

Solitary Fibrous Tumor of the Infratemporal Fossa

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ABSTRACT

Solitary fibrous tumors represent fewer than 2% of all soft tissue tumors, and only about 12-15% of them occur in the head and neck. We report a case of a 38-year-old male who presented with a six-month history of increasing right cheek swelling. Computed tomography of the paranasal sinuses with contrast demonstrated a well-circumscribed avidly enhancing mass in the right retroantral fat. On magnetic resonance imaging the lesion was homogeneously slightly hyperintense to muscle on T1 weighted and T2 weighted images and enhanced avidly with contrast. Surgical resection was performed and pathology was consistent with solitary fibrous tumor. There have been very few reported cases of solitary fibrous tumors in the infratemporal fossa and none described as originating in the retroantral fat.

CASE REPORT

CASE REPORT

A 38-year-old male presented with a six-month history of slowly progressive right facial swelling. Physical examination demonstrated a firm lesion, the inferior margin of which could be palpated via a transoral approach. The patient had diminished facial sensory function to light touch in the right V2 distribution. Head and neck examination was otherwise normal with no other lesions and no enlarged lymph nodes.

Imaging Findings

CT scan of the paranasal sinuses with intravenous contrast demonstrated a 4.0 x 2.6 x 1.3 cm avidly enhancing, well-circumscribed lesion centered in the right retroantral fat extending caudally anterior to the superior alveolar ridge of the mandible. The lesion abutted the lateral wall of the right maxillary sinus resulting in bone remodeling without cortical erosion (Figure 1). No other lesions were identified.

On MRI, the lesion was well encapsulated and separate from the adjacent muscle. It was homogeneously slightly hyperintense to muscle on T1 weighted (T1W) and T2 weighted (T2W) images and homogeneously enhanced on post-contrast images (Figure 2).

The radiological features, particularly the bone remodeling on CT, suggested a non-aggressive entity and the differential primarily included a nerve sheath tumor in that location.

Management

Resection was performed via a transoral approach. Gross examination of the specimen demonstrated a well-encapsulated, firm mass measuring 3.8 x 2.8 x 1.4 cm and weighing 8.0 grams. Microscopically, the mass consisted of cytologically bland spindle cells arranged in a patternless architecture along with thin-walled blood vessels arranged in a stag-horn branching pattern (Figure 3). The cells were round

and spindle-shaped with indistinct borders, rare mitoses, and were separated by thick collagen bands. The immunohistochemical staining profile of the tumor cells was strongly and diffusely positive for cluster of differentiation (CD) 34, B cell lymphoma 2 (BCL2), and CD99 and negative for epithelial membrane antigen (EMA), keratin, and S100 protein. No calcifications were present. Both the morphologic and immunohistochemical features were consistent with a diagnosis of solitary fibrous tumor.

Follow-up

The patient recovered well from the surgery and his facial swelling gradually resolved. Follow-up CT at 6 months showed complete resection of the mass with no signs of recurrence. There was mild thickening of the right retroaural fat plane, which was likely postsurgical in nature (Figure 4).

DISCUSSION

Etiology and demographics

Solitary fibrous tumors (SFTs) are a rare group of mesenchymal tumors, suspected to be of fibroblastic origin. They are typically composed of bundles of spindle cells arranged in a patternless architecture with a background of collagen [1] and a dilated 'staghorn'-like vascular network [2]. Some can show myxoid changes, fibrosis, hyalinization, necrosis, or focal calcifications [3]. SFTs usually stain positive for CD34 (95%) and CD99 (70%) [2,4]. They may be positive for BCL2, EMA and smooth muscle actin (SMA) (~30%) [2,4,5]. They are usually negative for keratin and desmin.

SFTs can occur in any site, but most commonly occur in subcutaneous tissue (40%)[2]. A study conducted by Demicco et al of 110 cases found about 32% to be in the abdomen/pelvis, 32% to be pleural, 16% in the extremities, 12% in the trunk, and 12% in the head and neck. SFTs tend to occur in adults aged 20-70 and equally in males and females [2]. They are very rare in children [3].

Clinical findings

Presenting symptoms depend on the location. Soft tissue SFTs present with a slowly enlarging painless mass [2-3]. Head and neck SFTs may also present with obstructive symptoms, headaches, epistaxis, or rhinorrhea [6]. Two paraneoplastic syndromes have been reported: hypoglycemia due to production of an insulin-like growth factor (Doegge-Potter syndrome) or hypertrophic osteoarthropathy, neither of which was present in our patient.

The World Health Organization (WHO) classifies SFTs as intermediate risk for aggressive behavior. The behavior is usually benign but can be difficult to predict [1]. The percentage of SFTs that are found to be malignant varies in the literature, with most studies reporting ranges from 5 to 26% of cases [2-3, 7]. Most metastases are to the lung, followed by the bone and liver. Malignancy potential increases with the presence of necrosis, 4 or more mitotic figures per 10 high power fields, increased cellularity, and cellular polymorphism [3]. Increased tumor size (greater than 10cm), the presence of

hemorrhage, infiltrative margins, and the presence of anaplastic foci also point to malignancy. Local recurrence after resection can also occur, with reported rates of around 6% in solitary disease and around 23% in malignant disease [3].

Imaging findings

On imaging studies, SFTs typically have a non-aggressive appearance irrespective of the location. On CT imaging, soft tissue SFTs frequently appear as a well-defined hyperdense to isodense avidly enhancing mass [7-9]. Calcifications may be present in as many as 10% of cases [3]. Bone erosion may be present [10]. Hemorrhage and central necrosis may be visible in larger lesions. On MRI, SFTs can be isointense [9] or slightly hyperintense [11] to skeletal muscle on T1W and T2W images. The mass enhances avidly post-contrast. Enhancement pattern is prominent [12] and may be homogenous, heterogeneous or patchy [3,9,11]. Intralesional cystic areas and cystic degeneration have been reported [9]. SFTs do not typically show restricted diffusion. [12] On ultrasound, the expected finding is a homogenous hypoechoic or mostly hypoechoic mass with heterogeneous components [13]. A vascular pedicle may be seen as well [10]. On positron emission tomography (PET), SFT shows increased metabolic activity [10]. Therefore, PET can be helpful when multiple lesions are suspected.

Differential Diagnosis

The lesion in our patient had the classic CT and MRI features described for SFT and this entity should have been considered in the differential. Other tumors on the differential based on the non-aggressive appearance included neurofibroma, nerve sheath tumor, and leiomyoma, although these each have distinct imaging features when considering multiple modalities (Table 2) [13-17].

Schwannomas tend to be well circumscribed and demonstrate intermediate to high signal on T2W and intermediate signal on T1W [13]. They are diffusion weighted imaging (DWI) negative like SFTs, and exhibit homogenous enhancement in small lesions. On CT, schwannomas appear as ovoid to fusiform hypodense masses. Larger schwannomas with cystic change demonstrate nonenhancing areas (Figure 5). On ultrasound, they are homogenous and hypoechoic with posterior acoustic enhancement with peripheral nerve continuity. They are often positive on PET scan [14].

Neurofibromas, like SFTs, can be isointense on T1W and hyperintense on T2W, although the hyperintensity tends to be darker centrally, leading to a "target" sign [13]. They are DWI negative. On CT, neurofibromas are hypodense, unlike SFTs. On ultrasound, they are hypoechoic and homogenous, with posterior acoustic enhancement and peripheral nerve continuity. They are often positive on PET scan [15].

Leiomyomas, albeit exceedingly rare in the head and neck region, are on the differential as they have a similar appearance to SFTs. On MRI, they are isointense to hyperintense on T1W and homogenous to heterogeneous hyperintense to muscle on T2W. They are DWI negative and well defined, often with calcifications. On CT, they show increased density and show prominent contrast enhancement.

On ultrasound, they are hypoechoic if uncomplicated [16]. On PET scan, leiomyomas are often positive [17].

Treatment & prognosis

Most SFTs are surgically removed in order to end compressive symptoms and to establish a histological diagnosis. Conservative therapy for asymptomatic SFTs may yield to surgery as the tumor grows and becomes symptomatic; long-term follow-up is mandatory since 10-15% tend to behave in a locally aggressive fashion [2]. Adjuvant chemotherapy is rarely successful, and SFTs are regarded as chemoresistant [3]. Even with treatment, local and distant relapses can occur more than 20 years later. Resection with clear margins yields the best chance for curing SFTs with or without malignant features [7]. Radiotherapy can be considered if tumor displays evident criteria for malignancy or if the surgical margins were positive [7].

In summary, an adult presenting with a well circumscribed, avidly enhancing mass that is iso- to hyperintense compared to skeletal muscle on T1W and T2W MRI images in the head and neck should prompt a consideration of SFT. While most SFTs are benign, surgical resection is advised as these tumors continue to grow, can be locally invasive, and have the potential to be malignant.

TEACHING POINT

SFTs are uncommon soft tissue lesions in the head and neck that present as a well-defined solid, enhancing mass that is iso- to hyperintense compared to skeletal muscle on T1W and T2W MRI images. Surgical resection with wide margins is the treatment of choice as these tumors continue to grow, can be locally invasive, and have the potential to be malignant.

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FIGURES

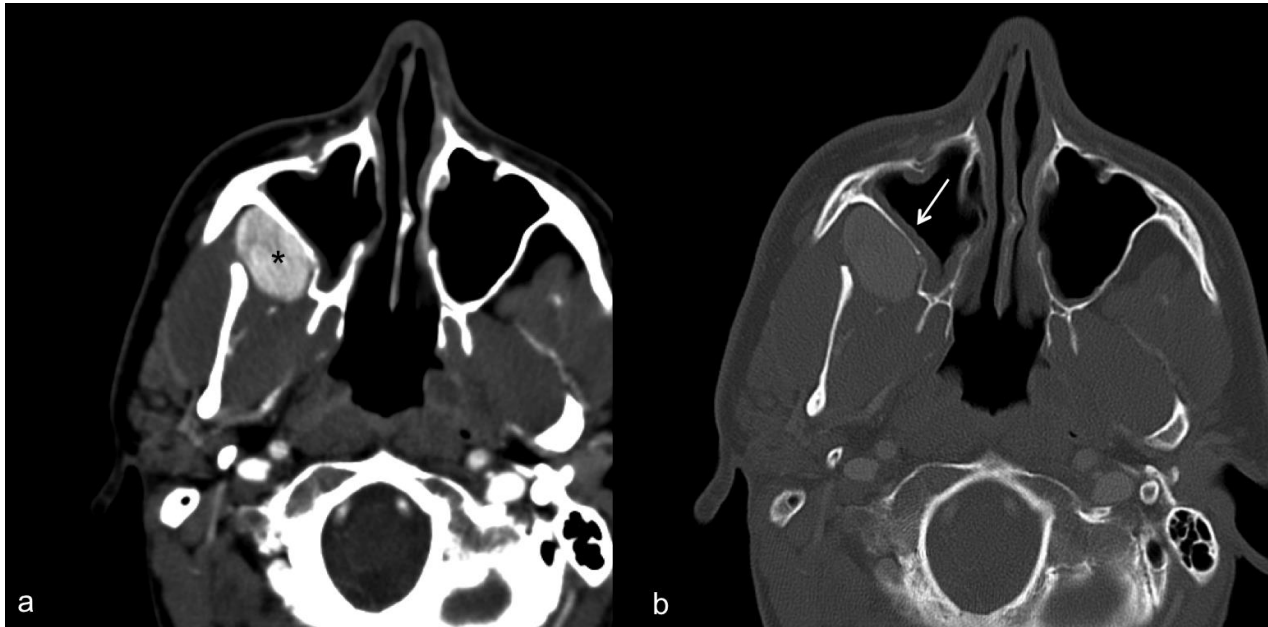


Figure 1: 38-year-old male with solitary fibrous tumor of the retroantral fat on the right side.

FINDINGS: Axial CT scan with intravenous contrast, venous phase, at the level of the retroantral fat. (a) Soft tissue window: There is a well defined, oval shaped solid enhancing mass (asterisk) measuring 4.0 x 2.6 x 1.3 cm in the retroantral fat on the right. (b) Bone window: the lesion scallops the posterolateral wall (arrow) of the right maxillary sinus.

TECHNIQUE: Axial CT, a. KV 120, mAs 100, slice thickness 3 mm ww/wl 441/161. Contrast: Isovue 300, 100 ml with 65 second delay. b. KV 120, mAs 100, slice thickness 3 mm ww/wl 2118/690. Contrast: Isovue 300, 100 ml with 65 second delay.

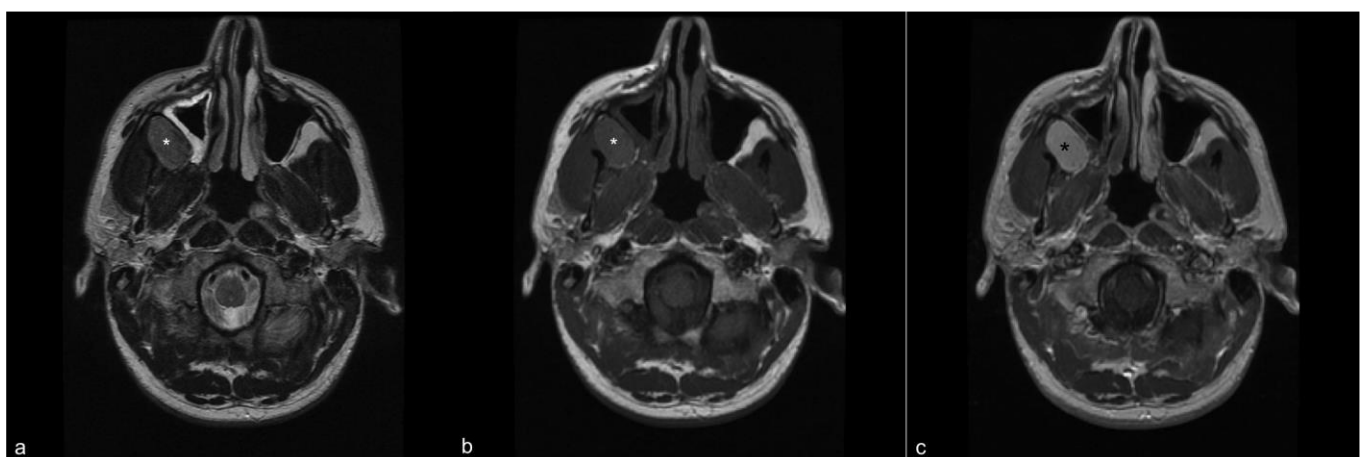


Figure 2: 38-year-old male with solitary fibrous tumor of the retroantral fat on the right side.

FINDINGS: On axial MR images at the level of the retroantral fat, there is a well-defined oval shaped solid mass (asterisks) measuring 4.0 x 2.6 x 1.3 cm in the retroantral fat on the right. The lesion is hyperintense to muscle on T2W (a), slightly hyperintense on T1W without contrast (b) and shows avid, homogeneous enhancement on T1W post contrast (c).

TECHNIQUE: Axial MRI, 1.5 tesla a. T2W: TR 4266, TE 98, slice thickness 4 mm, ww/wl 1612/829. b. T1W: TR 416, TE 14, slice thickness 4 mm ww/wl 1622/811. c. T1W + C 416, TE 14, slice thickness 4 mm ww/wl 2356/178. Contrast: 12 ml MultiHance.

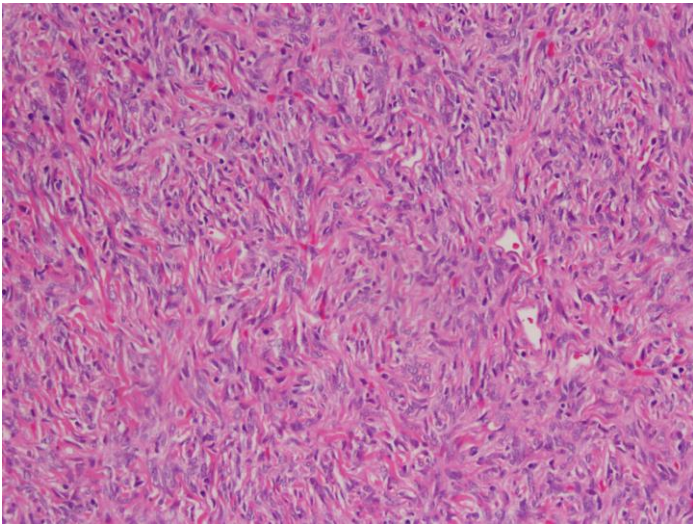


Figure 3 (left): 38-year-old male with solitary fibrous tumor of the retroantral fat on the right side.

FINDINGS: Histological examination (40x) with H&E stain revealed a solitary fibrous tumor consisting of spindle cells with a patternless architecture, thin-walled blood vessels, and collagen bands.



Figure 4 (right): 38-year-old male with solitary fibrous tumor of the retroantral fat on the right side.

FINDINGS: On six month postoperative axial CT with intravenous contrast at the level of the retroantral fat, the lesion has been completely resected with no evidence of recurrence. There is minimal stranding of the retro antral fat on the right (arrow), likely post surgical.

TECHNIQUE: Axial CT, KV 120, mAs 77, slice thickness 3 mm ww/wl 304/29. Contrast: Isovue 300, 100 ml.

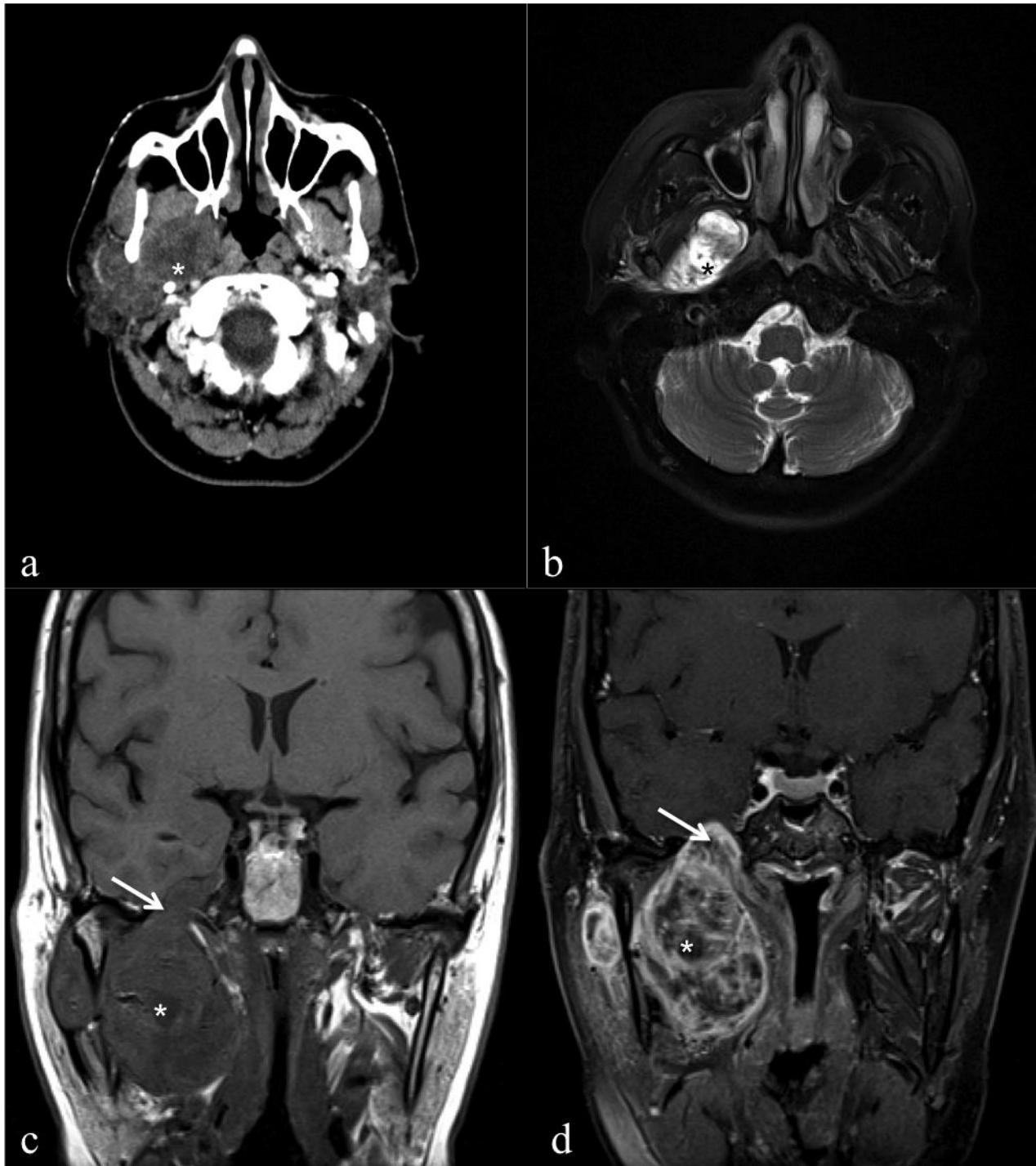


Figure 5: 49-year-old female with right parapharyngeal space schwannoma. Patient presented with an enlarging right neck mass. On axial CT post-contrast soft tissue window (a), the lesion is hypodense with central necrotic areas (asterisk) as is characteristic of large schwannomas. On axial T2W image (b) the lesion is hyperintense with high signal central areas of necrosis (asterisk.) A coronal T1W image (c) shows the lesion to be isointense to skeletal muscle with areas of necrosis (asterisk). On the post-contrast T1W image (d), the lesion enhances heterogeneously with the necrotic areas not enhancing (asterisk). Note the superior extent of the lesion to the foramen ovale. Technique: a. Axial CT scan with contrast, soft tissue window, KV 140, mAs 140, slice thickness 3mm ww/wl 196/53. Contrast: Isovue 300, 100 ml with 65 second delay. b. Axial T2W MRI, 1.5 Tesla, TR 4910, TE 113, slice thickness 5 mm, ww/wl 1011/504. c. Coronal T1W MRI, SE without contrast, slice thickness 3mm, TR 611, TE 18, ww/wl 852/471. d. Coronal T1W FS post-contrast, slice thickness 3mm, TR 568, TE 118, ww/wl 1242/708. Contrast: 12 mL MultiHance.

Etiology	Mesenchymal tumor of probable fibroblastic type
Incidence	Rare; 1-2% of soft tissue tumors are SFTs, of which 12-15% are in the head and neck.
Gender ratio	1:1
Age Predilection	20 to 70 years; median age 45-50. Occasionally in children and adolescents.
Risk Factors	No known risk factors
Treatment	En-bloc resection.
Prognosis	Local recurrence after surgical removal: 0-6%. Malignant transformation with metastatic disease: 5-26%, most commonly to lungs, bone and liver. 5-year survival 89-100%.
Findings on Imaging	CT: hyperdense / isodense, avid enhancement MRI: T1W: isointense / hyperintense to skeletal muscle. T2W: isointense / hyperintense to skeletal muscle. Avid enhancement.

Table 1: Summary table of clinical and imaging findings in patients with extrapleural solitary fibrous tumor (SFT)

	US	CT	MRI – T1, T2, DWI	Enhancement Pattern	FDG PET	Other
SFT	<ul style="list-style-type: none"> Homogenous hypoechoic or mostly hypoechoic 	<ul style="list-style-type: none"> Hyperdense to isodense 	<ul style="list-style-type: none"> T1W and T2W: hyperintense DWI: negative 	<ul style="list-style-type: none"> Strong homogeneous enhancement 	<ul style="list-style-type: none"> Positive 	<ul style="list-style-type: none"> Case reports of positive thallium scan
Schwannoma	<ul style="list-style-type: none"> Hypoechoic and homogeneous, with posterior acoustic enhancement and peripheral nerve continuity 	<ul style="list-style-type: none"> Ovoid to fusiform hypodense mass often with cystic changes 	<ul style="list-style-type: none"> T1W: isointense T2 W: isointense/hyperintense DWI: negative 	<ul style="list-style-type: none"> Strong homogeneous enhancement in small lesions Cystic change in larger lesions: nonenhancing areas 	<ul style="list-style-type: none"> Positive 	<ul style="list-style-type: none"> Case reports of positive Thallium scan Rarely positive on Octreotide scan
Neurofibroma	<ul style="list-style-type: none"> Hypoechoic and homogeneous, with posterior acoustic enhancement and peripheral nerve continuity 	<ul style="list-style-type: none"> Hypodense 	<ul style="list-style-type: none"> T1W: Isointense T2W: Hyperintense, darker centrally ("target" sign) DWI: negative 	<ul style="list-style-type: none"> Mild homogeneous or patchy enhancement 	<ul style="list-style-type: none"> Positive 	<ul style="list-style-type: none"> Case reports of positive Thallium scan Typically negative on Octreotide scan
Leiomyoma (superficial)	<ul style="list-style-type: none"> Hypoechoic if uncomplicated 	<ul style="list-style-type: none"> Well defined. Typically have calcifications resulting in increased density 	<ul style="list-style-type: none"> T1W: Isointense to slightly hyperintense relative to muscle T2W: homogeneous to heterogeneous signal hyperintense to muscle DWI: negative 	<ul style="list-style-type: none"> Strong contrast enhancement 	<ul style="list-style-type: none"> Positive 	<ul style="list-style-type: none"> Rare in the neck.

Table 2: Presentation and imaging of the most important differential diagnoses for extrapleural solitary fibrous tumor (SFT)

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ABBREVIATIONS

CT - Computed Tomography
DWI - Diffusion Weighted Imaging
MRI - Magnetic Resonance Imaging
SFT - Solitary Fibrous Tumor
TE - Echo Time
TR - Repetition Time
T1W - T1 Weighted
T2W - T2 Weighted
WHO - World Health Organization

KEYWORDS

Solitary fibrous tumor; infratemporal fossa tumor; fibrous tumor; soft tissue tumor; head and neck; retroantral fat; extrapleural; CT; MRI

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