Nasal Solitary Fibrous Tumor Arising from the Anterior Cranial Fossa

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ABSTRACT

The solitary fibrous tumor (SFT) is a mesenchymal, spindle cell neoplasm that was originally found in pleural tissue. Recently, however, numerous extrapleural sites have been discovered, including the nasal cavity. We present the 15th case of a nasal SFT, and the first such tumor to arise from the cribriform plate and extend into the anterior cranial fossa. In addition to highlighting the aggressive nature of this tumor, we review its clinical features and the diagnostic difficulties posed by SFT.

KEYWORDS: Solitary fibrous tumor, nasal, anterior skull base

The solitary fibrous tumor (SFT), a spindle cell lesion that occurs in adults, was first described in 1931 by Klemperer and Rabin.¹ Much debate concerning its exact histogenesis ensued, and early reports attributed this tumor to pleural mesothelial tissue. Currently, however, SFT is commonly described as a mesenchymal tumor.² This distinction may be of minor significance: mesothelial cells are derived from the primitive mesoderm, and SFTs therefore have always been “mesenchymal” tumors. Numerous extrapleural sites of SFTs have recently been discovered. Within the head and neck, examples include the orbit,^{3,4} salivary glands,^{5,6} soft tissue of the neck⁷ and face,⁸ oral cavity,^{9,10} upper respiratory tract,¹¹ thyroid,¹² and temporal region.¹³ We found reports of 14 cases of SFTs located in the nasal cavity, often with extensions into the paranasal

sinuses and nasopharynx.^{2,13–19} We present a 15th case of an SFT within the nasal cavity, and the first case of an SFT to arise from and extend through the cribriform plate. We describe the potential for intracranial involvement and the diagnostic dilemma posed by this rare tumor.

CASE REPORT

A 31-year-old woman sought treatment at an outside institution with a 2-year history of headaches, midfacial pain, and nasal congestion of gradually increasing severity. Originally, she was diagnosed with sinusitis and treated with nasal steroid sprays and antibiotics. Her condition failed to improve.

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Figure 1 (A) Axial CT and (B) coronal MRI showing a large mass filling the right nasal cavity, extending into the right ethmoid sinuses, displacing the nasal septum to the left, and laterally displacing both laminae papyracea. The mass appears to be pedicled superiorly to the dura of the anterior cranial fossa. CT, computed tomography; MRI, magnetic resonance imaging.

She underwent computed tomography (CT) and magnetic resonance (MR) imaging (Figs. 1A, B), which showed a large mass occupying the right nasal cavity. The mass extended into the right ethmoid sinus and displaced the nasal septum and both lamina papyracea laterally. Bony destruction of the cribriform plate was evident. A biopsy specimen was obtained under general anesthesia, but the procedure was complicated by extensive hemorrhage that was ultimately controlled by nasal packing. The patient was then referred to our institution for further treatment.

Embolization of the mass was considered, but angiography showed that the right anterior ethmoid artery was the primary vascular supply, contraindicating this nonoperative treatment. The mass was subsequently resected surgically via a transcranial approach, with a bicoronal skin flap and subfrontal extradural exposure (Fig. 2). Intraoperative findings included a visible defect in the anterior skull base

with tumor adherence to the dura overlying the right cribriform plate. Excision of a small piece of dura in this region was necessary to allow complete removal of the tumor, which measured $5 \times 4 \times 3$ cm. The anterior skull base was reconstructed with layered temporalis fascia and fibrin glue, and osteoplastic frontal sinus obliteration was performed with an abdominal fat graft. Total blood loss of almost 3,000 cc necessitated the infusion of two units of autologous blood, two units of packed red blood cells, and 450 cc of autologous blood harvested with a cell saver. The patient recovered uneventfully and has been free of tumor for 3.5 years.

Histopathologically, the tumor was well circumscribed and firm to palpation. Microscopic analysis revealed a variable proliferation of cells whose appearance ranged from plump to spindled. The cells were dispersed in a random herringbone pattern (Fig. 3). Numerous blood vessels of

Figure 2 Intraoperative appearance of the SFT described in the current case. The bicoronal skin flap is retracted inferiorly (top). The nasal bones and inferior portion of the frontal bone have been removed, allowing adequate access to the tumor. SFT, solitary fibrous tumor.

different sizes were identified, some of which were thick-walled. There was almost no mitotic activity, and no necrosis was present. Glandular structures, psammoma bodies, and keratin pearls were also absent. Immunohistochemical testing resulted in prominent staining for CD-34 and variable reactivity for vimentin. Negative results were obtained

after staining for epithelial membrane antigen (EMA), S-100 protein, HMB 45, SMA (smooth muscle actin), cytokeratin, LCA (leukocyte common antigen), actin, factor XIII, and factor VIII. A separate specimen of olfactory dural tissue revealed no definitive evidence of involvement with the tumor.

Figure 3 Hematoxylin and eosin-stained microscopic image of the patient's SFT, shows a varying herringbone pattern of plump and spindled cells with prominent vascularity. SFT, solitary fibrous tumor.

DISCUSSION

The macroscopic pathological features of SFTs include a well-circumscribed, unencapsulated mass with a nodular surface, often tethered by a pedicle. The cut surface is firm and white or gray.^{11,17} Microscopically, spindled cells of various widths are scattered across a collagenous background. Histological arrangement is often described as patternless, storiform, fascicular, or herringbone. SFTs also often display prominent vascularity.²

In addition to examination of the pathologic features, immunohistochemistry is essential for the correct diagnosis of SFTs. Positive staining for CD-34, CD-99, and vimentin is common. Markers for which staining is negative include keratin, EMA, S-100 protein, factor VIII-related antigen, carcino-embryonic antigen, desmin, HHF-35, and glial fibrillary acidic protein.^{9,15}

Previously reported cases of SFT in the nasal cavity are summarized, with the current case, in Table 1. Several of these tumors infiltrated the ethmoid sinuses and extended posteriorly to the nasopharynx. No previous case, however, eroded the anterior skull base and involved the anterior cranial fossa. Nasal SFTs typically lead to nasal

obstruction and also may be associated with epistaxis, rhinorrhea, anosmia, headache, facial pain, and visual disturbances caused by orbital pressure. Based on these symptoms and appropriate radiologic imaging, the clinical differential diagnosis includes benign anterior cranial lesions such as meningiomas, malignant lesions such as fibrosarcomas, hemangiopericytomas, nasopharyngeal carcinomas, or esthesioneuroblastomas. However, the diagnosis must be confirmed pathologically. Even then diagnosis can be difficult without performing immunohistochemical analysis. The histologic differential diagnosis of SFTs includes hemangiopericytomas, schwannomas, fibrous histiocytomas, fibrosarcomas, and nasopharyngeal angiofibromas.¹¹

Effective treatment involves complete resection of the tumor, which has been attained by a transcranial approach (the current case), local excision, maxillectomy, sphenoidectomy, or external ethmoidectomy.¹⁴ The possibility of profuse bleeding must be considered during resection and even during initial biopsies. Proximity to vital structures near the nasal cavity may limit exposure and necessitate two or more procedures to achieve total resection. Nonetheless, no nasal SFT has been reported to recur or metastasize. Prognosis can be

Table 1 Summary of Reported Nasal Cavity SFT

Case	Age/Sex	Location	Size (cm)	Original Diagnosis	Follow-up
1 ²	31/F	Nasal cavity	2	SFT	NA
2 ¹¹	64/F	Nasal cavity	"Polyp"	Esthesioneuroblastoma	NED 12 years
3 ¹¹	36/F	Nasal cavity, sphenoid sinus	7 × 4 × 3	Reparative process	NA
4 ¹¹	47/F	Nasal cavity, nasopharynx	4 × 3.5	NA	NED 4 years
5 ¹¹	55/M	Nasal cavity	NA	NA	NED 2 years
6 ¹³	61/F	Nasal cavity	6 × 3	SFT	NED 20 mos.
7 ¹³	51/F	Nasal cavity	NA	SFT	NA
8 ¹⁴	54/M	Nasal cavity	5 × 4 × 3	SFT	NED 2 years
9 ¹⁵	55/M	Nasal cavity, ethmoid sinus	NA	Hemangiopericytoma	NED 4 years
10 ¹⁵	53/M	Nasal cavity, ethmoid sinus, nasopharynx	NA	Granulation	NED 2 years
11 ¹⁶	33/F	Nasal cavity	4	Fibroma	NED 1 year
12 ¹⁷	59/F	Nasal cavity, ethmoid sinus, nasopharynx	5 × 3 × 3	SFT	NED 18 mos.
13 ¹⁹	48/F	Nasal cavity, ethmoid sinus	NA	Spindle cell tumor	NED 1 year
14 ¹⁹	45/F	Nasal cavity, ethmoid sinus	NA	Spindle cell tumor	NED 6 mos.
Current	31/F	Nasal cavity, maxillary and ethmoid sinuses, anterior cranial fossa	5 × 4 × 3	Spindle cell tumor	NED 3 years

SFT, solitary fibrous tumor; NA, not available; NED, no evidence of disease.

assessed based on the presence or absence of histological malignant features, which include high cellularity, high mitotic activity, and cellular pleomorphism. However, such features do not necessarily lead to clinically malignant behavior. Only one nasal SFT¹⁵ displayed several of these features, but it was resected successfully without recurrence. Conversely, the current case behaved aggressively, with erosion through the anterior skull base, but it lacked histological signs of malignancy. The predominantly benign nature of nasal and other extrapleural SFTs contrasts with the more aggressive clinical behavior of pleural tumors, displayed by as many as 23% of such masses.²⁰

In conclusion, we present the first case of an SFT arising from the anterior skull base and the 15th reported nasal SFT in the English literature. Our case demonstrates the diagnostic difficulty inherent in such tumors and emphasizes the potential complications associated with hemorrhage during biopsy and treatment. Furthermore, this case represents the only report of tumor extension through the roof of the nasal cavity into the anterior cranial fossa.

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