Olfactory Groove Schwannoma: A Rare Intracranial Tumor - Case Studies and Review

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Test

Olfactory Groove Schwannoma Test

1. What is the primary location of Olfactory Groove Schwannomas?
    1. [ ] a) Temporal lobe
    2. [x] b) Frontal lobe
    3. [ ] c) Occipital lobe
    4. [ ] d) Parietal lobe

1. Which of the following is NOT a common symptom of Olfactory Groove Schwannoma?
    1. [ ] a) Anosmia
    2. [ ] b) Visual disturbances
    3. [x] c) Auditory hallucinations
    4. [ ] d) Headaches

1. What is the primary treatment option for Olfactory Groove Schwannomas?
    1. [ ] a) Radiation therapy
    2. [ ] b) Chemotherapy
    3. [x] c) Surgical resection
    4. [ ] d) Medication

1. Which of the following statements is true regarding Olfactory Groove Schwannomas?
    1. [x] a) They often originate from Schwann cells in the olfactory groove.
    2. [ ] b) They primarily affect the optic nerve.
    3. [ ] c) They are associated with frequent auditory deficits.
    4. [ ] d) They cannot be treated surgically.

1. Olfactory Groove Schwannomas can sometimes be confused with which other tumors?
    1. [ ] a) Glioblastomas
    2. [x] b) Olfactory groove meningiomas
    3. [ ] c) Medulloblastomas
    4. [ ] d) Astrocytomas

1. What is the typical outcome after complete surgical resection of an Olfactory Groove Schwannoma?
    1. [ ] a) Poor prognosis
    2. [ ] b) Neurological deficits
    3. [x] c) Favorable prognosis
    4. [ ] d) Olfactory dysfunction

1. What is the primary goal of surgical resection in treating Olfactory Groove Schwannomas?

    1. [ ] a) Complete removal of the tumor with no concern for neurological function
    2. [x] b) Partial removal of the tumor while preserving neurological function
    3. [ ] c) Eliminating olfactory function
    4. [ ] d) None of the above

1. According to the provided information, what is the controversial aspect of Olfactory Groove Schwannoma origin?
    1. [ ] a) They always originate from the olfactory nerve.
    2. [x] b) Their origin remains unknown.
    3. [ ] c) They originate from the optic nerve.
    4. [ ] d) They develop due to hormonal imbalances.

1. In which cranial fossa are Olfactory Groove Schwannomas predominantly located?
    1. [ ] a) Middle cranial fossa
    2. [ ] b) Posterior cranial fossa
    3. [x] c) Anterior cranial fossa
    4. [ ] d) Inferior cranial fossa

1. Which specific cells are responsible for the formation of Schwannomas?
    1. [ ] a) Oligodendrocytes
    2. [x] b) Schwann cells
    3. [ ] c) Neurons
    4. [ ] d) Astrocytes

Abstract

Olfactory groove schwannoma is an uncommon intracranial tumor originating in the olfactory groove and is primarily associated with benign schwann cells. This review aims to provide insights into the clinical characteristics, diagnosis, treatment, and prognosis of this rare entity. Olfactory groove schwannomas are predominantly located in the anterior cranial fossa, often resulting in symptoms such as headaches, anosmia, visual disturbances, personality changes, and cognitive deficits. Diagnosis typically involves neuroimaging studies like MRI and CT scans. Surgical resection is the primary treatment option, with the goal of preserving neurological function, and sometimes complemented with radiation therapy for inoperable cases. While olfactory groove schwannomas are generally benign, each case presents unique challenges, and a multidisciplinary approach is essential for comprehensive patient care. Additionally, there is an ongoing debate about their origin, with both developmental and non-developmental hypotheses being proposed. Differential diagnosis should consider other tumors, such as olfactory groove meningiomas and esthesioneuroblastomas. This review also discusses case reports to shed light on the complex nature of olfactory groove schwannomas. Understanding the diverse aspects of this rare tumor is crucial for accurate diagnosis, management, and patient outcomes.

Keywords: Olfactory groove schwannoma, anterior cranial fossa, diagnosis, surgical resection, prognosis, differential diagnosis, case reports.

Introduction

Olfactory groove schwannoma is a rare type of tumor that arises in the olfactory groove of the skull. Schwannomas are typically benign tumors that originate from Schwann cells, which are responsible for the formation of the myelin sheath covering nerves. When a schwannoma develops in the olfactory groove, it usually arises from the olfactory nerve or its branches.

Here are some key points about olfactory groove schwannomas:

Location: Olfactory groove schwannomas are found in the anterior cranial fossa, where the olfactory bulb and tract are located. They can grow and compress nearby structures, including the frontal lobes of the brain.

Symptoms: The symptoms of an olfactory groove schwannoma can vary depending on its size and location. Common symptoms may include headaches, changes in smell (anosmia), visual disturbances, personality changes, and cognitive deficits.

Diagnosis: Diagnosis often involves neuroimaging studies, such as magnetic resonance imaging (MRI) or computed tomography (CT) scans. These tests can help visualize the tumor and its impact on adjacent structures.

Treatment: The primary treatment for olfactory groove schwannomas is surgical resection. The goal of surgery is to remove the tumor while preserving neurological function. In some cases, a combination of microsurgery and endoscopy may be used to access and remove the tumor. Radiation therapy may be considered for residual or inoperable tumors.

Prognosis: Olfactory groove schwannomas are typically slow-growing and benign, which generally results in a favorable prognosis after surgical removal. The long-term outcome depends on factors such as the extent of tumor resection and the patient's overall health.

Differential Diagnosis: Olfactory groove schwannomas may be mistaken for other tumors, such as olfactory groove meningiomas, which are more common in this region. A precise diagnosis is essential for determining the most appropriate treatment plan.

Multidisciplinary Approach: Managing olfactory groove schwannomas often requires a multidisciplinary approach involving neurosurgeons, otolaryngologists, and neurologists to provide comprehensive care.

It's important to note that while olfactory groove schwannomas are generally benign, each case is unique, and treatment decisions should be made in consultation with a medical team specializing in neurosurgery and neuro-oncology.

According to past reports, subfrontal schwannomas are occasionally described as olfactory schwannomas or olfactory groove schwannomas.

Epidemiology

Schwannoma arising from the olfactory system, often called olfactory groove schwannoma (OGS), is rare, as the olfactory bulb and tract, belonging to the central nervous system, should lack Schwann cells. Another rare entity called olfactory ensheathing cell tumor (OECT) has been reported, which

mimics clinical and radiological characteristics of OGS.

They are very rare tumors, leaving the issue of their origin controversial.

In 94 patients with anterior skull base (ASB) and sinonasal schwannomas, 44 (46.8%) were exclusively sinonasal, 30 cases (31.9%) were exclusively intracranial, 12 (12.8%) were primarily intracranial with extension into the paranasal sinuses, and 8 (8.5%) were primarily sinonasal with intracranial extension [1].

Li et al. gathered previous literatures and reported that results in 35 cases of olfactory schwannomas (between 1974 and 2010) has shown that 14 out of 30 cases (47%) (with the exclusion of five cases due to unknown olfactory function) had preserved olfactory function, but that the remaining 16 (53%) experienced either anosmia or hyposmia. Regarding the attachment sites of the schwannomas, they summarized that 12 cases were on the cribriform plate, 10 cases were on the olfactory groove, and 5 cases were on the skull base and skull base dura. When the tumor was attached to the cribriform plate, the rate of olfaction preservation was relatively high [9 of 11 cases (82%), excluding one case due to unknown olfactory function], compared to olfactory groove attachment [2 of 7 cases (29%), excluding three cases due to unknown olfactory function].

Figueiredo et al. systematically reviewed the literature concerning the anterior cranial fossa schwannomas to understand their pathogenesis, determine their origin, and standardize the terminology. They performed a MEDLINE, EMBASE, and Science Citation Index Expanded search of the literature; age, gender, clinical presentation, presence or absence of hyposmia, radiological features, and apparent origin were analyzed and tabulated. Cases in a context of neurofibromatosis and nasal schwannomas with intracranial extension were not included. Age varied between 14 and 63 years (mean = 30.9). There were 22 male and 11 female patients. The clinical presentation included seizures (n = 15), headache (n = 16), visual deficits (n = 7), cognitive disturbances (n = 3), and rhinorrhea (n = 1). Hyposmia was present in 14 cases, absent in 13 cases (39.3%), and unreported in five. Homogeneous and heterogeneous contrast enhancement was observed in 14 and 15 cases, respectively. The region of the olfactory groove was the probable site in 96.5%. Olfactory tract could be identified in 39.3%. The most probable origin is the meningeal branches of trigeminal nerve or anterior ethmoidal nerves. Thus, olfactory groove schwannoma would better describe its origin and pathogenesis and should be the term preferentially used to name it [2].

Etiology

Because the olfactory and optic nerves lack a Schwann cell layer, these are not prone to develop into a schwannoma.

Some hypotheses about the genesis of olfactory groove schwannoma are centered on its developmental and non-developmental origins.

The developmental hypotheses suggest whether mesenchymal pial cells to transform into ectodermal Schwann cells or neural crest cells to migrate within the substance of the central nervous system .

The non-developmental hypotheses postulate that intracranial schwannomas arise from the Schwann cells normally presenting in the adjacent structures, such as the perivascular nerve plexus, the meningeal branches of the trigeminal and anterior ethmoidal nerves innervating the anterior cranial

fossa and olfactory groove [3) 4)].

Yasuda et al. [5)] proposed the concept of an olfactory ensheathing cell (OEC) tumor in 2006. Olfactory ensheathing cells are glial cells that ensheath the axons of the first cranial nerve. Microscopically, both olfactory ensheathing cells and Schwann cells have similar morphological and immunohistochemical features. However, immunohistochemically olfactory ensheathing cells are negative for Leu7 and Schwann cells positive [6)].

Differential diagnosis

Often, these tumors can be confused for other entities, especially olfactory groove meningiomas and esthesioneuroblastomas [7)].

Treatment

Because most olfactory region schwannomas have a benign nature, a complete resection of the tumor is the treatment of choice, and adjunctive therapy is not usually required [8) 9)].

With the recent advances in endoscopic skull base surgery, various anterior skull base tumors (ASB) can be resected successfully using an expanded endoscopic endonasal transcribriform approach through a “keyhole craniectomy” in the ventral skull base. This approach represents the most direct route to the anterior cranial base without any brain retraction. Tumor involving the paranasal sinuses, medial orbits, and cribriform plate can be readily resected. In a video atlas report, Liu and Eloy demonstrate their step-by-step techniques for resection of an ASB olfactory schwannoma using a purely endoscopic endonasal transcribriform approach. They describe and illustrate the operative nuances and surgical pearls to safely and efficiently perform the approach, tumor resection, and multilayered reconstruction of the cranial base defect. The video can be found here: http://youtu.be/NLtOGfKWC6U [10)].

Endoscopic Endonasal Anterior Cranial Fossa Approach [11)]

Prognosis

The prognosis after complete resection is known to be favorable [12)].

When the tumor is attached to the cribriform plate, the preservation rate of olfactory function is higher compared to nearby structures [13)].

Case reports

A 65-year-old male patient who presented with olfactory groove meningioma and non-functioning pituitary adenoma as a collision tumor. The patient was admitted with a headache and right-sided vision loss. The patient's first neurologic examination was consistent with temporal anopsia in the right eye. Subsequent contrast-enhanced cranial MRI revealed a 65x55x40 mm heterogeneously contrast-enhanced lesion in the anterior skull base extending from the sellar region to the corpus

callosum. Because of the tumor size, a two-staged operation was planned. First, the tumor was partially excised via a right frontal craniotomy with a transcranial approach, and the tumor in the sellar region was left as a residue. The pathology reports after the first surgery showed pituitary adenoma and meningeal epithelial type meningioma (WHO Grade I). The residual tumor tissue was resected seven months later via an endoscopic endonasal approach, except for the part that invaded the right anterior cerebral artery. The optic nerve was decompressed. The patient was then referred to the radiation oncology clinic for radiosurgery. Collision tumors should be considered in the differential diagnosis in preoperative evaluation and surgical planning when heterogeneously contrast-enhanced areas significantly localized adjacent to each other are seen on cranial MRI. On the other hand, when the surgeon encounters sudden changes in the appearance or consistency of the tumor during the surgery, they should suspect these tumor complexes. The diagnosis of collision tumors is quite challenging but is of great importance regarding the patient's need for postoperative radiation therapy or the recurrence characteristics of tumors. However, more studies are needed on these complexes' etiology, surgical planning, and postoperative management [14].

A 59-year-old woman who presented with a paroxysmal headache for 1 year. The tumor appeared as hypointensity on T1-weighted images, hyperintensity on T2-weighted, and exhibited strong, heterogeneous enhancement. The tumor was removed through a lateral supraorbital approach. The final pathologic diagnosis was schwannoma. The postoperative period was uneventful after 4 months, and the headache disappeared [15].

2016

Bohoun et al report two rare cases of schwannoma-like tumor in the anterior cranial fossa that showed negative staining for Leu7, but positive staining for Schwann/2E, and discuss their origin. Two cases of mass lesions in the anterior cranial fossa in a 26-year-old man and a 24-year-old woman were successfully removed. Morphological examination of these tumors was compatible with a diagnosis of schwannoma. Immunohistochemically, both cases were negative for Leu7, yielding a diagnosis of olfactory ensheathing cell tumor (OECT), but were positive for the schwannoma-specific marker, Schwann/2E. Immunohistochemical staining results in this two cases question the current assumption that OGS and OECT can be distinguished only by Leu7 staining pattern. In conclusion, the origins of OGS and OECT remain to be determined, and further studies in larger numbers of cases are needed to characterize these rare tumors in the anterior cranial fossa [16]

2015

A case of a 49-year-old woman with an olfactory groove schwannoma attached to the cribriform plate without olfactory dysfunction. She had no specific neurological symptoms other than a headache, and resection of the tumor showed it to be a schwannoma. About 19 months after the operation, a follow-up MRI showed no evidence of tumor recurrence. Surgical resection through subfrontal approach could be one of the curative modality in managing an olfactory groove schwannoma. An olfactory groove schwannoma should be considered in the differential diagnosis of anterior skull base tumors [17].

2014

Okamoto et al. report two cases of subfrontal schwannomas treated with surgical resection. In one case, the tumor was located between the endosteal and meningeal layers of the dura mater. This rare case suggests that subfrontal schwannomas may originate from the fila olfactoria [18].

A 24 year old lady presented with hemifacial paraesthesias. Radiology revealed a large olfactory region enhancing lesion. She was operated through a transbasal approach with olfactory preservation. [19].

One patient had intradural intracranial extension and required an extended endoscopic endonasal transcribriform approach with anterior skull base resection [20].

2013

A 66-year-old woman presented with a 1-year history of progressive headaches. Clinical examination revealed hypoesthesia of the nasal tip. CT-scan and MRI studies revealed a large subfrontal tumor thought preoperatively to be a meningioma. Intraoperatively, a large extra-axial tumor arising from the floor of the right frontal fossa was encountered. Histopathology identified the tumor as a schwannoma. This current case gives strong clinical presumption of an origin from the anterior ethmoidal nerve. We reviewed the literature in order to establish the epidemiology of these tumors, from which there appear to be divergent profiles depending on tumor origin and histology. Despite close similarities with olfactory groove meningiomas, patient history and radiological findings provide substantial evidence for differential diagnosis [21].

2012

Liu and Eloy demonstrate their step-by-step techniques for resection of an ASB olfactory schwannoma using a purely endoscopic endonasal transcribriform approach.

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 tumor was attached to the cribriform plate, and achieved gross total resection without compromising her olfactory function [22].

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Conclusions

In conclusion, understanding the diverse aspects of olfactory groove schwannomas is crucial for accurate diagnosis, effective management, and improved patient outcomes. This rare tumor entity serves as a reminder of the complexity of intracranial pathologies and the need for ongoing research to better comprehend its origin and pathogenesis.

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