Endoscopic Resection of Solitary Fibrous Tumors of the Nose and Paranasal Sinuses

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ABSTRACT

Solitary fibrous tumors (SFTs) are uncommon neoplasms of mesenchymal origin that were first described as primary spindle-cell tumors of the pleura in 1931. Since then, infrequent case reports of extrapleural SFTs have been described including various subsites within the head and neck. Based on a review of the literature and a description of the endoscopic treatment of three patients with SFTs of the nasal cavity and ethmoid sinuses, the challenges associated with the management of sinonasal SFTs are discussed. Successful endoscopic resection was performed at a tertiary referral rhinology practice within a university center in three cases of sinonasal SFTs with no evidence of recurrence at 26, 35, and 49 months following resection. Summarized are the common presenting symptoms, appropriate diagnostic workup, and indicative computed tomography and magnetic resonance imaging appearance of SFTs. Further discussed are the challenge associated with accurate histological and immunohistochemical diagnosis, the difficulty in assessing the aggressiveness and malignant potential of these lesions, and the appropriate treatment and follow-up duration that these neoplasms require.

KEYWORDS: Solitary fibrous tumor, endoscopic resection, sinus

Solitary fibrous tumors (SFTs) are rare tumors that have been previously described as benign fibrous mesotheliomas, localized fibrous mesotheliomas, or submesothelial fibromas. They are spindle-cell tumors that were originally described originating from the pleura by Klemperer and Rabin in 1931.¹ Due to their mesenchymal origin, as opposed to the originally proposed mesothelial origin, several extrapleural sites with no relationship to serosal surfaces have since been described. In the head and neck, SFTs have been documented in the external auditory canal, lacrimal sac, epiglottis, larynx, thyroid, sublingual gland, parotid gland, tongue, gingiva, orbit, parapharyngeal space, nasopharynx, hy-

poglossal nerve, scalp, and infratemporal fossa as well as the nasal cavities and paranasal sinus. This series reports the endoscopic endonasal treatment of a series of three cases of SFTs arising from the nose and paranasal sinuses. Highlighted in the discussion are the challenges associated with their diagnosis, treatment, appropriate adjuvant therapy, and adequate length of follow-up.

CASE SERIES

Three cases of SFTs of the nose and paranasal sinuses were treated via endoscopic resection between June 2006 and April 2008. The patients included one woman and

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two men of ages 33, 36, and 41, respectively. One patient was completely asymptomatic. A second patient, aged 41 at the time of resection, presented solely with the complaint of unilateral nasal obstruction. The third patient, aged 36 at the time of resection, experienced symptoms of nasal obstruction, decreased sense of smell, unilateral epiphora, slight broadening of the nasal dorsum, and very mild lateral displacement of the ipsilateral globe (on clinical examination), which did not affect his vision. The endoscopic appearance of each of these lesions was that of a smooth, rubbery, encapsulated, unilateral nasal mass. Imaging revealed an expansile, homogeneous nasal mass that bowed portions of the septum in each case (Fig. 1). They filled the nasal cavities and ethmoid sinuses to variable degrees and in two of the cases caused pressure erosion of the anterior skull base with no evidence of dural invasion. Additionally, in one case, the tumor caused pressure erosion of the lamina papyracea and lateral displacement of the bony nasal dorsum.

None of the preoperative biopsies were reported as SFTs. Instead they were reported as hemangiopericytoma, hyalinized scar, and inflammatory polyp with overlying squamous metaplasia, respectively. Each of the lesions was removed completely with an endoscopic resection, without complication. In the cases with pressure erosion of the cribriform plate and/or lamina papyracea, there was no evidence direct involvement or invasion of the dura or periorbita. Final pathology in each case revealed a uniform spindle-cell proliferation with ill-defined borders and alternating regions of variable cellularity. The spindle cells were arranged haphazardly and interspersed thick collagen bundles were present. Scattered throughout were thin walled vascular spaces, which in some areas had a “staghorn” configuration. Other blood vessels had hyalinized walls. Rare mitoses were identified in one of the tumors. There was no evidence of malignancy in any of the specimens (Fig. 2 and Fig. 3). Hematoxylin and eosin

Figure 1 Computed tomography of right ethmoid solitary fibrous tumor—note bowing of nasal septum and pressure erosion of right lamina papyracea.

Figure 2 Hematoxylin and eosin–stained section at original magnification $\times 5$.

(H&E) section of one of the tumors showed the typical haphazardly arranged spindle cells with variable cellularity and thick collagen bundles. H&E section of the same tumor showed a cellular area with uniform spindle cells and associated vascular spaces. All tumors showed positive immunohistochemical staining for CD34, vimentin, and bcl2. They were negative for S-100, desmin, and actin. As such, they were reported as SFTs (Fig. 2 and Fig. 3).

All patients recovered uneventfully from their endoscopic resections and have been followed clinically, including serial endoscopy and computed tomography (CT). All patients were free of disease recurrence at 26, 35, and 49 months, respectively, following resection.

DISCUSSION

Twenty-three cases of SFTs of the nasal cavity and/or paranasal sinuses have been previously reported in the literature.² These reports vary in terms of their description of the inherent nature of these lesions. At present, there is no consensus as to the appropriate elements

Figure 3 Hematoxylin and eosin–stained slide at original magnification $\times 10$.

required for conclusive diagnosis, appropriate treatment, and adequate length of follow-up. This review aims to highlight these challenges in the management of SFTs and offers an evidenced-based opinion on these issues based on our cases and review of the existing literature.

SFTs have been described throughout adulthood and occur equally in men and women. Their common clinical presentation in the nose and sinuses is that of progressive nasal obstruction and/or secondary symptoms of sinus obstruction or displacement of adjacent structures, depending on their location. No particular area of involvement of the nose or sinuses has been identified as occurring more frequently, although this would be difficult to discern with the limited number of cases described.

The endoscopic appearance is routinely described as that of a firm, lobulated, and well-encapsulated mass without surrounding tissue reaction. The differential diagnosis of lesions with this clinical appearance is very broad and includes epithelial neoplasm, esthesioneuroblastoma, meningioma, hemangiopericytoma, lymphoma, schwannoma, leiomyoma, angiofibroma, fibromatosis, malignant fibrous histiocytoma, and fibrosarcoma. Reliable and accurate diagnostic criteria for SFTs are necessary to avoid overdiagnosis or confusion with more aggressive neoplasms in these locations.

CT findings are that of a smooth expansile soft tissue mass with surrounding bony remodeling or erosion from pressure effect. SFTs show some characteristic magnetic resonance imaging findings including a well-circumscribed solid mass that is hypo- to isointense on T1-weighted images and has a prominent, but heterogeneous, enhancement with gadolinium. In addition, variable hypo- or, more often, hyperintensity on T2-weighted images is seen³ (Fig. 4).

The major diagnostic feature of these tumors is the histopathologic finding of two alternating fundamental architectural patterns present in various degrees. These patterns consist of hypercellular plump spindle cells haphazardly intermixed in a dense hypocellular collagenous stroma with variable vascularity. These two distinct architectures have been described as “solid spindle type” and “diffuse sclerosing type.”⁴ The tumor cells have a faint eosinophilic cytoplasm, inconspicuous nucleoli, and evenly distributed fine nuclear chromatin. The cells are haphazardly arranged within a dense collagenous, sometimes keloid-like, stroma. Variable, but often prominent, branching vessels are seen within the tumor and may result in some confusion between the diagnosis of SFT and that of a hemangiopericytoma⁵ (Figs. 2 and 3).

Due to the broad differential diagnosis associated with their clinical presentation and histopathologic features, a panel of immunohistochemical markers has become paramount in accurately diagnosing SFTs.

Figure 4 Magnetic resonance scan of head—note characteristic findings of a well-circumscribed solid mass that is hypo- to isointense on T1-weighted images and has a prominent, but heterogeneous, enhancement with gadolinium.

CD34 is a transmembrane glycoprotein found on the surface of hematopoietic progenitor cells. It has recently been considered to be a positive marker for SFT. It should be noted that although CD34 is very sensitive for SFTs, it is not entirely specific for SFTs and is expressed in a variety of spindle-cell neoplasms, such as dermatofibrosarcoma protuberans or neural tumors.⁶ In addition, SFTs are strongly positive for vimentin and uniformly negative for keratin, desmin, and S100 protein. Although not specific for SFTs, the addition of Bcl-2 to the panel of immunostains may aid in diagnosis, as Bcl-2 is frequently positive in SFTs but not, for example, in hemangiopericytomas.⁷ In the present case series, histological analysis revealed a benign spindle cell lesion with positive immunohistochemical staining for CD34, vimentin, and bcl2. All specimens stained negative for S-100, desmin, and actin. These histological findings supported the diagnosis of SFT in each case.

The clinical behavior of pleural SFTs is unpredictable. Approximately 80 to 88% of pleural SFTs behave in a benign fashion and are cured with surgical excision. In contrast, the remaining 12 to 20% of the pleural SFTs are malignant and are associated with local invasion, recurrence, intrathoracic spread, and distant metastasis. These rare cases of malignancy are associated with hypercellularity, increased mitosis (> 4 mitoses/10 high-power field), pleomorphism/nuclear atypia, and necrosis. These features have been described in 5 to 10% of extrathoracic SFTs and have been associated with aggressive clinical behavior including local recurrence and distant metastasis.⁷ Understandably, much attention has been concentrated on the

incidence of aggressive lesions and the features that may allow clinicians and pathologists to predict this malignant behavior. In the literature, there has only been one reported case of a malignant SFT of the nasal cavity that invaded intracranially and extended into the orbit.⁸ It has therefore been quite difficult to ascertain the features that predictably herald poor outcomes, as most behave in an indolent fashion. Although the histological features mentioned above are considered noteworthy in terms of the ascertaining the potential for disease progression, the work of England et al and Witkin and Rosai concludes that resectability is the most important prognostic factor in the treatment of SFTs.^{9,10}

Surgery represents the treatment of choice for SFTs, although radiotherapy and chemotherapy have been used in the past. In fact, Goodlad and Fletcher showed a case of mediastinal SFT that was treated with radiotherapy alone, and the patient was free from recurrence 19 years later.¹¹ All of the published reports of nasal SFTs were treated with surgical resection, although two patients underwent adjuvant radiation and one patient underwent embolization of a persistent nasopharyngeal remnant.¹² The role of radiotherapy has been explored sporadically, but its benefit remains unproven and its appropriate use poorly understood.

Endoscopic endonasal surgery is ideally suited for the resection of benign, encapsulated lesions such as SFTs. An endoscopic approach permits a wide and mobile panoramic view, good visualization, and magnification. This type of treatment also has the advantages of no external incision, less blood loss, low postoperative morbidity, and shorter hospital stay.¹³ One of the challenges of an endoscopic approach occurs in the setting of sizable tumors and/or bleeding that may obscure visualization and lead to incomplete resection. This is particularly worth mentioning in the treatment of SFTs because of the potential for prominent vascularity and the fact that outcome is inherently tied to "resectability." As such, endoscopic resections of these tumors should only be considered when the size of these tumors allows for adequate visualization and the surgeon has the equipment and experience to tackle potentially brisk bleeding endoscopically.

There has been only one previous report of recurrence of an SFT in the upper respiratory tract. In other sites, the recurrence of SFTs has been noted up to 31 years after surgery.¹⁴ Incomplete resection of SFTs stands out as the most important factor in determining long-term outcome. This remains an issue in this case series as follow-up was relatively short, at 26, 35, and 49 months following resection. As such, these patients need continued follow-up and assessment. Accurate prognosis of extrapleural tumors and their clinical behavior, specifically those of the nose and paranasal sinuses, cannot be confidently described due to the short

duration of follow-up and low number of cases reported so far. As such, we would recommend long-term clinical and imaging follow-up, especially when there is any question of residual disease.

CONCLUSION

Sinonasal SFTs are rare. Clinically, they manifest themselves as benign nasal tumors. This diagnosis should be considered in the differential of all expanding soft tissue masses in the nasal and paranasal cavities. The diagnosis of these tumors is based on the histopathologic findings of spindle cells arranged in a characteristic "patternless" pattern—two fundamental architectural patterns present in various degrees: hypercellular spindle-cell areas and diffuse hypocellular sclerotic areas. Immunohistochemical analysis, most notably positive staining with CD34 and bcl-2, is vital to definitive diagnosis. Local excision is the first line of treatment. Endoscopic resection is a reliable treatment modality if the surgeon has experience in advanced endoscopic techniques. At present, long follow-up is recommended because of the unpredictable behavior.

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