

## Sphenoid Sinus Myxoma: Case Report and Literature Review

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### ABSTRACT

**Objectives:** We present the first known case in the English-language literature of a myxoma arising in the sphenoid sinus. By describing the patient's clinical course and the salient features of this rare neoplasm, we seek to increase the awareness of the presentation, histological features, and treatment considerations for myxomas of the head and neck. In the process, we intend to describe the work-up of isolated sphenoid sinus lesions and focus on the varying and evolving techniques for surgical access to the sphenoid sinus.

**Study Design and Methods:** Case report and literature review.

**Results:** We describe the clinical course of a patient with a myxoma of the sphenoid sinus. The patient underwent an external sphenoidectomy through a lateral rhinotomy approach with medial maxillectomy under MRI-guidance. He remains without evidence of recurrent disease after 8 months.

**Conclusions:** Myxomas of the head and neck are rare neoplasms. Their infiltrative nature and tendency to recur demand an aggressive surgical approach that may be accomplished with minimal morbidity using currently available image-guided techniques.

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### INTRODUCTION

Myxomas are benign neoplasms derived from primitive mesenchyme. These lesions occur most

frequently in the atria, but they can also be found in bone, skin, subcutaneous tissue, and skeletal muscle. Initially coined by Virchow in 1871, the term *myxoma* traditionally has been used to describe tumors that resemble the mucinous appearance of the umbilical cord.<sup>1</sup> However, other tumors, particularly certain soft tissue sarcomas, also exhibit myxomatous features, prompting Stout to further characterize myxomas as nonmetastasizing tumors of mesenchymal origin without histological evidence of other recognizable elements such as chondroblasts, lipoblasts, and myoblasts.<sup>2</sup>

In general, myxomas of the head and neck are rare neoplasms, infrequently reported in the literature. The majority of these tumors arise from the mandible and maxilla, with only scattered reports of myxomas of the temporal bone, pharynx, larynx, soft tissues of the face and neck, and the paranasal sinuses.<sup>3–5</sup> Of the paranasal sinus myxomas, the only traceable reports describe lesions predominately arising in the maxillary sinus. We describe the first known case of a myxoma arising in the sphenoid sinus with aggressive extension into surrounding structures.

### CASE REPORT

A 45-year-old male presented with a 1-year history of progressive left-sided nasal obstruction without rhinorrhea or epistaxis. Although he had a history of wood dust exposure, he denied exposure to chromium and nickel, as well as prior trauma to the head and neck. This history is pertinent since exposure to these products has been associated with the diagnosis of squamous cell carcinoma of the paranasal sinuses. His past medical and surgical histories were essentially unremarkable, and routine physical examination, including anterior rhinoscopy, was unrevealing. However, flexible fiberoptic nasopharyngoscopy demonstrated a pearly white mass with vascular markings just posterior to the middle turbinate that almost completely obstructed the left nasal cavity.

Due to the patient's sinonasal complaints, and the concerning findings on endoscopy, radiographic evaluation was undertaken with both computed

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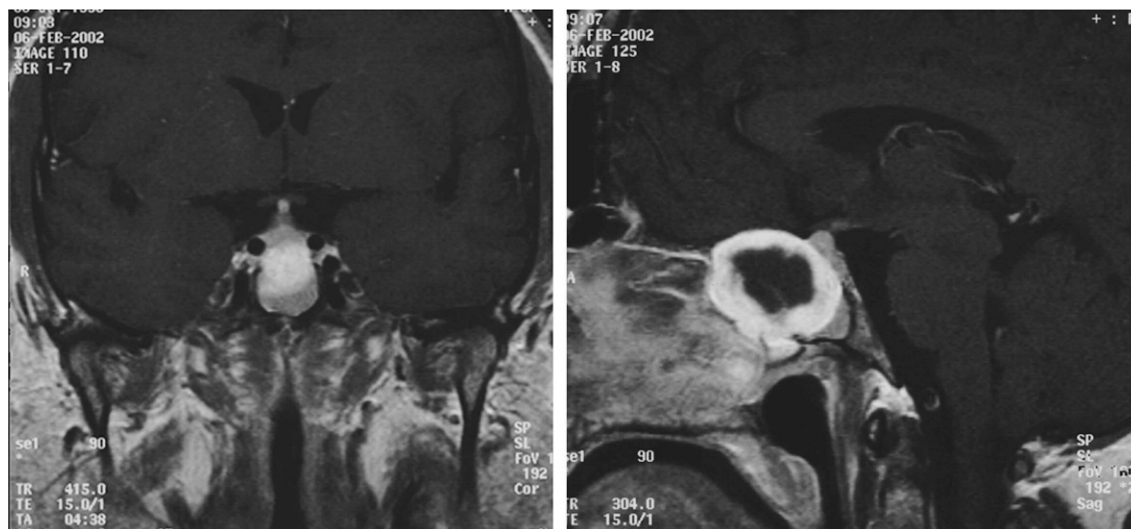
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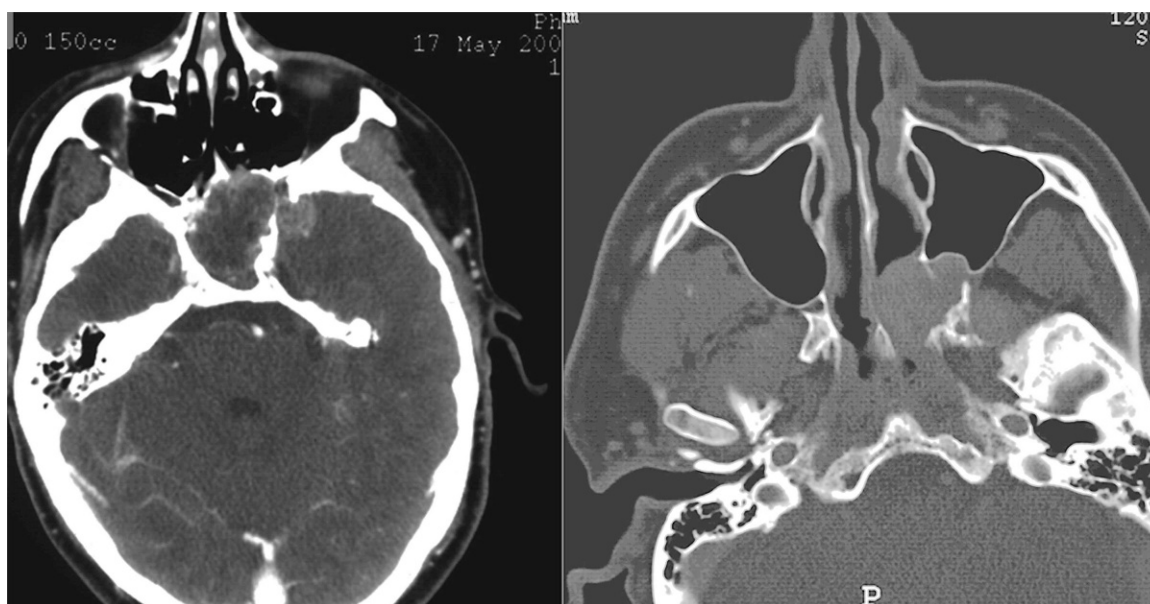
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**Figure 1. Contrast axial CT scan demonstrating soft tissue mass filling the sphenoid sinus, with extension into the left middle cranial fossa, pterygopalatine fossa, and posterior ethmoid sinus. Note the anterior bowing of the posterior wall of the left maxillary sinus, as well as the destruction of the pterygoid plates.**

tomography (CT) and magnetic resonance imaging (MRI). The CT scan exhibited a non-enhancing, 3 × 5 cm soft-tissue mass centered in the sphenoid sinus and extending into the left ethmoid sinuses and the pterygopalatine fossa, as well as the middle cranial fossa (Figure 1). The lesion appeared to have caused both erosion of the pterygoid plates and remodeling of the posterior maxillary sinus wall, suggesting an indolent growth pattern. Enhancement with contrast and central necrosis were evident on post-contrast T1-weighted images, which also demonstrated encroachment on the carotid arteries and pituitary

gland, with anterior extension along the planum sphenoidale (Figure 2). The mass extended laterally into the middle cranial fossa and filled the pterygopalatine fossa in the region of foramen rotundum (Figure 3). Inferiorly, it eroded the anterior sphenoid floor as well as the posterior aspect of the left middle turbinate. An endoscopic biopsy was performed at an outside institution, demonstrating myxoid fragments, fibrous tissue, and sinonasal mucosa with varying degrees of cellularity, as well as stellate neoplastic cells that suggested the diagnosis of myxoma. The patient was then referred to our institution for definitive care.



**Figure 2. Gadolinium-enhanced T1 weighted MRI images of the sphenoid sinus mass. It appears to encase both carotid arteries and closely approximate the pituitary gland, with anterior extension along the planum sphenoidale.**



**Figure 3. Coronal view of contrasted MRI. The mass spills anteriorly and laterally from the sphenoid sinus, extending throughout the pterygomaxillary space in the region of the vidian canal and foramen rotundum.**

After treatment options were discussed and informed consent was obtained, the patient underwent an extradural anterior skull base approach with lateral rhinotomy, medial maxillectomy, external ethmoidectomy, and sphenoidectomy with mini-facial translocation. Because of the complex anatomical relationships of the sphenoid sinus, we employed a 3-D MRI-based navigational system (Stealth®, Medtronic, Memphis, TN) to provide accurate intraoperative anatomic localization. A partially encapsulated mass was noted to have eroded the bone of the superior and lateral sphenoid sinus, with preservation of a thin bony covering over the carotid arteries. Although the dura of the anterior and middle cranial fossae was extensively exposed, it remained intact. The lesion extended grossly into the left pterygopalatine fossa where it eroded portions of both pterygoid plates. Despite the extent of disease, the mass was completely excised without evidence of dural or carotid injury. We utilized the image guidance system frequently during tumor extirpation as an adjunct to known anatomic landmarks of the skull base.

Histopathologic review of the surgical specimen supported the diagnosis of myxoma, with stellate cells in areas of low to normal cellularity visible on both light and electron microscopy in a background of mucoid material (Figure 4). Additionally, immunohistochemical staining was performed, revealing that the tumor cells were positive for vimentin and negative for S-100, low molecular weight cytokeratin (CAM 5.2), broad spectrum cytokeratin (AE1/AE3), epithelial marker antigen, and carcinoembryonic antigen. The



**Figure 4. Photomicrograph of tumor specimen at 10× magnification with hematoxylin and eosin stain.**

patient has done well postoperatively and remains without evidence of recurrent disease more than 1 year from diagnosis.

## DISCUSSION

Despite their description more than 130 years ago, myxomas of the head and neck remain rare neoplasms. Exhaustive reviews of pathologic specimens from major centers have identified myxomas in 0.1% of cases.<sup>6</sup> As demonstrated in this review, most myxomas in the head and neck occur in the mandible, followed by the maxilla.<sup>6</sup> Other described sites of origin for myxomas of the head and neck are listed in Table 1. Head and neck myxomas have been identified in patients ranging in age from 15 months to the 8th decade, although a peak incidence has been

**Table 1. Site of Origin of Head and Neck Myxomas**

Bone	Soft Tissue
Mandible	Paranasal sinuses
Maxilla	Oropharynx
Nasal bones	Larynx
Temporal bone	Parotid
Palate	Tonsils
	Oral cavity
	Subcutaneous tissue

**Table 2. Isolated Sphenoid Sinus Disease\***

Infectious/Inflammatory	Neoplastic	Other
Sinusitis (Acute/Chronic)	Carcinoma	Cerebrospinal fluid leak
Fungal infection	Inverting papilloma	Foreign body
Allergic fungal sinusitis	Fibro-osteoma	
Mycetoma	Lymphoma	
Polyp	Hemangioma	
Mucocele	Pituitary adenoma	
Mucus retention cyst		

\* Modified from Wang et al.<sup>13</sup> and Martin et al.<sup>14</sup>

described between the ages of 25 and 35 years. There is a slight female predominance, but there does not appear to be an ethnic predisposition.<sup>3</sup>

Paranasal sinus myxomas comprise a minority of the cases in the head and neck, and they may be among the most difficult to diagnose and treat. Heffner describes nine pediatric sinonasal myxomas from a survey of the Armed Forces Institute of Pathology registry of otorhinolaryngologic specimens between 1976 to 1981, eventually adding four more cases to the literature.<sup>7,8</sup> Other authors have reported myxomas arising in the paranasal sinuses, nearly all of which involve the maxillary sinus.<sup>4,9-12</sup> Although most of these lesions remain well localized, certain tumors have demonstrated more aggressive behavior, as evinced by involvement of the nasal cavity, as well as the ethmoid sinus, frontal sinus, skull base, orbit, and pterygopalatine fossa.<sup>4,11</sup> No cases of sphenoid sinus involvement have previously been reported in the English-language literature.

Regardless of etiology, sphenoid sinus disease remains a challenging clinical entity. Numerous recent reports document the inherent difficulties in diagnosing and treating problems in this poorly accessible anatomic site.<sup>13,14</sup> Common symptoms, such as headache, visual changes, epistaxis, and cerebrospinal fluid rhinorrhea, are often nonspecific and frequently precipitate a delay in diagnosis. The proximity of the sphenoid sinus to the vital structures of the cavernous sinus and sella turcica at the skull base and middle cranial fossa underscores the importance of early diagnosis and treatment, so that complications such as optic neuritis, cranial nerve palsies, pituitary dysfunction, and intracranial extension do not occur. Fortunately, increased utilization of CT and MRI scans in the work-up of sinonasal complaints has increased detection of isolated sphenoid sinus disease, with diagnosis further aided by endoscopic evaluation and biopsy.<sup>14</sup> Table 2 depicts a few of the most frequently encountered causes of isolated sphenoid sinus disease.

Although the histologic appearance of myxomas is well described, theories regarding the etiology of this tumor abound. The nomenclature of myxomas, although not universally employed, seeks to delineate the potential histogenesis of these lesions. Myxomas are regarded as either bone-derived or soft tissue-derived, depending on their location, with the bone-derived forms further subdivided into odontogenic and non-odontogenic forms.<sup>15-17</sup> Historically, myxomas were thought to arise from primitive mesenchyme, due to their similarity to the primitive connective tissue in the embryo.<sup>2</sup>

Myxomas may arise from rests of odontogenic mesenchyme or from modified fibroblasts associated with tooth germ; these altered fibroblasts may hypersecrete mucin.<sup>18-20</sup> An association between myxoma formation and the presence of unerupted teeth has provided support for this position.<sup>9</sup> However, evidence exists against the odontogenic theory as well; Slootweg and Wittkampf have demonstrated that the extracellular matrix of myxomas differs histologically from the extracellular matrix of dental and periodontal tissues.<sup>6</sup> The simple fact that a myxoma may arise in the non-odontogenic tissues of the sphenoid sinus challenges the odontogenic hypothesis, but no definitive answer has emerged.

On evaluation, a myxoma appears as a glistening, grey-white, gelatinous mass with discrete surface nodularity and a variable consistency.<sup>20</sup> The lesions demonstrate varying degrees of encapsulation, and a pseudocapsule may even develop as a result of compression of surrounding normal structures. The tumor discussed in our case exhibited intratumoral variation in the amount and presence of a capsule, with a well-formed capsule adjacent to the anterior cranial fossa, but no evidence of a capsule in the middle fossa and pterygopalatine fossa extensions. The amorphous, gelatinous texture of myxomas complicates their complete removal and has been identified as a prime contributor to tumor recurrence, which occurs in up to 25% of cases. The lack of a true

**Table 3. Differential Diagnosis of Myxoid Tumors**

Benign Lesions	Malignant Lesions
Neurofibroma	Myxoid chondrosarcoma
Neurilemoma	Myxoid liposarcoma
Schwann cell tumor	Malignant fibrous histiocytoma
Myxolipoma	Embryonal rhabdomyosarcoma
Myxochondroma	
Pleomorphic adenoma	

capsule and the potential for secretion of bioactive enzymes such as hyaluronic acid and acid phosphatase contribute to the locally infiltrative nature of this tumor.<sup>11</sup>

Myxomas are composed of stellate-shaped cells with slender cytoplasmic processes and small nuclei embedded in a mucoid matrix rich in polysaccharides.<sup>21,22</sup> Reticulin fibers are evident, although the amount of collagen is variable, potentially due to the production of mucopolysaccharides by myofibroblasts.<sup>23</sup> The cellularity of a myxoma is typically minimal, secondary to the vast mucoid stroma, although some variants with increased cellularity and enhanced spindle cell morphology have been documented.<sup>7</sup> Ultrastructurally, myxomas appear as scattered tumor cells containing abundant rough endoplasmic reticulum, well-developed Golgi apparatus, numerous filaments, and pinocytotic vesicles within a background of low electron density.<sup>17</sup>

The myxoid appearance of these tumors resembles several other lesions, both benign and malignant, which are summarized in Table 3. Ultimately, the diagnosis of a myxoma is a diagnosis of exclusion. Increased cellularity should raise suspicions for one of the malignant myxoid tumors, as a true myxoma is considered a relatively hypocellular tumor. Special staining techniques and immunohistochemistry are helpful adjuncts to clinical cues in identifying and differentiating these lesions. The mucoid matrix of a myxoma stains strongly with Alcian blue and mucicarmine, as well as periodic acid–Schiff, although these are not specific. Immunostaining reveals that myxomas stain positive for vimentin and actin, but negative for desmin, S-100, and cytokeratin.<sup>9,23</sup>

After an accurate diagnosis is achieved, complete surgical excision is the treatment of choice. Intranasal, transantral, transseptal, transethmoidal, and various external approaches have all been utilized to perform a complete surgical excision.<sup>3,4,8,13</sup> With recurrence rates often quoted at 25% and usually attributed to inadequate primary excision, radical primary resection has been advocated by some authors, with en bloc removal when possible.<sup>4</sup> Furthermore, aggressive variants of myxomas have been reported that

**Table 4. Approaches to the Sphenoid Sinus**

Endoscopic
Transnasal
Transseptal
Pterygoid fossa <sup>14</sup>
Transseptal
Sublabial
Midfacial degloving
External sphenoidectomy
Anterior craniofacial resection

have exhibited marked infiltration and erosion into surrounding structures, resulting in significant morbidity and even death.<sup>11</sup> Citing the risk of recurrence and the potential for the myxoma to endanger surrounding structures, Deron et al. advocate “radical primary resection in spite of significant functional and aesthetic sequelae.”<sup>11</sup> However, such radical resection is not always possible or practical, particularly at the skull base. Resection in these areas, as in our patient, requires excellent exposure and meticulous surgical technique to ensure complete tumor removal. The challenges inherent in skull base resection of myxomas have been previously described.<sup>24</sup> Despite these limitations, complete surgical resection provides the best chance for cure, as these tumors do not respond appreciably to radiation therapy or chemotherapy.<sup>11</sup>

Advances in surgical technique, particularly the use of image-guided systems, may allow complete resection of myxomas while minimizing the functional and aesthetic consequences. Although numerous options exist for the extirpation of sphenoid sinus lesions, we opted to perform a lateral rhinotomy, medial maxillectomy, ethmoidectomy, and sphenoidectomy with minifacial translocation in our patient to maximize the exposure of the tumor and its finger-like projections. Frozen section analysis provided an assessment of tumor margins in an area where en bloc resection is not reasonable for benign disease. Exposure provided by the lateral rhinotomy facilitated adequate visualization of the pterygopalatine fossa extension of disease. This approach provided an excellent view of the skull base with, ultimately, an acceptable oncologic and cosmetic result. Our position could be readily determined with the image-guided system, particularly as we approached the superior and lateral recesses of the sphenoid sinus with underlying carotid arteries and optic nerves. Although numerous other approaches to sphenoid sinus could have been employed, including endoscopic, sublabial, and craniofacial techniques (Figure 4), we maintain that our method provides excellent exposure while minimizing intraoperative risk and morbidity.

## CONCLUSIONS

Myxomas of the head and neck are rare neoplasms, commonly involving the mandible and maxilla. We describe the first known case of an aggressive myxoma arising from the sphenoid sinus and extending to the middle and anterior cranial fossae, the maxillary sinus, and the pterygopalatine fossa. Because myxomas are infiltrative lesions with a propensity to recur locally if not completely excised, we aggressively resected the lesion with frozen section control of margins through a lateral rhinotomy, medial maxillectomy, and external sphenoidectomy approach. The risks of radical excision must be balanced with the risks of tumor recurrence in determining the appropriate surgical technique. The use of image-guided surgery may allow for more precise and thorough tumor extirpation while minimizing cosmetic and functional sequelae.

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