Huge Sphenoid Sinus Olfactory Neuroblastoma: A Case Report

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Intranasal or paranasal sinus olfactory neuroblastoma is a rare malignant neoplasm of olfactory neuroepithelial origin, accounting for approximately 5% of paranasal sinus cancers. Most of the presenting symptoms include nasal obstruction, nasal bleeding, anosmia, rhinorrhea, and headache. In this present report, we describe a 79-year-old man who presented with bilateral nasal congestion for more than 1 year. Nasoendoscopy showed a huge, smooth, mucosal tumor in the nasopharynx with extension to the posterior nasal septum. The tumor was completely resected under endonasal endoscopy and the pathology revealed olfactory neuroblastoma. Olfactory neuroblastomas usually arise in the cribriform plate and superior turbinate. However, the origin and isolation of olfactory neuroblastomas to the sphenoid sinus is exceedingly rare. Only four cases of olfactory neuroblastoma isolated in the sphenoid sinus have been described in English literature and the frequency of presenting symptoms with cranial neuropathies and headache. We report a case of primary sphenoid sinus olfactory neuroblastoma with the greatest enlargement reported to date.

Key Words: cribriform plate, olfactory neuroblastoma, sphenoid sinus

bilateral inferior turbinate hypertrophy and a huge mass with smooth mucosa in the nasopharynx, which extended to the posterior nasal septum obstructing the bilateral choanal (Figure 1A). The high-resolution sinus computed tomography (CT) showed a 5×5 cm heterogeneous soft tissue density mass occupying the sphenoid sinus and the inferior sphenoid sinus wall was pushed downward to the posterior nasal cavity and choana. Sagittal view CT scans demonstrated clivus erosion and tumor extension to the posterior ethmoid sinus. Magnetic resonance imaging (MRI) showed a moderate signal intensity in both T1- and T2-weighted images. T2-weighted images revealed the enhanced heterogeneous mass was limited to both sphenoid sinuses without cavernous sinus invasion (Figure 2). After administration of contrast medium, the tumor showed slightly heterogeneous enhancement; therefore, angiofibroma and other tumors of vascular origin could be ruled out. A biopsy was performed and the pathologic findings revealed olfactory neuroblastoma in the sphenoid sinus. There was no neck lymphadenopathy and metastatic studies were negative.

Under endonasal endoscopy, the tumor was completely resected from the sphenoid sinus. The tumor originated from the floor of the sphenoid sinus with extension into the posterior ethmoid sinus, posterior septum, and clivus. The sellar dura was exposed with an eroded bony wall, but there was no evidence of intracranial invasion of tumor. Microscopic examination revealed uniform small round neoplastic cells with scant cytoplasm, dispersed coarse to fine chromatin, and focal nuclear pleomorphism. The immunohistochemical studies were positive for neuron-specific enolase, chromogranin A, synaptophysin and CD56 (Figure 3).

The patient received postoperative radiotherapy of 56 Gy over a 7-week period. The patient’s postoperative course was uneventful and he was discharged 4 days after the operation. After 16 months of follow-up, there was no evidence of recurrence.

**DISCUSSION**

Olfactory neuroblastoma is a locally extensive malignancy and frequently directly invades the skull base, orbit, and adjacent soft tissue from the typical “cribriform” site. Distant and regional metastases are present in 14–38% of cases at the time of diagnosis, and the cervical lymph nodes, lung and bone are the most commonly involved sites [3]. Olfactory neuroblastomas arising from an ectopic site including the maxillary sinus, inferior turbinate, pituitary gland and sphenoid sinus have been reported in the literature. Our case showed bilateral sphenoid sinus involvement, which explained the presence of ectopic olfactory cells resting in the sphenoid sinus.
The sphenoid sinus is an unusual site for a neoplasm, accounting for less than 1% of all paranasal sinus malignancies. Individual sphenoid sinus pathologies are variable and can include inflammatory lesions, foreign bodies, and tumors. Tumors of sphenoid sinus are often subject to late diagnosis because of their deep location in the skull. Most sphenoid sinus neoplasms have invaded adjacent structures at the time of diagnosis. Accordingly, the clinical presenting symptoms are very variable, which affects diagnosis of sphenoid sinus neoplasms. The most common source of tumors that originate in the sphenoid sinus is squamous cell carcinoma, accounting for approximately 60–75% of cases. However, olfactory neuroblastomas that originate and are isolated to the sphenoid sinus are exceedingly rare [4].

We reviewed the previous literature and only five cases (including ours) of olfactory neuroblastoma isolated in the sphenoid sinus have been reported (Table) [5–8]. The cases included two females and three males, with ages ranging from 40 to 79 years (mean age, 61 years). Most of the previous reports have described nearly equal distribution of olfactory neuroblastomas between males and females. The most

Figure 2. (A) Coronal CT scan reveals a heterogeneous soft tissue density mass occupying the sphenoid sinus with erosion of clivus. (B) Sagittal CT scan shows a huge mass extending to the posterior ethmoid sinus from the sphenoid sinus.
common presenting symptoms of sphenoid sinus olfactory neuroblastoma are headache and cranial neuropathies. Cranial nerve neuropathy occurs when the tumor erodes the bony walls of the sphenoid sinus. Headache is the most common symptom of isolated sphenoid sinus olfactory neuroblastoma and affects the occipital and vertex regions. Our case had only bilateral nasal obstruction without other symptoms of olfactory neuroblastoma, despite severe destruction of the sphenoid sinus wall. Using Kadish et al’s classification system [9], the disease was classified as stage B in three patients, and stage C for two patients; no patient was at stage A. The early stage is usually asymptomatic or has nonspecific symptoms. Therefore, this tumor often presents only at an advanced stage at the time of diagnosis.

Traditionally, the treatment of choice for olfactory neuroblastoma is a combined approach with craniofacial resection and postoperative radiotherapy. This invasive procedure entails serious complications, morbidity and increased length of hospitalization [10]. The most common complications include cerebrospinal fluid leakage, intracranial hemorrhage and infection. The sphenoid sinus is located deeply in the skull; therefore, craniofacial resection is not a suitable intervention for sphenoid tumors. Combined endonasal endoscopic resection and radiotherapy are minimally invasive procedures for the treatment of sphenoid

Table. Clinical data for five patients with primary sphenoid sinus olfactory neuroblastoma

<table>
<thead>
<tr>
<th>Case</th>
<th>Author</th>
<th>Age (yr)/sex</th>
<th>Symptoms</th>
<th>Kadish staging</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Chacko et al,</td>
<td>62/F</td>
<td>Right facial numbness, weakness &amp; hearing impaired 2 mo</td>
<td>C</td>
<td>Surgical excision</td>
<td>Died after operation</td>
</tr>
<tr>
<td>2</td>
<td>Sharma et al,</td>
<td>40/M</td>
<td>Intermittent blood-stained postnasal drip &amp; headache</td>
<td>B</td>
<td>Combined CT &amp; RT</td>
<td>Alive; NER at 11 mo</td>
</tr>
<tr>
<td></td>
<td>2002 [6]</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Chirico et al,</td>
<td>59/F</td>
<td>Headache, left facial pain, diplopia</td>
<td>B</td>
<td>Combined CT &amp; RT</td>
<td>Unavailable</td>
</tr>
<tr>
<td></td>
<td>2003 [7]</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>4</td>
<td>Morris et al,</td>
<td>63/M</td>
<td>Hyperprolactinemia</td>
<td>B</td>
<td>Endoscopic en bloc excision</td>
<td>Alive; NER at 15 mo</td>
</tr>
<tr>
<td></td>
<td>2004 [8]</td>
<td></td>
<td></td>
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<tr>
<td>5</td>
<td>Chan et al,</td>
<td>79/M</td>
<td>Bilateral nasal obstruction for &gt;1 yr</td>
<td>C</td>
<td>Endoscopic excision &amp; postoperative RT</td>
<td>Alive; NER at 16 mo</td>
</tr>
<tr>
<td></td>
<td>2009 [This case]</td>
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CT = chemotherapy; RT = radiotherapy; NER = no evidence of recurrence.
sinus olfactory neuroblastomas. These procedures can reduce morbidity and mortality, with short hospitalization time, and can improve the quality of life. However, endonasal endoscopic resection increases the risk of locoregional relapse [11]. In previous reports, Castelnuovo et al [2] and Suriano et al [11] described that endonasal endoscopic resection combined with postoperative radiotherapy offers a good quality of life and excellent outcomes for disease control. Chemotherapy is used in cases of advanced, unresectable tumors, or under poor operative conditions, where it is used in combination with radiotherapy. Morris et al [8] reported one case that was treated with endonasal endoscopic resection of a primary sphenoid sinus olfactory neuroblastoma and enjoyed disease-free actuarial survival for 15 months. Our patient, who underwent endonasal endoscopic resection combined with postoperative radiotherapy (56Gy), responded completely to the treatment.

Isolated sphenoid olfactory neuroblastoma is an uncommon neoplasm and the most frequent presenting symptoms include cranial neuropathy and headache. It is distinct from intranasal olfactory neuroblastomas. Therefore, we suggest considering sphenoid sinus disease if cranial neuropathies and persistent headache are present. Nasoendoscopy can provide a clearer field of vision, particularly in patients with severe hypertrophy of the inferior turbinate. Although bilateral nasal congestion is the most common presentation of many diseases, the differential diagnosis should include sphenoid sinus olfactory neuroblastoma.

REFERENCES

巨大蝶竇嗅覺神經母細胞瘤—病例報告

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嗅覺神經母細胞瘤是一種非常罕見源自嗅覺上皮的鼻腔或鼻竇惡性腫瘤，約佔鼻竇惡性腫瘤的 5%。最常見的症狀為鼻塞及流鼻血，其次尚有以嗅覺喪失、流鼻水、頭痛等症狀表現。本科經歷一位 79 歲男性病患，主訴雙側鼻塞已經一年且藥物治療無效，利用鼻腔內視鏡檢查發現後鼻孔被一巨大且黏膜平滑之腫瘤阻塞，且侵犯鼻中隔後方。經鼻竇內視鏡手術將腫瘤完全切除，病理報告證實為嗅覺神經母細胞瘤。嗅覺神經母細胞瘤常發生在篩狀板及上鼻甲，而單純發生在蝶竇十分罕見，目前全球只有四個病例被報導，多以顱內神經異常及頭痛表現。而如此巨大且侵犯雙側蝶竇的病例未曾被報導。

關鍵詞：篩狀板，嗅覺神經母細胞瘤，蝶竇
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