Benign Schwannoma in the Sphenoid Sinus: two cases report

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Schwannomas are common benign neoplasms which arise from the peripheral nerve sheath and embryonically from the neuroectoderm. About 25-45% of all schwannomas arise in the head and neck region and of these only less than 4% was located at sinonasal cavity in the reported literature. Radiologists may encounter preoperative diagnostic difficulties of schwannoma involving paranasal cavities due to their rarity, varied clinical symptoms and imaging features. For example, a sphenoid sinus schwannoma sometimes could be confused with other more common skull base tumor such as clival chordoma, chondrosarcoma and plasmacytoma. The differential diagnosis should be carefully laid out since they have different prognosis and treatment plan to be executed. Thus, we report two cases of sphenoid sinus schwannomas with discussion of related skull base tumors and reviewed literatures.

CASES REPORT

Case 1

A 49-year-old woman presented with a history of headache, blurred vision and nasal obstruction for two months. She visited the ENT clinic, where a nasal endoscope exam showed nasal septum deviation to the left side and a protruding boggy mass from the right posterior nasal wall which was visibly hyperemic. Neurological examination and laboratory tests were not contributory. With suspicion of skull base tumor, a MRI examination was ordered and revealed an expansile heterogeneous tumor at the basisphenoid (Fig. 1). Under the impression of skull base tumor, transsphenoid approach for tumor resection was performed and uncovered a 5-cm well-marginated, encapsulated solid tumor, with the tumor mass located mainly in the sphenoid sinus and with rarefaction of the sinus roof. The pathology confirmed the diagnosis of benign schwannoma (Fig. 2). The patient has been disease-free in the eight-month follow up interval after the surgery.
Case 2
A 52-year-old male presented with a history of headache and diplopia for six months. On physical examination, there was no other neurologic deficit and the pertinent laboratory data were also insignificant. A MRI study was performed which showed a large inhomogeneous mass in the right sphenoid sinus (Fig. 3) with appearance similar to that in case 1. Partial resection of the tumor mass was executed by the neurosurgeon via transsphenoid approach. A well-defined encapsulated solid tumor with maximum diameter of 4 cm was found at the sphenoid sinus and juxtasellar-cavernous region. The tumor was yellowish, with soft-elastic texture, and evidently had eroded the floor of sphenoid sinus. Most of the tumor was removed except for the most lateral part that attached to the cavernous sinus. Frozen section showed a spindle cell tumor, and the immunohistochemistry stain was positive for S100 which confirmed the diagnosis of schwannoma. The patient has undergone radiation therapy for the paracavernous remnant tumor component.

DISCUSSION
Schwannoma is a common benign tumor in the head and neck region and mostly found at the cerebellopontine angle, where acoustic schwannoma familiarly located, followed by paracavernous region where trigeminal nerve transverses. Schwannoma have been discovered infrequently in the sinonasal cavity with the reported incidence of less than 4% of the head and neck schwannoma [1, 2]. Schwannoma has prevalence of location, in decreasing order, in the maxillary sinus, ethmoid sinus, sphenoid sinus and lastly frontal sinus when found in the sinonasal region [3]. It is believed that when occurring in the particular region, schwannoma probably arises from the peripheral maxillary and ophthalmic branches of trigeminal nerve or ganglia and plexus of autonomic nerves [4, 5]. It is plausible to suggest that the close proximity with the pathway of cranial nerves and their division is a major attributor behind the statistics. For example, because the olfactory nerve is lack of Schwann cells, therefore it’s least likely to find schwannoma in the frontal sinus.

There may be different clinical and surgical implications as well as diagnostic difficulties if a schwannoma is situated in the anterior paranasal sinuses (frontal, ethmoid and maxillary sinuses) as opposed to the posterior paranasal sinus (sphenoid sinus). The signs and symptoms of anteriorly located sinonasal schwannoma vary, commonly presenting with nasal obstruction. Other complaints include epistaxis, anosmia and rhinorrhea. On the other hand, sphenoid schwannoma may present with cranial nerve palsy as exemplified by diplopia in previously reported cases and also shown in both of the presented cases.

When these tumors are discovered in the anterior paranasal sinuses, it may pose diagnostic challenges among the other more common benign
epithelial lesions such as polyps, papilloma, angioma, fibrous-osseous tumors or malignant counterparts of squamous cell carcinoma, adenocarcinoma, lymphoma and esthesioblastoma [6]. Imaging features are often nonspecific but can give some preoperative diagnostic clues. For instance, schwannomas usually enhance variably and often demonstrate bone remodeling rather than cortex destruction on computed tomography. The MRI often reveals intermediate T1WI and variable T2WI signal intensity masses. When the schwannoma is found in the less commonly located sphenoid sinus, as in both of these cases, tumor mass may occupy the entire sphenoid sinus cavity and extending to the basisphenoid portion, it causes further diagnostic dilemma with other skull base malignancies such as chordoma, chondrosarcoma and plasmacytoma. Clival chordoma frequently presents as a midline basisphenoid mass, but it is associated with cortex rarefaction, destruction of the sphenoid or with mass protruding into the prepontine cistern. Chondrosarcoma is usually situated more eccentrically and contains matrix of calcifications. Both chordoma and chondrosarcoma may enhance variably and can be confused with sphenoid schwannoma, especially when the tumor encroaches on the boundary of the sinus cavity.

There have been only sporadic cases describing schwannoma involving primarily in the sphenoid sinus in the literature [7-9]. Both of the cases presented herein demonstrate the rarity of sphenoid sinus schwannoma. The sphenoid sinus being the latest sinus to be pneumatized and because of its location near the skull base, symptoms are usually not presented until considerable tumor bulk has filled the entire sinus cavity. If we scrutinize the extent of tumor on coronal MR images, we

Figure 2. Hisopathologic slide shows typical pictures of schwannoma with uniformly shaped spindle cells with variable hypercellularity, focal nuclear palisading and hyalinized blood vessels.

Figure 3. Non-enhanced coronal T2WI a. and gadolinium-enhanced sagittal a. MR imagings demonstrate expansile hyperintense tumor in the right sphenoid sinus extending to the basisphenoid and paracavernous region.
can discern the minor tumor component found in the paracavernous area in both cases. This is consistent with the hypothesis of the sphenoid sinus schwannoma arising from divisions of the trigeminal nerve. The direct tumor extension to the paracavernous region is a supportive evidence of these tumors originating from branches of trigeminal divisions with eccentric growth into the sphenoid sinus, either through the bony erosion of lateral sinus wall or through the pathway of skull base sutures or foramen. The characteristics of the heterogeneity and enhancement patterns of sphenoid sinus schwannoma are similar to those occurring elsewhere in the head and neck, including the other sinonasal regions. Nevertheless, it is difficult to discriminate sphenoid sinus schwannoma from other skull base tumor by its paracavernous component even on multiplanar MRI, especially after the tumor has grown to considerable size.

The diagnosis of schwannomas relies mainly on histopathology. The pathology shows uniform spindle cells with features of nuclear palisading. They have been classified as Antoni A with dense cellular proliferation and Antoni B with sparse cellularity. Immunohistochemical staining positive for S-100 protein is useful for differentiating schwannomas from other neurogenic tumors. The two cases of sinonasal schwannomas we reported were histologically encapsulated, which were different form the previous studies most sinonasal schwannoma were not encapsulated although lack of encapsulation does not imply malignant potential but merely reflected in a growth pattern that was more infiltrative than schwannoma found elsewhere [1, 2, 10].

Differentiating schwannoma at the basisphenoid region from other skull base malignancies is also crucial for the initiating different treatment strategies. Surgical resection and adjuvant radiotherapy are usually sufficient for managing schwannoma whereas more aggressive adjuvant therapies are required in treating skull base malignancies. Endonasal endoscopic surgery or combination of lateral rhinotomy and Caldwell-Luc and external frontoethmoidectomy suffices for completely excision of tumors located in the nasal cavity, ethmoid or maxillary sinuses. Transsphenoid approach for excision of schwannoma is uncommon and has been reported for removal of schwannoma mimicking pituitary adenoma [11]. In case 1, complete curettage of the tumor mass was achieved through transsphenoid approach, while in case 2, adjuvant radiation therapy following transsphenoid surgery was performed because the tumor component in the paracavernous region could not be completely excised and evidence has suggested radiotherapy was beneficial in reducing schwannoma [12].

CONCLUSION

Diagnosis of sinonasal schwannoma could sometimes be difficult to clinicians and radiologist due to its infrequency, varied symptoms and non-specific imaging features. Even though, we should always keep this entity in mind.

REFERENCES

10. Mey KH, Buchwald C, Daugaard S, Prause JU. Sinonasal schwannoma-a clinicopathological analysis of five rare cases. Rhinology 2006; 44: 46-52
蝶竇之良性神經鞘瘤：兩病例報告

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根據已報告文獻，竇內良性神經鞘瘤是一少見的腫瘤，發生率約占頭頸部神經鞘瘤的不到百分之四，大多數已報告的竇內良性神經鞘瘤病例主要分布於上頜竇以及篩竇。蝶竇的良性神經鞘瘤非常罕見，由於位置上和顱底非常接近，往往無法在術前影像上被正確診斷出來。我們在本篇文章中提出了兩個病例，無論在臨床症狀及影像上均不容易跟顱底常見的腫瘤區分。因此對於在蝶竇這位置的腫瘤，良性神經鞘瘤也是我們影像上必須考慮的鑑別診斷之一。