

Intracranial schwannomas are frequently located in the VIII cranial nerve, followed by the V and VII cranial nerves [1–2]. Olfactory schwannoma is extremely rare. Schwannomas originate from Schwann cells and thus can grow anywhere in the nervous system that contain Schwan cells. Olfactory and optic nerves lack a Schwann cell layer, making it impossible for schwannomas to develop here [2–3]. The most common tumor developed within the anterior cranial fossa is a meningioma, with olfactory schwannoma and olfactory ensheathing cell tumor (OECT) being the rarest. Olfactory schwannoma is similar to OECT with respect to clinical and radiological characteristics, and the only way to distinguish between them is immunohistochemical staining [4].

Here, we report about a patient with an olfactory schwannoma who underwent craniotomy for surgical excision to improve its diagnosis and treatment.

Case report

A 51-year-old man with vertigo, without headache, nausea, emesis, anosmia, and seizures was referred to our hospital. Physical examination revealed a body temperature of 36.4 °C, pulse rate of 83 beats/min, and blood pressure of 122/80 mmHg. Brain MRI revealed a homogeneous, well-defined, extra-axial neoplastic lesion measuring 4.1 cm × 3.6 cm × 3.1 cm and located on the anterior cranial base to the right of the midline near the cribriform plate and sphenoidal plane, with the ipsilateral rectus gyrus compressed. The lesion extended from the cribriform plate into the nasal cavity, was hypointense on T1-weighted and hyperintense on T2-weighted images, and was heterogeneously reinforced after the application of a contrast (Fig. 1). Computed tomography (CT) scans showed a thin anterior skull base and a partly calcified

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neoplastic mass (Fig. 2). This mass was initially considered to be an olfactory groove meningioma.

The right frontopterional craniotomy approach for surgical resection was performed successfully on September 6, 2018. With an operating microscope and microsurgical technique, dissection was performed, and the tumor was completely resected and the skull base was reconstructed at the same time. There were no surgical complications, and the patient recovered well without postoperative complications such as cerebrospinal fluid leakage. Microscopically, the mass consisted of spindle-shaped cells that were partly arranged in palisades. Histological and immunohistochemical (IHC) staining was positive for S-100 protein and Leu 7 but negative for EMA and GFAP (Fig. 3). Based on histological and IHC analyses, a diagnosis of schwannoma was made.

Discussion

The most common subfrontal neoplasm is a meningioma, whereas subfrontal olfactory schwannoma is uncommon [5]. Figueiredo [6] reported an average age of diagnosis of 30.9 years among mainly male patients who have main clinical symptoms such as headache, seizures, and anosmia.

Intracranial schwannomas are frequently located in the VIII cranial nerve, followed by the V and VII cranial nerves. Optic and olfactory cranial nerves lack a Schwann cell layer; thus, the development of schwannomas at this location is impossible [2–3]. They have been labelled as subfrontal schwannoma, anterior skull base schwannoma, olfactory groove Schwannoma, olfactory schwannoma, etc. [3].

The origin of pathogenesis remains unclear. Various theories have been proposed; the developmental hypothesis suggests that pial cells transform into Schwann cells, aberrant neural crest cells in the central nervous system, and ectopic Schwann cells, and the non-development theories maintain that olfactory schwannomas originate from Schwann cells located in the perivascular nerve plexus, meningeal branches of the trigeminal nerve, anterior ethmoidal nerve, or dural nerves of the anterior cranial fossa and subarachnoid space [2–3, 6–8, 10]. Other theories include the existence of

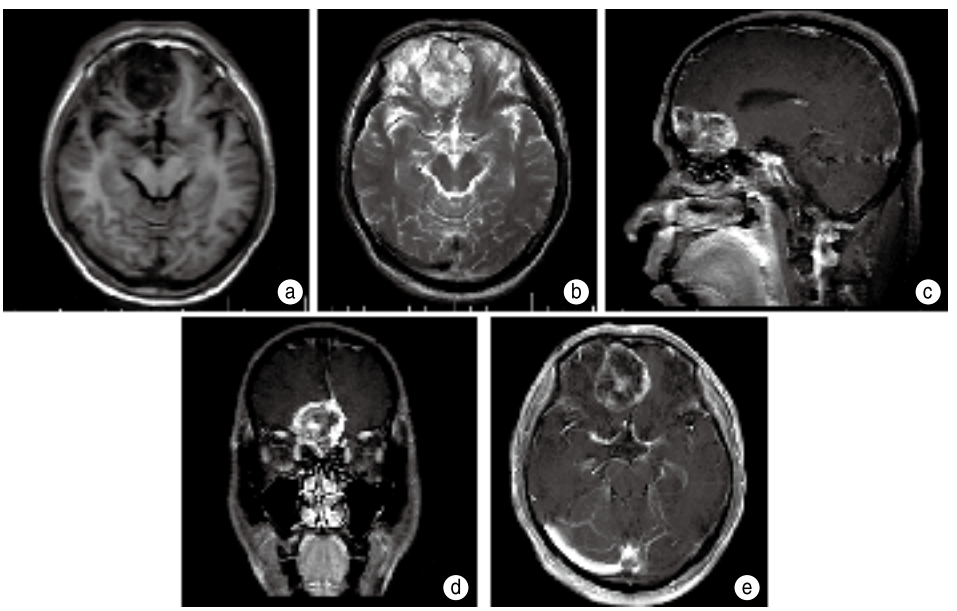

Fig. 1 MRI shows an extra-axial lesion of neoplastic, 4.1 cm x 3.6 cm x 3.1 cm, homogeneous, well defined and located on the anterior cranial base to the right of midline and throuth the cribriform plate into the nasal cavity. (a) T1 weighted is hypointense; (b) T2 weighted is hyperintense; (c–e) After application of contrast there was heterogeneous reinforcement, the margin of tumor are strongly reinforcement

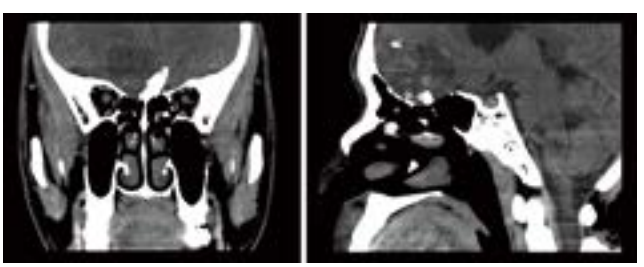

Fig. 2 The computed tomography scan reveal the anterior skull base became thin and the part of neoplastic has calcification

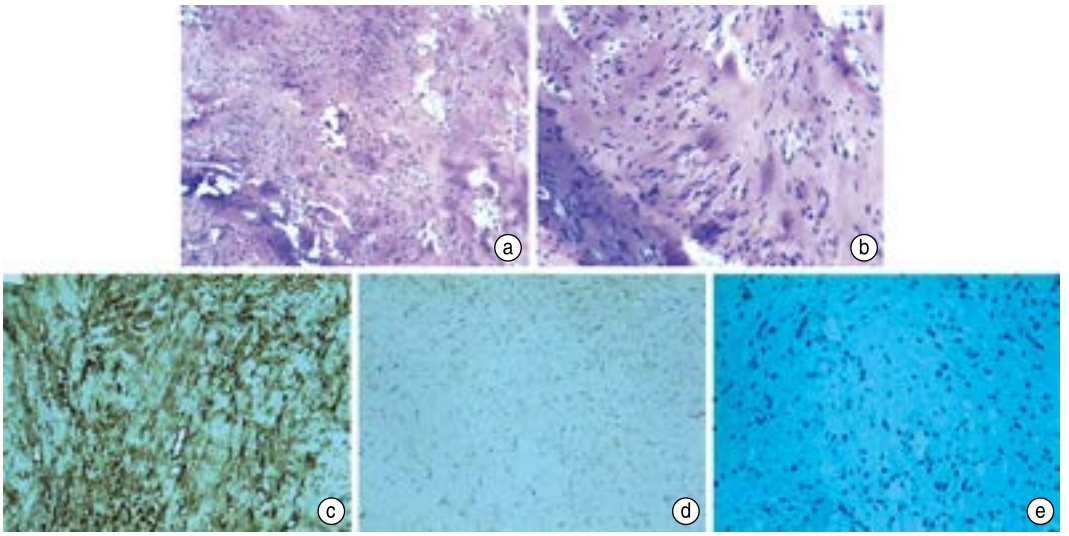

Fig. 3 Photomicrograph show the mass consisted by spindle-shaped cells and sparsely arranged, part of them arranged in palisades (HE staining, a × 100; b × 200); IHC analysis: The neoplastic cells were positive for S-100 protein (c) and Leu 7 (d) and negative for EMA (e) (× 200)

an embryological remnant terminal nerve and reactive changes as a result of cerebral infarctions or multiple sclerosis [3].

Yasuda [9] reported a case of OECT in 2006 and revealed that OECTs are located in the nerve fiber layer of the olfactory bulb and can promote axonal growth along the primary olfactory pathway of normal adult animals. Similar to Schwann cells, only immunohistochemical staining can identify them. Both Schwann cells and OECTs test positive for S-100 protein and negative for EMA, enabling their differentiation from meningiomas. However, Schwann cells are positive for Leu 7 staining, while OECs are negative [4, 9].

It is extremely difficult to diagnose olfactory schwannoma before surgery due to its uncommonness; most patients are regarded as having olfactory groove meningioma before operation [5]. Histological and IHC analyses, including positive staining for S-100, are vital for pathological diagnosis [4, 9]. Leu7 markers can be used to differentiate OECT from olfactory schwannoma [3–4, 8–9]. Therefore, to improve the diagnostic accuracy, additional IHC staining of Leu7 should be performed.

Our patient who presented vertigo, without headache, anosmia, and seizures was diagnosed based on histological and IHC staining. Pathology of the biopsy specimen showed schwannomas, with the appearance of spindle-shaped tumor cells sparsely and partly arranged in palisades. IHC analysis revealed tumor cells that were positive for S-100 and negative for EMA, similar to OECT as previously published [4]. IHC staining of Leu7(+) suggested an olfactory schwannoma.

The standard treatment of olfactory schwannoma remains unclear due to the disease rarity. As previous literature reported, most cases were treated via a bifrontal craniotomy for surgical excision [1–2, 3, 7–8, 10–11], while some cases underwent an endoscopy-aided or endoscopy resection [5, 12–14]. In our case, we performed a right frontopterional craniotomy approach for surgical resection and obtained a satisfactory surgical result. We reviewed literature where most patients with gross-total resection without postoperative radiotherapy during the subsequent MRI showed no evidence of tumor recurrence [1–2, 11]. However the patient had subtotal resection (STR) whether should have adjuvant radiotherapy still unknown, Kim [8] reported that patients with STR received gamma knife radiosurgery, and within 5 years following radiosurgery, the nasal cavity mass had not grown.

In conclusion, olfactory schwannomas are extremely rare in the anterior skull base; only 67 cases of the olfactory schwannoma have been reported [10]. Although schwannomas are regarded as benign tumors, the prognosis of olfactory schwannoma still needs further studies. Here, we report a patient with olfactory schwannoma to improve its diagnosis and treatment.

Acknowledgments

The authors thank all the patients and their families for participating in this research.

Conflicts of interest

The authors indicated no potential conflicts of interest.

References

1. Zheng K, Jiang S, Xu JG, *et al*. Giant cranionasal schwannoma. J Clin Neurosci, 2010, 17: 520–522.
2. Kim DY, Yoon PH, Kie JH, *et al*. The olfactory groove schwannoma attached to the cribriform plate: a case report. Brain Tumor Res Treat, 2015, 3: 56–59.
3. Figueroa-Ángel V, Rodríguez-Aceves CA, Calderon-Miranda WG, *et al*. Subfrontal schwannoma: case report and review of literature. World Neurosurg, 2018, 111: 386–390.
4. Darie I, Riffaud L, Saïkali S, *et al*. Olfactory ensheathing cell tumour: case report and literature review. J Neurooncol, 2010, 100: 285–289.
5. Kanaan HA, Gardner PA, Yeaney G, *et al*. Expanded endoscopic endonasal resection of an olfactory schwannoma. J Neurosurg Pediatr, 2008, 2: 261–265.
6. Figueiredo EG, Soga Y, Amorim RL, *et al*. The puzzling olfactory groove schwannoma: a systematic review. Skull Base, 2011, 21: 31–36.
7. Yuen A, Trost N, McKelvie P, *et al*. Subfrontal schwannoma: a case report and literature review. J Clin Neurosci, 2004, 11: 663–665.
8. Kim SH, Lee JH, Sung SK, *et al*. Subfrontal schwannoma extended broadly to nasal cavity treated by gamma knife Radiosurgery Following Surgical Excision: a case report. Brain Tumor Res Treat, 2017, 5: 116–119.
9. Yasuda M, Higuchi O, Takano S, *et al*. Olfactory ensheathing cell tumor: a case report. J Neurooncol, 2006, 76: 111–113.
10. Taha MM, AlBakry A, ElSheikh M, *et al*. Olfactory nerve schwannoma: a case report and review of the literature. Surg J (N Y), 2018, 4: 164–166.
11. Mirone G, Natale M, Scuotto A, *et al*. Solitary olfactory groove schwannoma. J Clin Neurosci, 2009, 16: 454–456.
12. Blake DM, Husain Q, Kanumuri VV, *et al*. Endoscopic endonasal resection of sinonasal and anterior skull base schwannomas. J Clin Neurosci, 2014, 21: 1419–1423.
13. Liu JK, Eloy JA. Expanded endoscopic endonasal transcribriform approach for resection of anterior skull base olfactory schwannoma. Neurosurg Focus, 2012, 32 Suppl 1: E3.
14. Pereira MC, Peris-Celda M, Qian J, *et al*. Unilateral endoscopic endonasal surgery for resection of an olfactory schwannoma of the anterior cranial base in a patient with hereditary hemorrhagic telangiectasia. World Neurosurg, 2016, 89: 729.e15–729.e20.

DOI 10.1007/s10330-018-0316-6
Cite this article as: Wu WD, Zhan XZ, Wang KZ, *et al*. Olfactory schwannoma: a case report. Oncol Transl Med, 2018, 4: 259–262.