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A Rare Tumor of the Retropharyngeal Area: Solitary Fibrous Tumor

Case Report

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Abstract

We describe a case of retropharyngeal solitary fibrous tumor (SFT) in a 46-year-old female patient. Total removal of the tumor without any complications was performed by transoral surgery. Intraoral resection of retropharyngeal SFT can be preoperatively performed by meticulous assessment of the extent and vasculature of the tumor with radiological guidance.

Keywords: Pharynx neoplasms, solitary fibrous tumor, surgery, oral

Introduction

A solitary fibrous tumor (SFT) is a rare, benign, mesenchymal neoplasm that was first described in the pleura, in 1931 (1). Since 1931, it has been found in almost every organ, including the head and neck region. However, the presentation of SFT in the retropharyngeal area is extremely rare, with very few reports in English literature (2, 3).

Case Report

A 46-year-old female presented with a two-year history of mild inspiratory dyspnea and progressive dysphagia. The symptoms worsened in the last six months. Physical and endoscopic examination of the patient revealed a large, submucosal, and smooth-surfaced mass on the posterior pharyngeal wall with partial obstruction of the pharyngeal inlet (Figure 1). Doppler ultrasonography of the neck revealed an 88 (craniocaudal)×40 (anteroposterior)-mm solid, hypoechoic, well-circumscribed retropharyngeal mass without marked vascularity. No lymphadenopathy with pathological configuration was detected by Doppler ultrasonography. Magnetic resonance imaging (MRI) revealed a 10×64-cm well-circumscribed, submucosal, and retropharyngeal mass extending from the oropharynx to the level of the first tracheal ring with left-side predominance. The mass was hypointense on T1- and heterogeneously hyperintense on T2-weighted images (Figure 2a, b). Diffuse and prominent enhancement with gadolinium was also noted within the mass.

After a meticulous physical and radiological examination, total transoral removal of the tumor was planned under general anesthesia with written informed consent obtained from the patient to access the transcervical approach if necessary. Preoperative biopsy was not performed because of the possible increase in dyspnea because of edema. An inverted U-shaped pharyngeal mucosal incision was applied to access the mass. The tumor was apparently encapsulated and smooth-surfaced with a rubbery consistency. It was not adherent to adjacent structures but was too large to be removed in "en bloc" fashion with direct visualization; hence, piecemeal removal was adopted. Complete intraoral removal of the tumor was successfully performed without any complication, and macroscopically, the entire mass was a 9×4×3-cm multi-lobulated solid tumor with a whitish-grey color.

Microscopically, the tumor had a patternless pattern with proliferation of bland spindle and ovoid cells, accompanied by a collagenous stroma with branching hemangiopericytoma-like vessels (Figure 3a). Immunohistochemically, the neoplastic cells were strongly and diffusely positive for CD34 (Figure 3b), vimentin (Figure 3c), and Bcl-2 and were less than 1% positive for Ki67. These diverse histological and immunohistochemical findings resulted in a diagnosis of SFT. The patient had no signs of recurrence one year after the surgery in endoscopic and MRI examinations. Written informed consent was obtained from the patient.

Discussion

In the head and neck region, SFT was first described by Witkin and Rosai in 1991 (4). To date, a number of different sites of the head and neck have been reported, with a predominance in the oral cavity and sinonasal tract (5-7). SFTs usually constitute a slow-growing, mobile, submucosal, non-tender, and painless lesion in the head–neck region. As a slow-growing painless mass, SFT can reach a considerable size before presentation. In

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the reported retropharyngeal cases, SFT usually presents with symptoms of dysphagia and dyspnea because of obstruction of the nasopharynx and oropharynx (2, 3). In this case, the lesion caused mild inspiratory dyspnea and progressive dysphagia, which dramatically improved after surgical removal.

Radiological studies have a critical role in ensuring an appropriate treatment plan for these rare tumors, although there are no absolutely distinctive radiological findings. A smooth soft-tissue mass with adjacent expansile bone and cartilage remodeling can be demonstrated on computed tomography (CT) scans, depending on the location. The MRI findings of SFTs show a well-circumscribed, solid mass, which is hypo- to iso-intense on T1-weighted images with variable hypointense or more frequently, hyperintense on T2-weighted images (8). Although intense contrast enhancement on CT and MRI is the most consistent feature, variable enhancements due to scattered collagen and fibroblast components of the tumor have been reported (9). In the present case, the mass was hypointense on T1- and heterogeneous-hyperintense on T2-weighted images in the MRI findings. Diffuse and prominent enhancement with gadolinium was also noted within the mass.

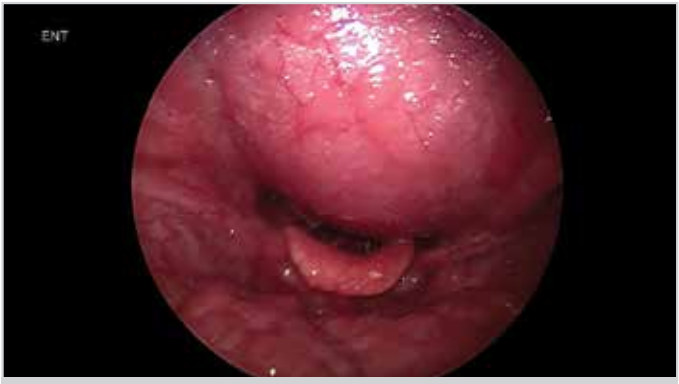

Figure 1. Endoscopic examination revealed a large, submucosal, and smooth-surfaced mass on the posterior pharyngeal wall

The differential diagnosis of SFT includes fibrosarcomas, solitary myofibromas, metastatic malign mesotheliomas, synovial sarcomas, hemangiopericytomas, neurofibromas, and benign and malignant nerve sheath tumors (7). Although preoperative fine-needle aspiration biopsy or image-guided biopsy may be helpful for diagnosis, the definitive diagnosis of SFT can only be made by histological examination and immunohistochemical studies of the surgical specimen. In our case, preoperative biopsy was not performed because of the possible increase in dyspnea due to edema. The microscopic findings of SFT include the proliferation of ovoid or spindle-shaped cells randomly distributed in a patternless-pattern within a collagenous stroma of variable vascularity. SFT is characteristically positive for vimentin and CD34 but is generally negative for neural, melanoma, and muscle markers and cytokeratins (3). SFTs are usually considered to be benign neoplasms. Malignancy can occur in up to 10% of extrapleural SFTs. The histological features of malignant lesions are marked hypercellularity pleomorphism and greater than four mitoses per 10 high-power fields. A large tumor size is another indicator of the aggressiveness of the tumor (10). Metastasis to the lungs, liver, and bone has been reported; however, it is ex-

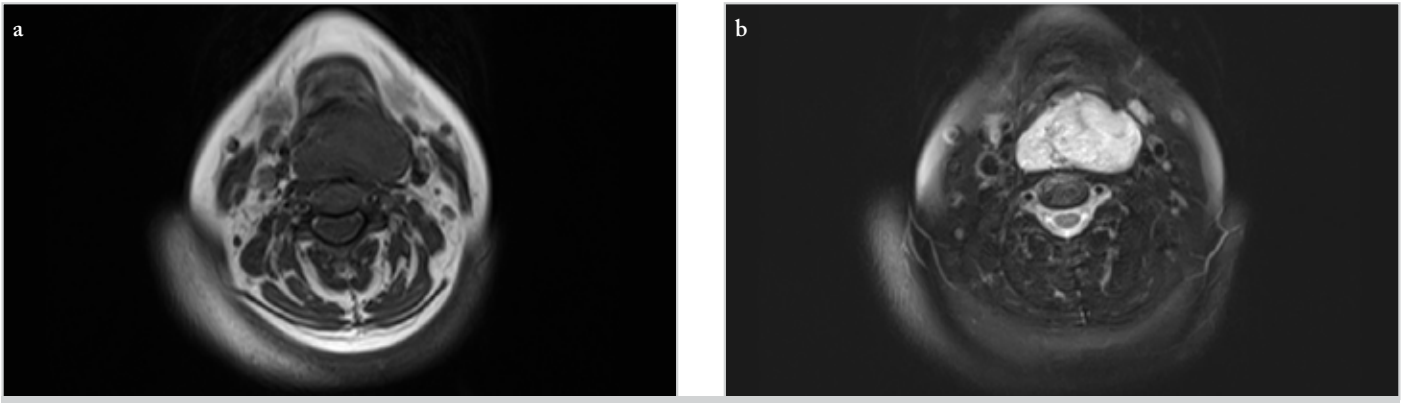

Figure 2. a, b. Axial fat-suppressed T2-weighted MRI (a) Sagittal fat-suppressed T2-weighted MRI (b)

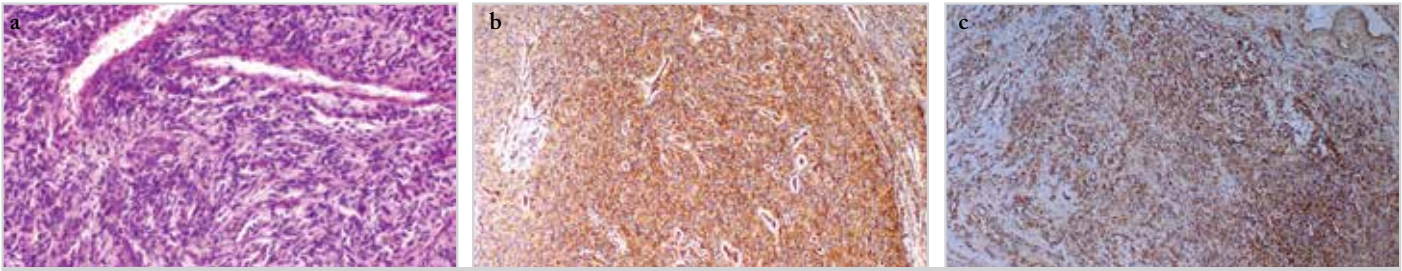

Figure 3. a-c. Higher magnification microphotograph demonstrating spindle and ovoid cells, without atypia and the presence of thin branching vessels within the tumor (hematoxylin-eosin stain, original magnification x200) (a). Positive immunohistochemical staining of the neoplastic cells for CD34 (original magnification x100) (b). Positive immunohistochemical staining of the neoplastic cells for vimentin (original magnification x100) (c).

ceptionally rare (11). No metastasis or malignant feature was detected in the present case.

A surgical approach with the guidance of imaging studies is the first choice of therapy, aas in previously reported cases of retropharyngeal SFT (2, 3). Right cervicotomy along the anterior border of the sternocleidomastoid muscle and transoral excision via a soft palate split approach have been used to remove the lesions in previously reported cases in this area (2, 3). Transoral excision of the mass without soft palate splitting was performed in the present case. The dense nature of the tumor with fibrotic coverage and well-defined boundaries facilitated the complete removal of the present tumor via the transoral approach. Radiotherapy and chemotherapy are not indicated in the treatment of SFT as long as total surgical resection can be performed (10). Long-term clinical follow-up is required because there are previously reported recurrent cases with malignant characteristics, even up to six years after surgical removal (12). In the present case, the patient did not require any adjuvant therapy because of total surgical removal of the tumor and absence of malignant features in the histopathological examination. No signs of recurrence were encountered in the one-year follow-up.

Conclusion

Intraoral resection of retropharyngeal SFT can be performed after meticulous preoperative assessment of the extent and vasculature of the tumor with radiological guidance.

Informed Consent: Written informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

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