

# Solitary fibrous tumor of the post-styloid parapharyngeal space

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## Abstract

Solitary fibrous tumor (SFT) is a well-known tumor composed of spindle cells found most commonly in the pleura. Recently, accounts of their rare occurrence at other sites, including the head and neck area, have been reported. The parapharyngeal space is a rare location even for head and neck SFTs, and thus, could be confused with a variety of other tumors that can originate in this area. Here, we report a case of SFT originating from the post-styloid parapharyngeal space and discuss the possible differential diagnosis on radiographic findings.

## Keywords

Head and neck, neoplasm, parapharyngeal space, solitary fibrous tumor

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## Introduction

Solitary fibrous tumor (SFT) is a rare, but well-known submesothelial-origin tumor composed mainly of relatively uniform spindle cells in collagenous stroma. Recent studies have shown that SFTs can originate in various parts of the body, although described most often in the pleura. Most of these tumors are clinically and histologically benign. However, malignant transformation over long follow-up periods has been reported. These tumors are known to affect mainly middle-aged adults, with no tendency towards a particular sex (1). Due to its rarity in extrapleural spaces, it may be difficult to identify SFT as a differential diagnosis of a soft tissue mass. Thereby, we report here a very rare case of a SFT originating from the post-styloid parapharyngeal space and discuss the radiologic features and its possible differential diagnosis.

## Case report

A 32-year-old woman presented with progressive dull aching pain in the throat of 1 year's duration. She also complained of foreign body sensation in the left oropharyngeal area and gradual aggravation of facial asymmetry. However, she did not have any difficulty swallowing or breathing.

In the oropharyngeal examination, diffuse swelling that occupied the left tonsillar fossa was seen with evidence of uvula deviation to the right. Examination of the neck revealed a non-tender, fixed, palpable mass in the left submandibular area.

Contrast-enhanced computed tomography (CT) scans showed a 4 × 4.5 × 6 cm sized relatively well-defined, dumbbell-shaped mass in the left post-styloid parapharyngeal space, with extension of the mass into the left prevertebral area. The mass was heterogeneously enhanced and had inner cystic changes (Fig. 1a and b). There was no evidence of calcifications on the pre-contrast CT images. The mass was separated from the left parotid gland, but was displacing the left

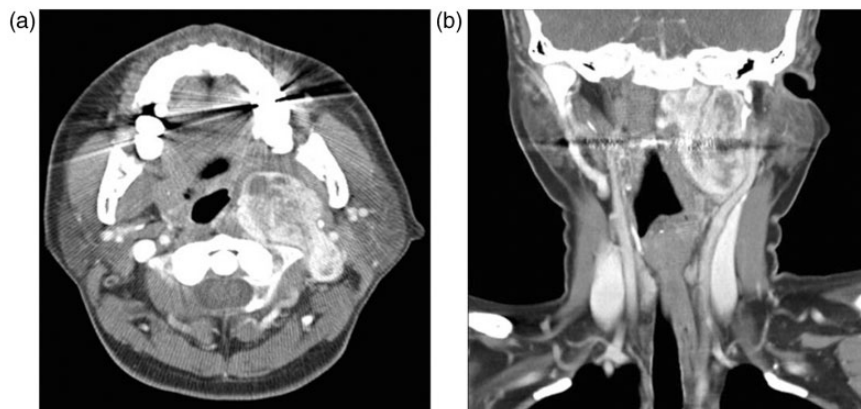
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**Fig. 1.** Contrast-enhanced CT scans of (a) axial and (b) coronal images showing a relatively well-defined, dumbbell-shaped mass in the left post-styloid parapharyngeal space, with extension of the mass into the left prevertebral space. The mass was heterogeneously enhanced and had inner cystic changes. There was no evidence of calcifications in the pre-contrast CT scan (not shown).

internal carotid artery laterally. There were no signs of bony erosion in the adjacent vertebral column.

Magnetic resonance (MR) images revealed that the mass lesion in the left post-styloid parapharyngeal space was iso-intense relative to muscle on T1-weighted (T1W) images (Fig. 2a), and heterogeneously hyperintense relative to muscles on T2-weighted (T2W) images (Fig. 2b). Multiple cystic changes were seen in the mass lesion. The mass was homogeneously and strongly enhanced on contrast-enhanced T1W images (Fig. 2c and d).

The mass was excised, via a trans-cervical approach, below the left submandibular area. The mass was well encapsulated and was easily removed from the post-styloid parapharyngeal space, except for the anterior portion where it showed some adhesion to the surrounding soft tissue. Nevertheless, complete removal was accomplished, successfully avoiding injury to the adjacent structures such as the hypoglossal and vagus nerves.

Microscopic evaluation of the specimen showed a pattern less architecture, consisting of small fusiform cells, randomly arranged between collagen bundles (Fig. 3a). Branching hemangiopericytoma-like vessels with perivascular hyalinization were prominent. The tumor cells were round to spindle-shaped and had little cytoplasm, with indistinct borders and dispersed chromatin within vesicular nuclei. However, focally pleomorphic tumor cells were also seen. Foci of hemorrhage were noted. Mitosis was scarce and necrosis was absent. Immunohistochemically, the tumor cells were CD34, CD99, and BCL2 positive (Fig. 3b), but negative for desmin, actin, cytokeratin, and the S-100 protein. All dissected lymph nodes were free from tumor cells. On the basis of these characteristic findings, the final diagnosis was made as SFT.

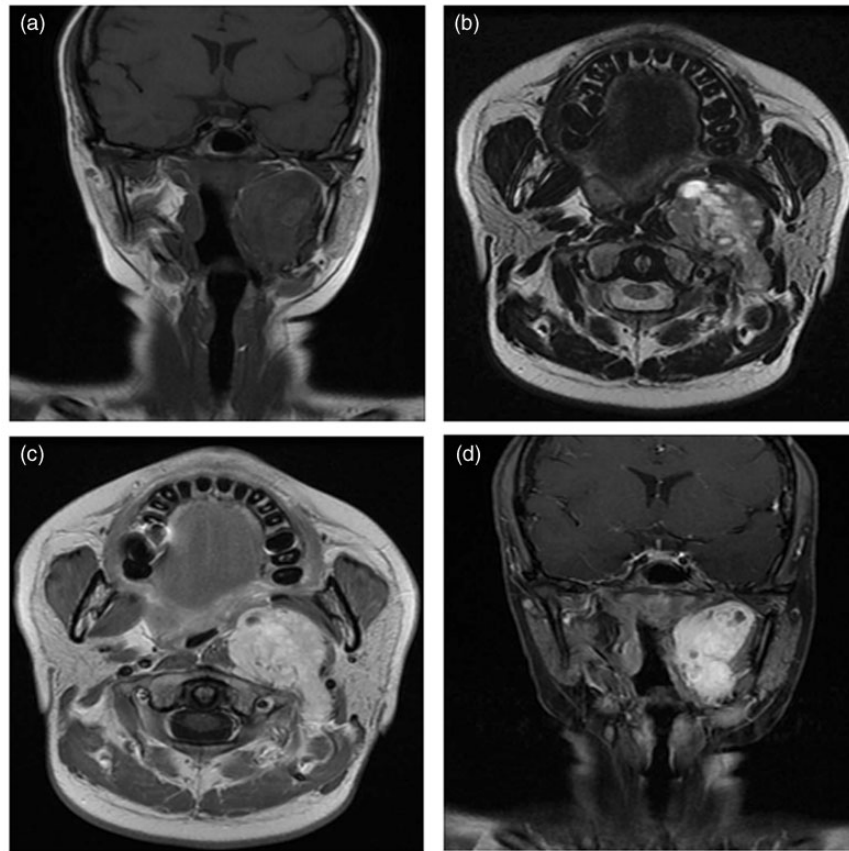
The patient recovered, with a normal postoperative clinical course.

## Discussion

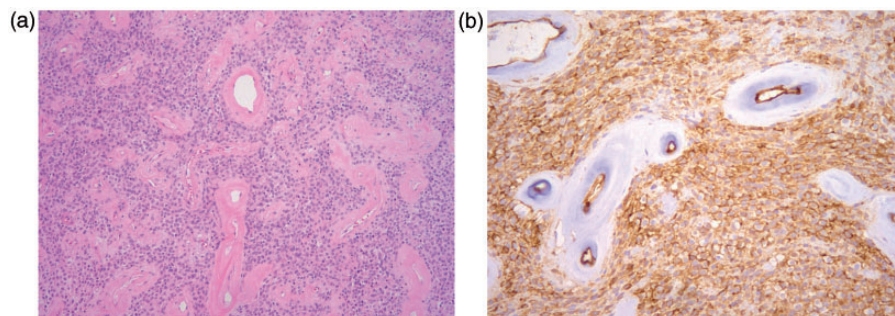
Parapharyngeal space tumors are a rare neoplasm, comprising less than 0.5–1% of tumors of the head and neck. Parapharyngeal space (also termed the lateral pharyngeal space) is divided into two parts by fascial condensations, the pre- and post styloid compartment. Nearly all tumors that arise in the post-styloid parapharyngeal space are neurogenic tumors, paragangliomas, venolymphatic malformation, metastatic masses, and tumors of the minor or ectopic salivary glands. Although extremely rare, wide variety of other cellular lesions has been reported in the post-styloid parapharyngeal space, such as rhabdomyosarcomas and lymphomas (2).

In recent years, a number of reports of SFTs originating in extrapleural sites have been reported. These body areas other than the pleura include the retroperitoneum, the deep soft tissues of the extremities, the abdominal cavity, and various areas of the head and neck. Although rarely found, there have been case reports of SFTs originating in the infratemporal fossa, parapharyngeal space, nasal and paranasal sinuses, soft palate, and epiglottis (3).

SFTs occur with equal frequency among men and women. They are more commonly diagnosed in patients aged 30–64 years, and no risk factors have been identified. Most SFTs are clinically benign; only 12–20% of SFTs are associated with local recurrence, distant metastasis, and histological malignancies, such as hypercellularity, severe nuclear atypia, high mitotic count, tumor necrosis, or infiltrative margins. Symptoms are non-specific and usually absent, which



**Fig. 2.** Left post-styloid parapharyngeal space mass shows iso-intense signal intensity relative to muscles on T1W coronal images (a), and heterogeneously high signal intensity on T2W axial images (b). The mass is strongly enhanced on contrast-enhanced T1W axial (c) and coronal (d) images.



**Fig. 3.** (a) Patternless architecture consisting of small fusiform cells randomly arranged between collagen bundles, separated from each other by thick bands of hyalinized hemangiopericytoma-like vessels. (H&E, 100 $\times$  magnification). (b) Tumor cells show diffuse strong expression of CD34 (200 $\times$  magnification).

may explain why these tumors often grow to a considerable extent before the initial diagnosis. The most common symptoms are due to the mass effect of the tumor itself, as in our case. Paraneoplastic manifestations, such as hypoglycemia and osteoarthropathy, have also been reported (3). En bloc surgery is the recommended treatment of choice in both benign and

malignant SFT, as surgical removal of the tumor will resolve all of these symptoms.

Microscopic findings revealed the characteristic features of SFTs, which appeared to be relatively uniform spindle cells in randomly arranged heavy collagen tissue, in a “patternless architecture”, usually with areas of prominent “hemangiopericytoma-like”

vascularity. Thus, histologically, SFT can be confused with a variety of other soft tissue tumors, such as hemangiopericytoma and Schwannoma. Immunohistochemically, SFT can be differentiated from these tumors by their strong positivity for CD34 and vimentin, while being negative for CD31, cytokeratin, desmin, and the S-100 protein (4).

It has been reported that on MR images, SFTs are usually a well-defined lobulating mass with iso- to low-signal intensity on T1W images, and heterogeneously high signal intensity on T2W images. Cystic and myxoid degeneration causes focal areas of T1 low signal intensity within the tumor. The hypervascularity of the tumor is shown as heterogeneous and intense contrast enhancement in both CT and MR images (5,6). In our case, the differential diagnosis included neurogenic tumors such as Schwannoma, or paraganglioma, hemangioma and lastly, pleomorphic adenoma.

Neurogenic tumors, such as Schwannoma, can occur in the parapharyngeal space. However, primary neurogenic tumor of the parapharyngeal space is extremely rare and they are usually an extension from neurogenic tumors of the carotid space. About half of the neurogenic tumors in this location originate from the vagus nerve; the others originate from the lower cranial nerves or the sympathetic chain. If the tumor originates from the vagus nerve, tumor displaces the ICA anteromedially and the internal jugular vein posterolaterally. If the tumor originates from the lower cranial nerves, neurogenic tumors do not separate them but only displace the carotid vessels posterolaterally. Lastly, if the tumor originates from the sympathetic chain, the carotid sheath will be displaced anterolaterally (7). Thus, differential diagnosis of the parapharyngeal space tumor by the direction of the vessel deviation is fairly difficult. In our case the ICA and the internal jugular vein was both deviated laterally by the tumor. Although hypovascular, Schwannoma is a fairly homogenous, significantly enhancing mass on both CT and MR contrast-enhanced images. Areas of necrosis and hemorrhage can occur.

Paraganglioma is a post-styloid-compartment-originating tumor that can extend into the prestyloid compartment. Paraganglioma originate from paraganglionic cells with significant homogenous contrast enhancement and early washout, due to its highly vascular structure. On MR images, paragangliomas have relatively intermediate signal intensity on all sequences, with a characteristic "salt-and-pepper" appearance. Scattered high T2-signal intensities ("salt") are due to slow flow within the tumor; the T2-signal void areas ("pepper") are due to its prominent vascular structure (7). This characteristic appearance of paraganglioma was absent in our case.

Due to the well enhancing feature of the mass lesion, vascular tumors such as hemangioma can also be

included in the differential diagnosis. Hemangioma is usually a lesion of infancy which mostly affects the head and neck. However, it is also known to occur in adults, although the incidence is very rare. On non-enhanced CT, hemangioma appear as iso-attenuated mass lesions compared to muscles. With contrast enhancement, the lesion show rapid enhancement due to its high flow (8). On MRI, hemangiomas appear as an intermediate T1-signal intensity mass lesion with moderately high T2-signal intensities and intense contrast enhancement (8,9).

Finally, possibility of a pleomorphic adenoma can be considered as a differential diagnosis. It is the most commonly found tumor known that occur in the prestyloid parapharyngeal space, which is also by far the most common primary salivary gland tumor. Malignant salivary gland neoplasms, such as mucoepidermoid carcinoma and adenoid cystic carcinoma, are rare (7). On contrast-enhanced CT images, pleomorphic adenoma is a relatively homogeneously enhancing mass. When larger than 2 cm, multiple foci of low density may occur, due to inner cystic changes or necrosis. On MRI scans, overall the tumors show low-to-intermediate signal intensity on T1W images, and high signal intensity on T2W images (7). Although the overall density and signal intensity of the tumor in our case was favorable for pleomorphic adenoma, the mass was located between the transverse process and the carotid sheath, so the mass was located in post-styloid parapharyngeal space. Therefore, the possibility of a pleomorphic adenoma was low.

In conclusion, SFT is a rare tumor that can involve the head and neck area, which in our case was in the post-styloid parapharyngeal space. The tumor was a well-defined dumbbell-shaped mass, with iso-signal intensity on T1W images and heterogeneously high signal intensity on T2W images. The mass showed significant contrast enhancement on both CT and MRI images. Soft tissue tumors such as Schwannoma or paraganglioma can be thought of as a possible differential diagnosis in a mass arising in the parapharyngeal space. However, when the mass shows CT and MR findings described above, SFT can also be thought of as a possible differential diagnosis of parapharyngeal space tumors.

#### Conflict of interest

None declared.

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