Rare localizations of malignant Solitary Fibrous Tumors (SFT): clinical presentation and radiological characteristics

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Learning objectives

Our aim is to identify and discuss, with a special focus on differential diagnosis, solitary fibrous tumors (SFTs) arising in unusual localizations with their clinical presentations.

Background

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms commonly arising from the pleura and the peritoneum. Most of them are benign, but 10-20% are malignant. SFTs are defined malignant when histopathological analysis shows at least one among high mitotic activity, high cellularity, necrosis, hemorrhage or pleomorphism.

Typical radiological, clinical and laboratory characteristics include:

- pleural or peritoneal localization
- round shape and well-defined margins
- increased contrast enhancement
- inhomogeneous enhancement directly proportional to the size of lesions (due to central necrosis, hemorrhage and cystic changes with a patchy pattern)
- branching vascular pattern
- middle-aged adults with no sex predilection

- nonspecific signs and symptoms, mainly related to the tumor's mass effect (dysphagia, bowel/respiratory obstruction), hemorrhage (epistaxis) or IGF production (Doege-Potter syndrome)
- clubbing and hypertrophic pulmonary osteoarthropathy (HPO)

- CD34+, CD117-, vimentine+

Very few reports have been published about rare tumor localizations with consequent atypical clinical presentations. These include pelvis, abdomen, retroperitoneum, buccal space, maxillary sinuses, visceral organs and extremities [1].

We identified in our Institute selected cases of atypical extrathoracic SFTs which underwent multimodality imaging (US/CT/MRI/PET).

Findings and procedure details

We searched though our Institutional databse all biopsy-proven diagnosis of SFTs with atypical presentations (extrapleural and extraperitoneal tumors).

In particular, we describe 3 cases of atypical localizations of SFTs with their CT, MR, US and PET characteristics.

CASE 1

Initially misdiagnosed as lymphadenopathy, this submandibular mass was later confirmed to be a SFT (CD34+, Bcl2+, CD99+).

Color Doppler US shows a vascularized hypoechoic mass in the submandibular region. US-guided core needle biopsy was subsequently performed to confirm the diagnosis.

MRI showed hyperintense mass in CE axial/coronal/sagittal T1W, mildly hipointense in both axial T2W STIR and T2 and highy hyperintense in DWI b=800.

CASE 2

Diffuse aspecific arthralgia was caused by a SFT localized in the left thigh. Pathology confirmed the diagnosis of SFT (CD34+, S100-, EMA-, Somatostatin receptor II, III, V A/B).

68Ga-DOTA-TOC PET/CT shows an avid soft tissue lesion in the left thigh.

MRI showed mildly hyperintensity in coronal FS-T2W, as well as in CE coronal and axial T1W.

CASE 3

Nasal obstruction and bleeding were caused by nasal localization of SFT.

Pathology confirmed the diagnosis of SFT (MIB1 10%, CD34 +, CD31 +, ActinML+, Factor XIII+, Patient 1).

CT showed a hypodense lesion within the left nasal cavity at coronal/sagittal unenhanced CT.

MRI showed a strongly hyperintense lesion in CE coronal T1W, as well as in the unenhanced sagittal T1W sequence.

In sagittal T2W the lesion appeared slightly hyperintense.

Images for this section:

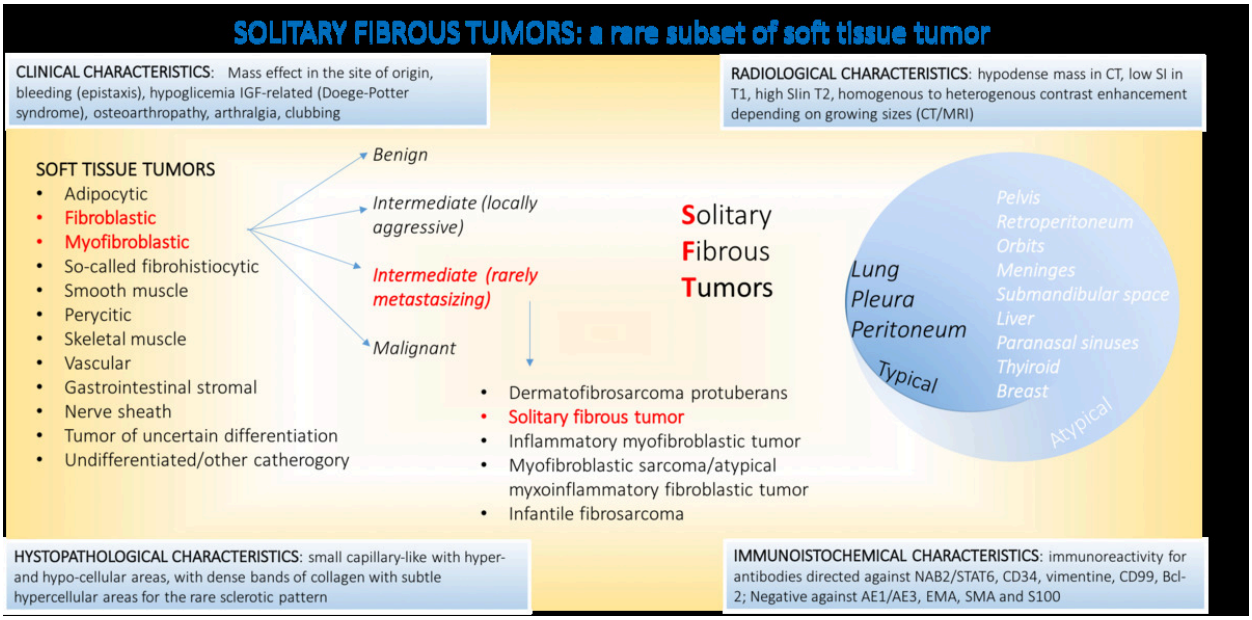

Fig. 1: Diagram showing the classification of soft tissue tumors, typical and atypical localizations of SFT, a rapid overview of clinical, radiological, histopathological and immunoistochemical characteristics.

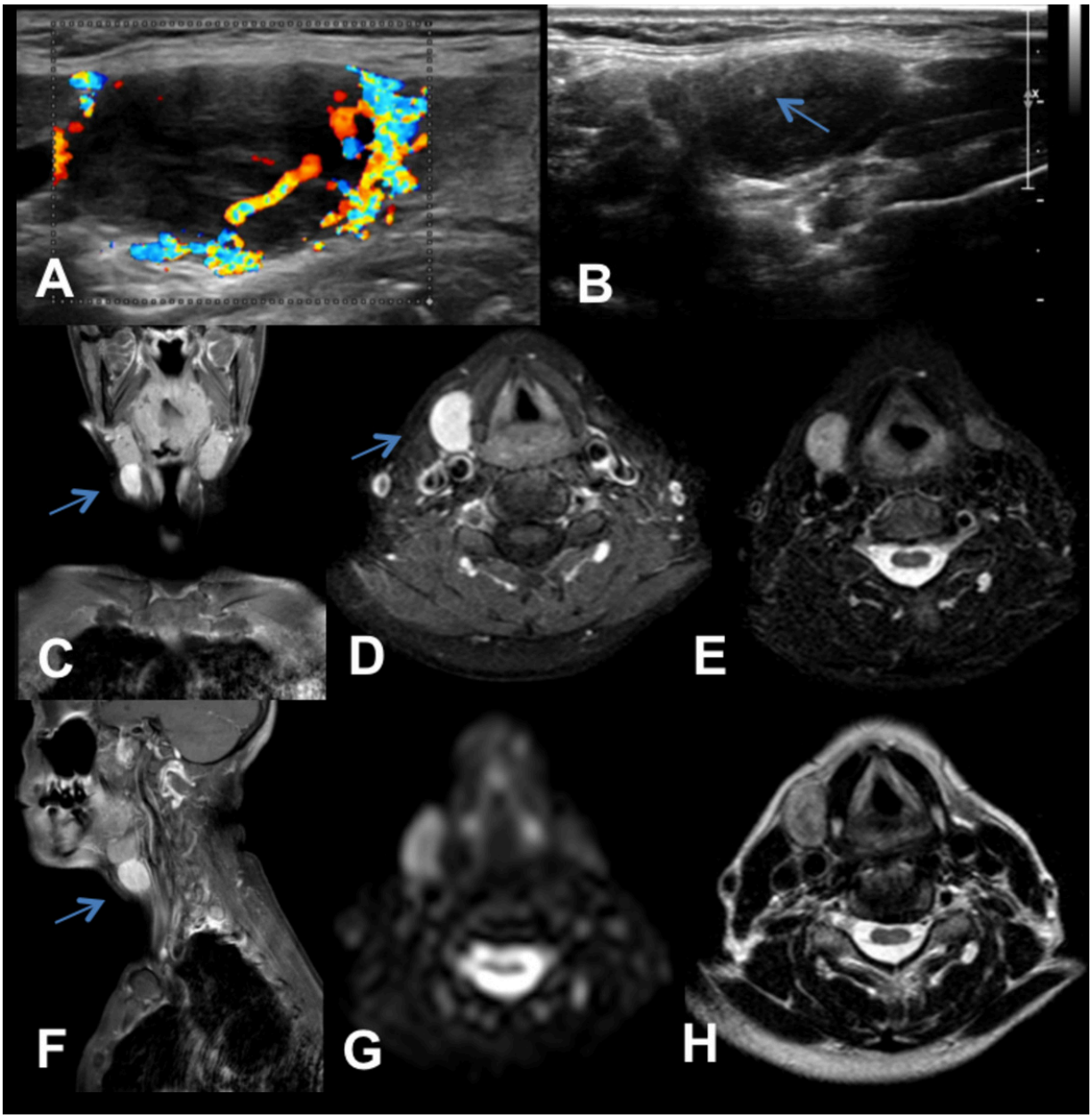

Fig. 2: A submandibular SFT is shown. Initially it was misdiagnosed as a lymph-adenopathy. Pathology confirmed the diagnosis of SFT (CD34+, Bcl2+, CD99+). A) Color Doppler US B) Percutaneous US-guided fine core needle biopsy (axial technique, arrow shows the needle within the lesion) C/F) CE coronal/sagittal T1W E) axial T2W STIR G) DWI b=800 H) axial T2W.

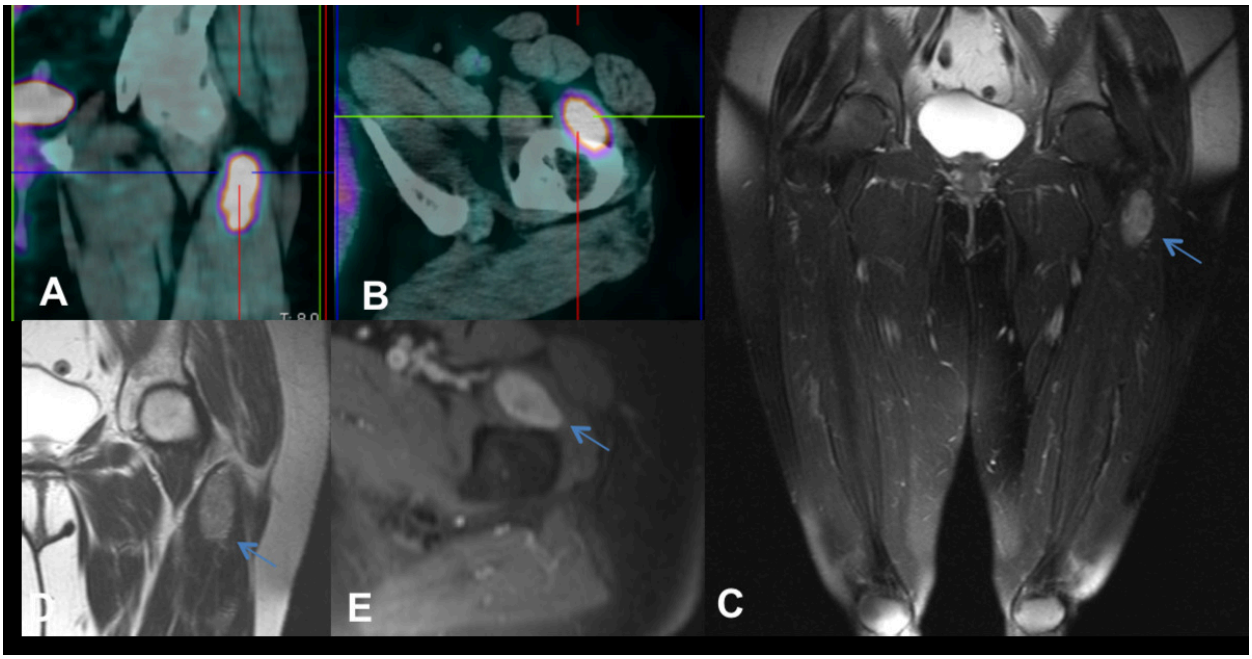

Fig. 3: An atypical left thigh localization of SFT is shown. The patient suffered from diffuse aspecific arthralgia Pathologdiagnosis of SFT (CD34+, S100-, EMA-, Somatostatin receptor II, III, V A/B) 68Gaist confirmed the -DOTA-TOC PET/CT imaging shows an avid soft tissue lesion in the left thigh C) coronal FS-T2W D) CE coronal T1W E) CE axial T1W

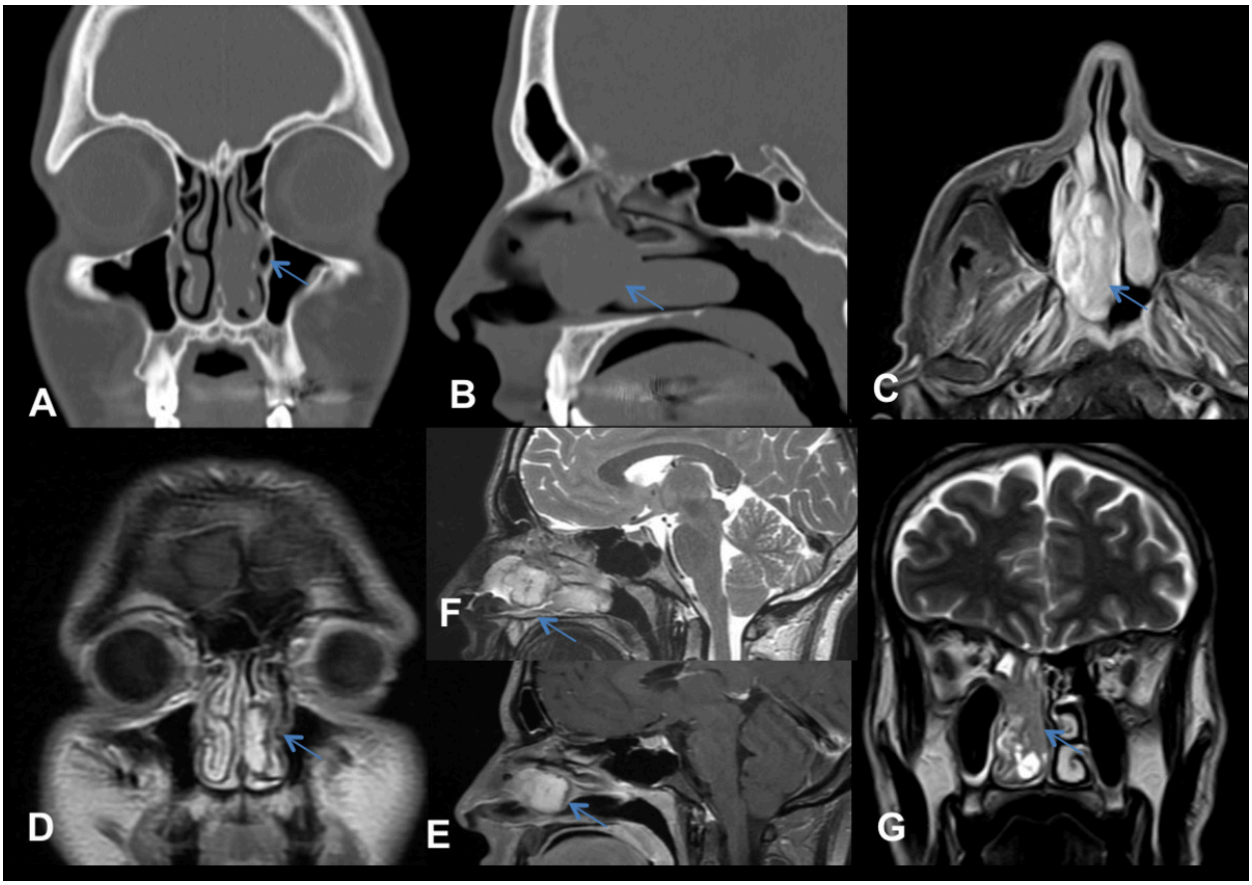

Fig. 4: An atypical nasal localization of SFT is shown in 2 with nasal obstruction and bleeding. Pathologist confirmed the diagnosis of SFT (MIB1 10%, CD34 +, CD31 +, ActinML+, Factor XIII+, Patient 1). Patient 1 A/B) coronal/sagittal unenhanced CT D) CE coronal T1W E) unenhanced sagittal T1W F) sagittal T2W Patient 2 C) CE axial T1W G) coronal T2W

Conclusion

STSs are rare but extremely heterogeneous tumors with a wide spectrum of clinical and radiological presentations. The majority of STSs are not locally invasive, however, a subset of STSs does show invasiveness and metastatic potential.

Being aware of atypical locations allows the radiologist to include potentially invasive tumors like SFTs in the differential diagnosis of extra-pleural/peritoneal soft tissue lesions.

Even though the final diagnosis relies on histopathological analysis, radiologists play a fundamental role in the diagnosis of SFTs through initial identification, biopsy guidance, surgery planning, and follow-up.

References

1. Zhanlong M, Haibin S, Xiangshan F, Jiacheng S, Yicheng N. Variable solitary fibrous tumor locations: CT and MR imaging features. Medicine (Baltimore) 2016;95:e3031. doi:10.1097/MD.0000000000003031.