Transoral Excision of pre-styloid parapharyngeal space Solitary Fibrous Tumor: A case report and review of the literature

Case Report

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ABSTRACT

Solitary Fibrous Tumors (SFTs) are relatively rare benign neoplasms that commonly occur in the pleura and barely reported in parapharyngeal space. SFT is often misdiagnosed due to its microscopic resemblance to several other spindle cell tumors; however, there are specific diagnostic features on MRI and immunohistochemical.

We herein report a case of a 65-year-old male patient with SFT arisen in the parapharyngeal space completely resected through a transoral approach. Also, a comprehensive review of previously published cases and case series of parapharyngeal space SFTs and hemangiopericytoma (HPC) reported globally in English up to August 2018 using the PubMed/Medline databases was presented. In addition to the current case, there are thirteen SFTs and thirteen HPCs reported cases. All cases were presented and analyzed. Males are predominantly affected. Actually, 23% of HPCs reported cases presented with the histological indicator of aggressive behavior while none of SFTs reported cases. Thus, Parapharyngeal space SFT presented herein is the first case that had been completely resected using the transoral technique. Hemangiopericytoma is a cellular form SFTs; it seems to behave more aggressively than SFTs from studying the previously reported cases.

Key Words: Hemangiopericytoma, immunohistochemistry, parapharyngeal space, solitary fibrous tumor, transoral approach

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INTRODUCTION

Solitary Fibrous Tumors (SFTs) are well-characterized spindle cell tumors first described in the pleura by Klemperer and Rabin in 1931[1]. SFTs are a heterogeneous group of tumors with a highly variable appearance. The SFTs are now considered ubiquitous that have been identified in many extra-pleural sites[2]. Pleural SFTs account for about 30% of all cases followed by meninges of about 27%. The most common extra-pleural and extra-meningeal locations include abdominal cavity in 20 %, trunks in 10 %, extremities in 8%, and neck in 5 %[3]. Head and neck SFTs are rare, and parapharyngeal space is even rarer. The exceedingly rarity of reported literature in this particular location made them not commonly considered in the differential diagnosis of parapharyngeal space soft tissue mass. Moreover, it is often misdiagnosed due to its microscopic resemblance to several other spindle cell tumors[4]. Hence it remains a diagnostic challenge with controversial biological behavior that requires an integrated approach including specific radiological, histological, immunohistochemical and even molecular findings.

Herein, we present a case of SFT located in the left pre-styloid parapharyngeal space that successfully excised by transoral approach. Also, a comprehensive review of previous published cases and case series of parapharyngeal space SFTs and hemangiopericytoma (HPC) are presented. In fact, the SFT and HPC tumors are commonly considered the same entity and almost indistinguishable.

MATERIALS AND METHODS:

This study complies with regional and institutional ethical guidelines and with declaration of Helsinki. A written informed consent was obtained from our case for his participation.

A comprehensive review via Medline, Pubmed, LILACS, SciELO, and Cochrane Library databases research using the keywords "Parapharyngeal -Solitary - Fibrous - Hemangiopericytoma -tumor" was conducted. English articles with a complete data and a confirmed histopathlogical diagnosis were only included. Careful studying for the histopathlogical case data was conducted to definitely identify if the case is SFTs or Hemangiopericytoma. Patients' data, clinical presentations, tumor size, tumor site, histopathological criteria, surgical procedures, complications, and history of recurrence data were collected and analyzed.

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Case Report:

A 65-year-old male patient presented to our clinic with a history of six months slowly progressive foreign body sensation in the throat, dysphagia, snoring, obstructive sleep apnea, and muffled voice. He is known to be hypertensive and diabetic type II for more than five years on oral medications.

In the oropharyngeal examination, a diffuse bulge was seen on the left lateral pharyngeal wall pushing the left tonsil downwards and medially. It extends into the soft palate and down to the level of the tip of epiglottis with normal mucosal covering. On palpation, swelling was firm, not tender; not bleed with manipulation. Head and neck examinations otherwise were normal. The patient is not a smoker with no significant past medical or family history.

Contrast-enhanced computed tomography (CT) scans showed a well-defined heterogeneous enhanced mass, involving the left parapharyngeal space with no infiltration of adjacent tissue and partial obliteration of oropharyngeal airway. Magnetic resonance imaging (MRI) revealed that the mass was uniform isointense in T1, heterogeneously enhancing hyperintense with central area of low signal on T2 and enhanced after intravenous administration of gadolinium DTPA (Figure 1a–c). The mass measure was approximately 8.5 × 4.3 cm.

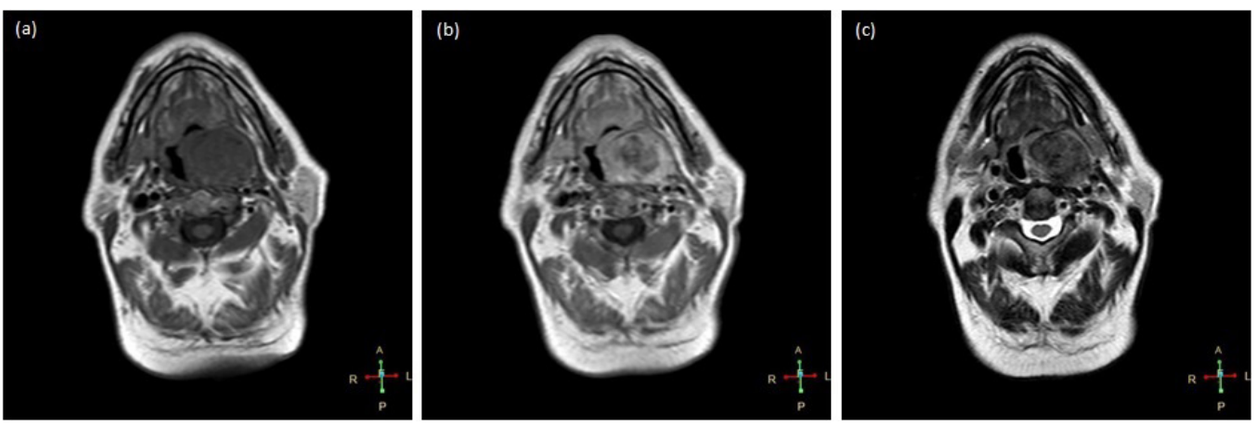

Fig. 1: Preoperative Axial MRI (a) T1W isointense PPS mass. (b) T1W with contrast. (c) Axial T2W showing heterogeneously enhancing hyperintense with central area of low signal of the Solitary Fibrous Tumor in parapharyngeal space.

Transoral biopsy under local anesthesia was taken. A single fibrous tumor was a diagnosis which confirmed by immunohistochemistry (IHC) (positive for CD 34, Bcl2 and negative for pancytokeratin, S100, B catenin, SMA, and P63) (Figure 2). Surgical excision was planned through transoral endoscopic guided approach.

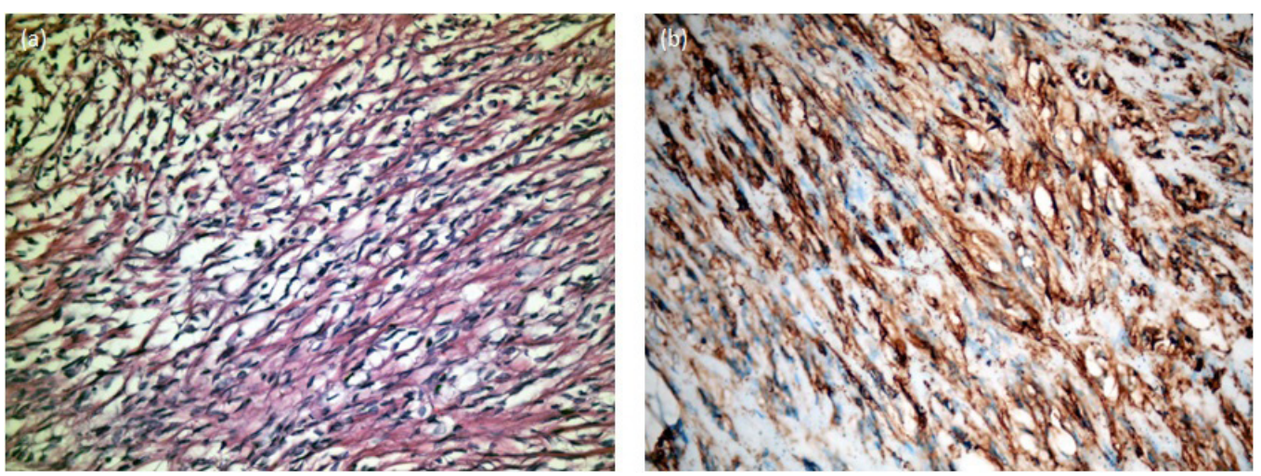

Fig. 2: (a) Histopathology H&E; X400 magnification showing a cellular spindle cell proliferation, with absent mitosis and necrosis. The cells have moderate to abundant esinophilic cytoplasm and separated by collagenous stroma. Scale bar = 100 µm (b) Immunohistochemistry X100 magnification showing diffusely and strongly positive staining for CD34. Scale bar = 50 µm

Transoral approach Technique:

The patient was intubated through a wakeful flexible fiberoptic nasotracheal intubation. He was placed in Rose's position. Oral cavity and oropharynx exposure were achieved using Boyle-Davis mouth gag and Cheek and Lip Retractors. An incision was fashioned to extend from the posterior edge of the hard palate, passing along the lateral edge of the soft palate and into the oropharynx. The mucosa, submucosa and superior constrictor muscle were sequentially divided with unipolar cutting electrocautery. The tumor popped out in view with its glistening grayish white capsule with gentle blunt dissection assisted with 0 and 30 rigid endoscopes. It was enucleated from its surrounding tissues and delivered into oral cavity as a whole. Then, the pharyngeal wall and soft palate was repaired primarily in three layers using interrupted Vicryl 3-0 suture (Figure 3). Macroscopically, the tumor was well encapsulated and had firm and yellowish white cut surface. Immunohistochemistry (IHC) confirmed the diagnosis of SFT.

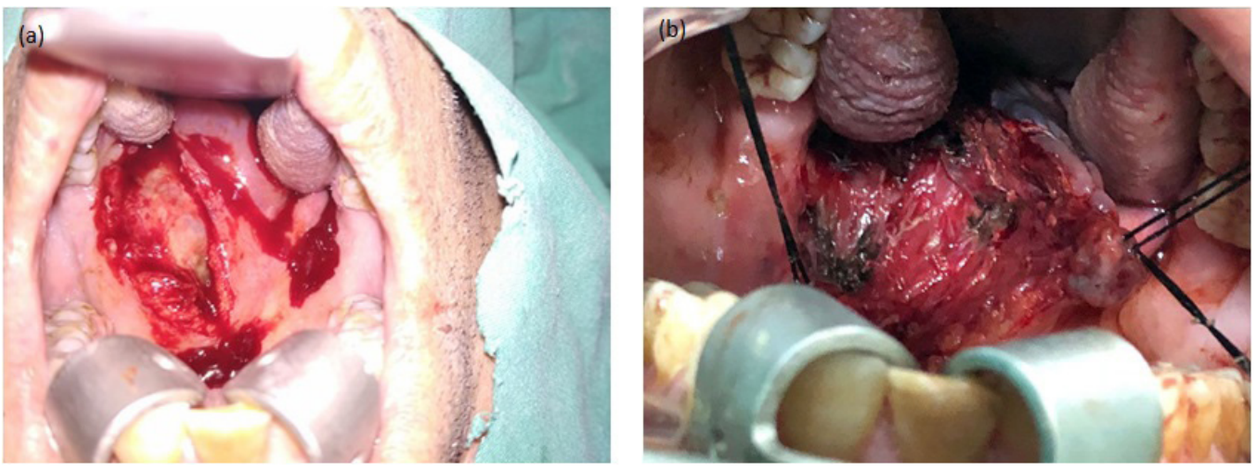

Fig. 3: Intraoperative view (a): showing palatopharyngeal incision with the mass popped out in view, (b) After complete excision of the mass.

Postoperatively, there were no complaints with respect to swallowing, breathing and speech. The patient was discharged home after three days of surgery. Now the patient is on more than nine months of follow-up; his symptoms have been completely resolved with no evidence of recurrence.

RESULTS

A total of 40 (17 SFT and 23 HPC) articles published with 41 (18 SFTs and 23 HPC) cases until August 2018. However, only 25 (12 SFT and 13 HPC) cases fulfilled the inclusion criteria with the present case. All cases are summarized in (Table 1).

Painless swelling, dysphagia, snoring, obstructive sleep apnea, F.B sensation and voice change were the most mentioned clinical presentations. The size range of tumor was 2.5 to 12 cm in its maximum diameter while all cases of HPC tumor size as above 10 cm. The left side was commonly affected than right side. None of the SFT s reported cases showed a histological indicator of aggressive behavior or history of recurrence. While 23% of HPC total cases reported to show Aggressive histopathological behavior features.

Except for the current case, the reported SFTs and HPC tumors were excised through trans-cervical approach. The current case is the first case in literature to be totally excised trans-orally. Postoperative adjuvant radiotherapy has been used only with one SFT s case and four HPC cases.

DISCUSSION:

SFTs are rare, usually benign, spindle cell neoplasms. Before the widespread acceptance of the term SFTs, there were varieties in nomenclature of the pleural SFTs which are a reflection of the presumed histologic origin of the deranged cells, spectrum of structural and ultra-structural features of this tumor and the broad differential diagnosis[5]. The WHO (2013) classification abandons the entity HPC and categorizes extrapleural SFT within the group of fibroblastic/ myofibroblastic tumors of intermediate biologic potential with intermediate -rarely metastasizing - biological behavior[3].

A total of 25 (12 SFT and 13 HPC) cases fulfilled the inclusion criteria including the present case are summarized in (Table 1)[4, 6-16]. Similar to other extrapleural SFT; the age range of the reported cases was 19-77 years with a peak of incidence in the fourth and sixth decades with an average age of 49.4 and it is rarely reported in children and adolescents. Similar to other extrapleural SFTs, a male predominance was reported for Parapharyngeal SFTs[3].

Table 1: Analysis of 26 (13 SFT and 13 HPC) cases found in the literature - including our case.

	SFT	HPC
< 20 ys	1 (7.7 %)	1 (7.7 %)
20-40 ys	3 (23 %)	5 (38.5%)
41-60 ys	5 (38.5%)	4 (30.8%)
> 61 ys	4 (30.8 %)	3 (23%)
Average age:	49.4	45.7
Sex:		
Male	9 (69.2%)	8 (61.5)
Female	4 (30.8%)	5 (38.5%)
Site:		
Right	4 (30.8%)	6 (46.2%)
Left	9 (69.2%)	7 (53.8)
Main Complaint:		
Swelling	5 (38.5%)	6 (46.2%)
Dysphagia	4 (30.8%)	3 (23%)
Snoring and OSA	3 (23%)	1 (7.7%)
Change of voice	2 (15.4%)	1 (7.7%)
Pain	2 (15.4%)	2 (15.4%0
Accidentally discovered	2 (15.4%)	1 (7.7%)
F.B sensation	2 (15.4%)	3 (23%)
Size:		
<10 cm	10 (77%)	13 (100%)
>10 cm	3 (23%)	0
Surgical Approach:		
Transcervical	12 (92.3%)	13 (100%)
Mandibulotomy	4 (30.8%)	_
Trans-parotid	2 (15.4%)	4
Transoral (our case)	1 (7.7%)	_
Tracheostomy	1 (7.7%)	1 (7.7%)
Aggressive histopathological behavior features:	Non	3 (23%)
Postoperative Radiation	1 (7,7 %)	4 (30.8%)

The preoperative SFT diagnosis remains a difficult task, although some specific diagnostic features on MRI and Immunohistochemical study have been suggested. The reported MRI findings of SFTs include an isointense signal on T1-weighted images, the signal intensity increased with intravenous gadolinium and it was variable on the T2-weighted images. It has been suggested that this variable signal intensity corresponds well to the histologic findings and the differences in the main components of the tumor (the amount of collagen and fibroblasts), as well as the presence of the degeneration. The mature fibrous tissue usually has lower signal intensity on T1- and T2-weighted images, and this is related to the area of hypocellularity and abundant collagen in the stroma. Intense enhancement is generally due to its high vascularity[17, 18]. On the other hand, characteristic Immunohistochemical of SFTs is the strong positive reaction with CD34 and vimentin. A negative immunoreactive reaction with epithelial, vascular, neural crest and muscle markers (cytokeratin, CD31, CD21, CD35, S-100, desmin SMA and EMA) would, in turn, exclude other differential diagnoses such as epithelial tumor, fibrosarcoma, and neurogenic tumor. In fact, a reliable diagnosis of SFTs could be confirmed by immunohistochemical and MRI[8, 17, and 19].

Recent research on the biology of hemangiopericytoma (HPC) tumor leads to question the traditional separation between HPC and SFTs as a two distinct entities. In fact, this separation has been resulted from the difference in their histological grading (cellularity and mitotic index). Unlike SFT fibrous form characterized by hyalinized, thick-walled vessels with opened lumina and strong CD34 reactivity, the cellular form representing a conventional HPC is characterized by staghorn-branching vascular pattern, thin-walled vessels and focal or absent CD34 reactivity[3, 6].

The clinical behavior of SFTs is unpredictable. Poor prognosis is expected in cases associated with recurrence, local invasion and occasionally distant metastasis (75% of those patients die within 1.8 to 3.8 years). Careful studying of the previous reported cases of SFT and HPC leads to the conclusion of that the HPC tumors look more aggressive and show history of recurrence in comparison to the SFTs. Some histological appearances could suggest an aggressive behavior and high chances of relapse as a large tumor size (>10 cm), lack of circumscription, hypercellularity, increased mitosis (mitotic index >4/10 HPF), nuclear atypia, infiltrative margins, pleomorphism, and necrosis. However, these features were still shown to be unreliable as SFTs cases with benign microscopic appearances may behave aggressively while others with malignant microscopical appearance can behave benignly. Thus, the prognosis is substantially unpredictable and not strictly dependent on histological features. The most important prognostic factor, however, is the complete resection of the tumor[3,8, 17].

The current case is the first case of parapharyngeal space SFT that successfully resected using trans-oral approach. This approach seems promising as the intraoperative bleeding is less, the postoperative is uneventful, and the patient recovered well with no visible scars.

Postoperative adjuvant radiotherapy has been used in some of the reported cases, its effect remains underestimated. It was reported that the radiotherapy could have a role in larger SFT mass particularly with positive surgical margin after resection[12, 13].

Post-operative follow-up is mandatory since there were some cases reported in the literatures developed recurrent with malignant characteristics especially for HPC, even up 30 years after excision[20, 21]. There were also rare cases with sudden transition from conventional benign-appearing to overtly high-grade sarcoma[22].

CONCLUSION

The current paper presents the first case of parapharyngeal space SFT, which excised completely using transoral approach. Review of literature of previous reported cases of parapharyngeal space SFTs and HPC tumors revealed that Hemangiopericytoma seems to behave more aggressive.

CONFLICT OF INTERESTS

There are no conflicts of interest

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