INTRODUCTION

Solitary fibrous tumor (SFT) is a rare neoplasm also known as benign fibrous mesothelioma or submesothelial fibroma. It was first described as a primary spindle cell tumor of the pleura by Klemperer and Rabin in 1931.\(^1\)\(^2\) SFTs were initially thought to be originated from mesothelium, but later studies showed them to arise from mesenchyme.\(^3\) SFTs mostly arise from the pleura, but they are also found in various extra-pleural sites including the abdomen, extremities, and vulva.\(^4\)

SFTs in the head and neck area have been reported in several studies, but occurrence in nasal cavity and paranasal sinus is still extremely rare.\(^4\) The patients are usually presented with well-defined soft tissue mass initially causing no symptoms but later resulting in various symptoms such as nasal obstruction, epistaxis, hyposmia, and facial pain due to the mass effect.\(^5\) For the diagnosis of SFT, computed tomography (CT), magnetic resonance imaging (MRI), and biopsy for pathologic results with immunohistochemical stains are usually performed. Once the SFTs are diagnosed, complete surgical excision is the treatment of choice. In this case, we report a huge SFT originated from the frontal recess and extended to nasal cavity, which was successfully excised by transnasal endoscopic tumor removal.

CASE REPORT

A 32-year-old male patient with chief complaint of headache and left facial pain visited the outpatient clinic which started several months ago. He also reported left-sided, persistent nasal obstruction that had been aggravated pro-

---

Somi Ryu, MD\(^1\), Sang Yun Lee\(^1\), Sang-Wook Kim, MD, PhD\(^1\)\(^2\), and Hyun-Jin Cho, MD, PhD\(^1\)\(^2\)

\(^1\)Department of Otorhinolaryngology, Gyeongsang National University School of Medicine, Gyeongsang National University Hospital, Jinju; and \(^2\)Institute of Health Sciences, Gyeongsang National University, Jinju, Korea

Received: January 17, 2021 / Revised: March 6, 2021 / Accepted: March 15, 2021

Address for correspondence: Hyun-Jin Cho, MD, PhD, Department of Otorhinolaryngology, Gyeongsang National University School of Medicine, Gyeongsang National University Hospital, 79 Gangnam-ro, Jinju 52727, Korea
Tel: +82-55-750-8935, Fax: +82-55-759-0613, E-mail: goma0320@gmail.com
gressively, but otherwise he was in good general condition without other medical issues. A smooth, round, and pale mass occupying the entire left nasal cavity and nasopharynx was found in rigid nasal endoscopy (Fig. 1). The laboratory findings and ophthalmological examination were also conducted, and there were no significant clinical findings, except mild left exophthalmos. He was further evaluated with CT and MRI of paranasal sinuses. The image findings revealed a huge, expansile, and heterogeneously enhancing mass with highly vascular nature in left nasal cavity, maxillary sinus, ethmoid sinus and both frontal sinuses, and it was extended to left orbit (Fig. 2).

Incisional biopsy in operating room under general anesthesia revealed spindle cell proliferated tumor with moderate cellularity, mild pleomorphism, and increased capillaries. The pathologic reports were consistent with low grade mesenchymal tumor, such as SFT, and there was no evidence of malignancy. After the reports were confirmed, the 2nd operation, tumor removal via transnasal endoscopic approach under a general anesthesia, was performed at a week after the primary incisional biopsy. Highly fibrotic and hemorrhagic tumor adhesive to diffuse nasal cavity mucosa was found, and a focal hyperostosis in the frontal recess was suspicious for the origin site (Fig. 2B). Several arterial bleedings occurred during the surgery, but they were successfully controlled by electrocoagulation. Gross total tumor removal was established without additional external incision or exposure of crucial structures around the lesion (e.g., skull base and the orbit).

Preoperative and postoperative CT images revealed successful excision of the tumor by transnasal endoscopic surgery (Fig. 3). The pathology results of main tumor were consistent with SFT; the spindle cells were stained positively for CD34 and negatively for both S-100 protein (neural marker) and smooth muscle actin. One mitosis per 10

---

Fig. 1. A smooth white mass occupying the entire left nasal cavity found in rigid nasal endoscopy.

Fig. 2. A huge heterogeneous enhancing mass with high vascularity in left nasal cavity, maxillary sinus, ethmoid sinus, and both frontal sinuses was found in paranasal sinus CT (A, B) and T1-weighted, T2-weighted and T1-enhanced MRI (C, D, E). The suspicious origin site was frontal recess, and it was labeled by an arrow.
high-power fields (HPFs) was found in the excised specimen (Fig. 4). Upon follow-up, the postoperative course was uneventful with no evidence of recurrence for 6 months.

DISCUSSION

SFTs are relatively rare tumors originating from mesenchymal spindle cells. They most commonly occur from the pleura, but various extra-pleural sites of tumor origin have been reported. SFTs in the head and neck area are well described in several studies, but involvement of the nasal cavity and paranasal sinuses have been still rarely reported. Up to 20% of SFTs have been reported as malignant neoplasms, and SFTs found in extra-pleural sites including nasal cavity and paranasal sinus tend to be predominantly benign while pleural tumors are more aggressive. Patients with sinonasal SFTs generally visit clinics for not only slowly growing, painless mass lesion but also nasal obstruction, rhinorrhea, and intermittent epistaxis, if symptomatic.

In terms of the morphologic characteristics, SFTs are usually encapsulated and fibrous mass with rich vascularities, as found in this case. Ahn et al. reported that preoperative angiographic embolization could be helpful for treating sinonasal SFT due to their high vascularity and bleeding risk. Through consultation with the radiologic specialist with the contrast enhanced and high-resolution CT, we planned the surgery without embolization as there was no dominant, single, and targetable feeding artery of the tumor.

For the diagnostic process, radiologic examinations with CT and MRI are important. The tumors usually show homogeneous density compared with gray matter in non-contrast CT, while they are well enhanced in contrast enhanced CT and MRI. Also, they show the iso to hypo signal intensity on T2-weighted MRI that is useful finding to make a diagnosis of SFTs. It is very important to differentiate SFTs through the radiologic images from other spindle-cell neoplasms including angiofibroma, fibrous histiocytoma, schwannoma and fibromatosis, since punch biopsy of the SFTs might be very dangerous due to their rich vascularity.

Histologically, the tumor is composed of spindle cells arranged without typical patterns in collagenous background. In the aspect of immunohistochemical staining, the majority of SFTs are positive for CD34, CD99, and Bcl-2 protein while they are negative for S-100 protein (neural marker), epithelial membrane antigen, and keratin. Even though CD34, a type of transmembrane phosphoglycoprotein, is not entirely specific for SFTs, the expression of CD34
in conjunction with the lack of other markers is definitely helpful to find SFTs. In our case, immunohistochemical assessment was positive for CD34 but negative for S-100 and smooth muscle actin. Additionally, there was only 1 mitosis per 10 HPFs in our specimen. For the diagnosis of the malignant SFT, more than 4 mitoses per 10 HPFs should be identified by pathologic exam.

The treatment of choice of benign SFTs is complete surgical resection, even though radiotherapy and chemotherapy have been also adapted. A number of surgical methods such as lateral rhinotomy and maxillectomy have been reported, but transnasal endoscopic resection is widely performed these days to treat paranasal SFTs. It might be meaningful that our case of paranasal SFT, not only relatively large (7 × 7.5 cm in CT and MRI findings) but also destructing frontal sinus and orbit, was successfully treated by transnasal endoscopic resection without external approaches.

Therefore, transnasal endoscopic approach can be useful to completely resect large paranasal SFTs without additional external incision. However, cautious preoperative assessment with radiologic images should be performed for evaluating the vascularity and location of the tumor.

REFERENCES