Malignant fat-forming solitary fibrous tumor (lipomatous hemangiopericytoma) in the neck: Imaging and histopathological findings of a case

Alice Duarte de Carvalho[1], Lucas Faria Abrahão-Machado[2], Cristiano Ribeiro Viana[2], Renato de Castro Capuzzo[3], Augusto Elias Mamere[1*]

1. Department of Radiology, Barretos Cancer Hospital, Barretos, Brazil

2. Department of Pathology, Barretos Cancer Hospital, Barretos, Brazil

3. Department of Head and Neck Surgery, Barretos Cancer Hospital, Barretos, Brazil

* **Correspondence:** Dr. Augusto Elias Mamere, Barretos Cancer Hospital, Avenida Antenor Duarte Vilela, 1331 - Barretos - SP, CEP: 14784-400, Brazil
(*mamere@uol.com.br*)

Radiology Case. 2013 Mar; 7(3):1-7 :: DOI: 10.3941/jrcr.v7i3.1336

ABSTRACT

Fat-forming solitary fibrous tumor (SFT) is a rare variant of solitary fibrous tumor, a mesenchymal fibroblastic neoplasia with a particular branching hypervascular pattern. This tumor is usually classified as benign and only very few fat-forming SFTs with malignant histologic features have been reported. We report a histologically malignant fat-forming solitary fibrous tumor in a 61-year-old man, located in his neck. Ultrasonography examination was first performed showing a heterogeneous lesion, predominantly hyperechoic, with sound beam attenuation, containing two hypoechoic solid nodules. Magnetic resonance imaging and computed tomography examinations demonstrated a heterogeneous and predominantly adipose mass, containing post contrast enhancing solid nodules and thin septations. Treatment consisted of total removal of the lesion. Histologically, the tumor showed hypercellularity, numerous mitoses and cytological atypia, fulfilling the criteria for malignancy. The patient had no metastasis. This rare tumor may be confused with other fat-containing lesions on imaging examinations, mainly liposarcoma.

CASE REPORT

CASE REPORT

A 61-year-old man presented with a more than 10-year history of a slowly growing, minimally tender left supraclavicular fossa mass. Family history was noncontributory. Physical examination revealed a 5-cm painless mass within the deep soft tissue of the left supraclavicular fossa. Motor and sensory examination of the respective limb was normal. There was no lymphadenopathy, and the remainder of the physical examination was unremarkable.

Ultrasonography (US) examination (linear transducer; frequency: 10.0 MHz) of the neck was performed first and showed a heterogeneous lesion that was predominantly hyperechoic, with sound beam attenuation, containing two hypoechoic solid nodules. The lesion demonstrated low vascularization on color Doppler (figure 1).

Computed tomography (CT) examination (General Electric, multidetector helical CT; 16 channels; 350 mAs; 120 kVp; 1.25 mm slice thickness, 100 ml of intravenous iodinated contrast medium) demonstrated a predominantly adipose and heterogeneous mass in the left supraclavicular fossa, with ill-

defined borders, containing solid nodules. It demonstrated scattered fat attenuation areas (mean attenuation value (in Hounsfield units) of -64). The solid nodules inside the adipose mass showed moderate post contrast enhancement. Some thin septations were also present inside the lesion (figure 2). The lesion measured 5.5 cm in its longest axis in the transverse plane.

Magnetic resonance imaging (MRI) confirmed the predominantly fatty matrix of the lesion (General Electric, Signa HDx, 1.5 Tesla). The correspondent areas with low attenuation on the CT images presented with high signal intensity on the T1-weighted images, similar to that observed in the subcutaneous adipose tissue. On the fat-suppression sequences, the signal intensity decreased considerably in all these areas. The solid nodules inside the lesion were isointense on the T1-weighted images and moderately hyperintense on the T2-weighted ones. Similarly to what was noted on the CT examination, these nodules also demonstrated moderate post contrast enhancement on the MRI evaluation (figure 3). Facing these imaging findings, liposarcoma was considered the main hypothesis.

Treatment consisted of total excision of the lesion. At gross examination, the tumor showed a tan and whitish cut surface, with a firm appearance and yellowish fatty areas. Nodules of elastic consistency were present within the mass. At microscopy, the tumor was characterized by a hypercellular proliferation of round and spindle cells in a patternless architecture, with alternating areas of thick bands of collagen and branching hemangiopericytoma-like vessels (figure 4). A considerable number of mature adipocytes were observed throughout the neoplasia. The tumor showed marked cytological atypia in addition to hypercellularity and mitotic figures. At immunohistochemical analysis, tumor tissue was positive for CD34, Vimentin, CD99 and BCL-2. Based on these findings, the lesion was diagnosed as a histologically malignant fat-forming solitary fibrous tumor (SFT).

The perioperative course was uneventful and the patient was asymptomatic after surgery. At 9 months follow-up, there was no evidence of local recurrence or distant metastasis.

DISCUSSION

Fat-forming solitary fibrous tumor (SFT), also known as lipomatous hemangiopericytoma, is a rare variant of SFT, usually occuring in middle-aged patients (typically between 40 and 60 years old), with no obvious gender predilection. This entity was first described by Nielsen et al in 1995 (1). They have described three cases of a singular tumor composed of mature adipocytes and hemangiopericytomatous areas, for which they proposed the term lipomatous hemangiopericytoma. Lately, the term "fat-forming solitary fibrous tumor" has gained increasing popularity (2, 3, 4, 5). To date, there have been only a small number of cases reported in the literature, most of them as single cases or small series (2). Only three fat-forming SFTs were reported in this region, and just one of them was malignant. This is the second malignant fat-forming SFT of the neck region described so far (2, 3, 6).

This tumor is a fibroblastic neoplasia characterized by an admixture of variable amounts of collagen and proliferation of vascular spaces in a pattern called hemangiopericytoma-like, associated with a mature adipocytic component. It usually shows benign histologic features and, more frequently, it has an indolent clinical course (4, 6, 7, 8). Only very rarely have fat-forming SFT's been described as containing malignant morphologic characteristics, and just a couple of them metastasized (2, 9). Complete surgical excision of the tumor is the main treatment.

Fat-forming SFT often presents as a heterogeneous lesion, with solid enhancing regions and scattered low-attenuating adipose areas on CT examination. The adipose areas show high signal intensity on T1-weighted MRI that decrease in intensity with fat-suppression sequences. The signal intensity of the solid areas can vary. These areas may be hypo, hyper or isointense on T2-weighted images and hypo or isointense on the T1-weighted sequences. The solid areas usually demonstrate post-contrast enhancement. Thin septations may be found inside the tumor. Benign and malignant SFT cannot be differentiated based only on imaging findings. With US examination, fat-forming SFTs are seen as heterogeneous and predominantly hyperechoic lesions with sound beam attenuation due to their fatty nature. They are usually associated with solid hypoechoic areas.

Histologic parameters involving increased cellularity, pleomorphism, mitoses > 4/10 high-power fields (HPF), hemorrhage and necrosis were grouped as malignant features for SFTs (2). Bai et al (9) reported a malignant pleural SFT with "liposarcomatous differentiation" exhibiting marked nuclear atypia, numerous mitoses and necrosis, with subsequent metastasis. Guillou et al (3) reported two cases, both of which had > 4 mitoses per 10 HPFs and either atypia or necrosis. Our case met three of the currently accepted criteria for malignant SFT (hypercellularity, mitoses > 4/10 HPF and atypia), consistent with the concept that these findings seem to be among the most significant prognostic factors in SFT (2).

Regarding the variety of combinations and extent of different morphologic elements in an atypical or malignant fat-forming SFT, a number of etiologies may be considered for differential diagnosis. Taking into account the intimate juxtaposition of fat and the spindle cell component, liposarcoma should be considered as one of the most important on the list, particularly dedifferentiated liposarcoma.

Teratoma is another fatty tumor that can be present in the cervical region, but it has different features compared to our case. It is congenital, seen almost exclusively in children and young adults, and it usually has cystic areas (10). Calcification may be seen in up to 50% of these lesions and it is considered a pathognomonic feature. Predominantly fatty dermoid cysts may look very similar to the fat-forming SFT in US examination (hyperechoic lesion with sound beam attenuation), but its cystic aspect can be identified with MRI

and CT examinations, especially when a fat-fluid level is present. Unlike the SFT, dermoid cysts usually do not contain solid areas.

The adipose component of the fat-forming SFTs reported in the literature were composed mostly of benign mature adipocytes, as in our case, with few exceptions. Ceballos et al (4) described one malignant fat-forming SFT formed by lipoblasts, which made it difficult to distinguish from a dedifferentiated liposarcoma. Immunostaining or molecular studies are often necessary. The presence of an obvious SFT component, which typically stains for CD34 and/or CD99, is often enough to make the correct diagnosis.

Regardless, the imaging findings of SFTs are not specific. They usually show isoattenuation relative to the adjacent muscles on pre contrast CT scans, and on MRI they are mostly isointense to the muscle on T1-weighted images, with variable signal intensity on T2-weighted images, specially due to the presence of an intratumoral fatty component. They also show heterogeneous post contrast enhancement because of the combination of dense collagenization, increased cellularity and dilated vascular spaces (8).

Although it is a rare neoplasm, fat-forming SFT should be considered in the differential diagnosis along with liposarcoma when the image shows a heterogeneous mass containing macroscopic fat in an adult patient.

TEACHING POINT

Fat-forming solitary fibrous tumor (also known as lipomatous hemangiopericytoma) is a rare neoplasia and it may be confused with other fat-containing lesions, mainly liposarcoma. In general, it shows macroscopic fat tissue on imaging examinations (regions with low attenuation on the computed tomography images and with high signal intensity on the T1-weighted magnetic resonance images) associated with solid nodules.

REFERENCES

1. Nielsen GP, Dickersin GR, Provenzal JM, Rosenberg AE. Lipomatous hemangiopericytoma. A histologic, ultrastructural and immunohistochemical study of a unique variant of hemangiopericytoma. Am J Surg Pathol. 1995 Jul;19(7):748-56. PMID: 7793472

2. Lee JC, Fletcher CD. Malignant fat-forming solitary fibrous tumor (so-called "lipomatous hemangiopericytoma"): clinicopathologic analysis of 14 cases. Am J Surg Pathol. 2011 Aug;35(8):1177-85. PMID: 21716088

3. Guillou L, Gebhard S, Coindre JM. Lipomatous hemangiopericytoma: a fat-containing variant of solitary fibrous tumor? Clinicopathologic, immunohistochemical, and ultrastructural analysis of a series in favor of a unifying concept. Hum Pathol. 2000 Sep;31(9):1108-15. PMID: 11014579

4. Ceballos KM, Munk PL, Masri BA, O'Connell JX. Lipomatous hemangiopericytoma: a morphologically distinct soft tissue tumor. Arch Pathol Lab Med. 1999 Oct;123(10):941-5. PMID: 10506450

5. Liu X, Zhang HY, Bu H, Meng GZ, Zhang Z, Ke Q. Fat-forming variant of solitary fibrous tumor of the mediastinum. Chin Med J (Engl). 2007 Jun 5;120(11):1029-32. PMID: 17624277

6. Alrawi SJ, Deeb G, Cheney R, et al. Lipomatous hemangiopericytoma of the head and neck: immunohistochemical and DNA ploidy analyses. Head Neck. 2004 Jun;26(6):544-9. PMID: 15162357

7. Hasegawa T, Hirose T, Seki K, Yang P, Sano T. Solitary fibrous tumor of the soft tissue. An immunohistochemical and ultrastructural study. Am J Clin Pathol. 1996 Sep;106(3):325-31. PMID: 8816589

8. Kim MY, Rha SE, Oh SN, et al. Case report. Lipomatous haemangiopericytoma (fat-forming solitary fibrous tumour) involving the perineum: CT and MRI findings and pathological correlation. Br J Radiol. 2009 Feb;82(974):e23-6. PMID: 19168684

9. Bai H, Aswad BI, Gaissert H, Gnepp DR. Malignant solitary fibrous tumor of the pleura with liposarcomatous differentiation. Arch Pathol Lab Med. 2001 Mar;125(3):406-9. PMID: 11231493

10. Hasiotou M, Vakaki M, Pitsoulakis G, et al. Congenital cervical teratomas. Int J Pediatr Otorhinolaryngol. 2004 Sep;68(9):1133-9. PMID: 15302143

FIGURES

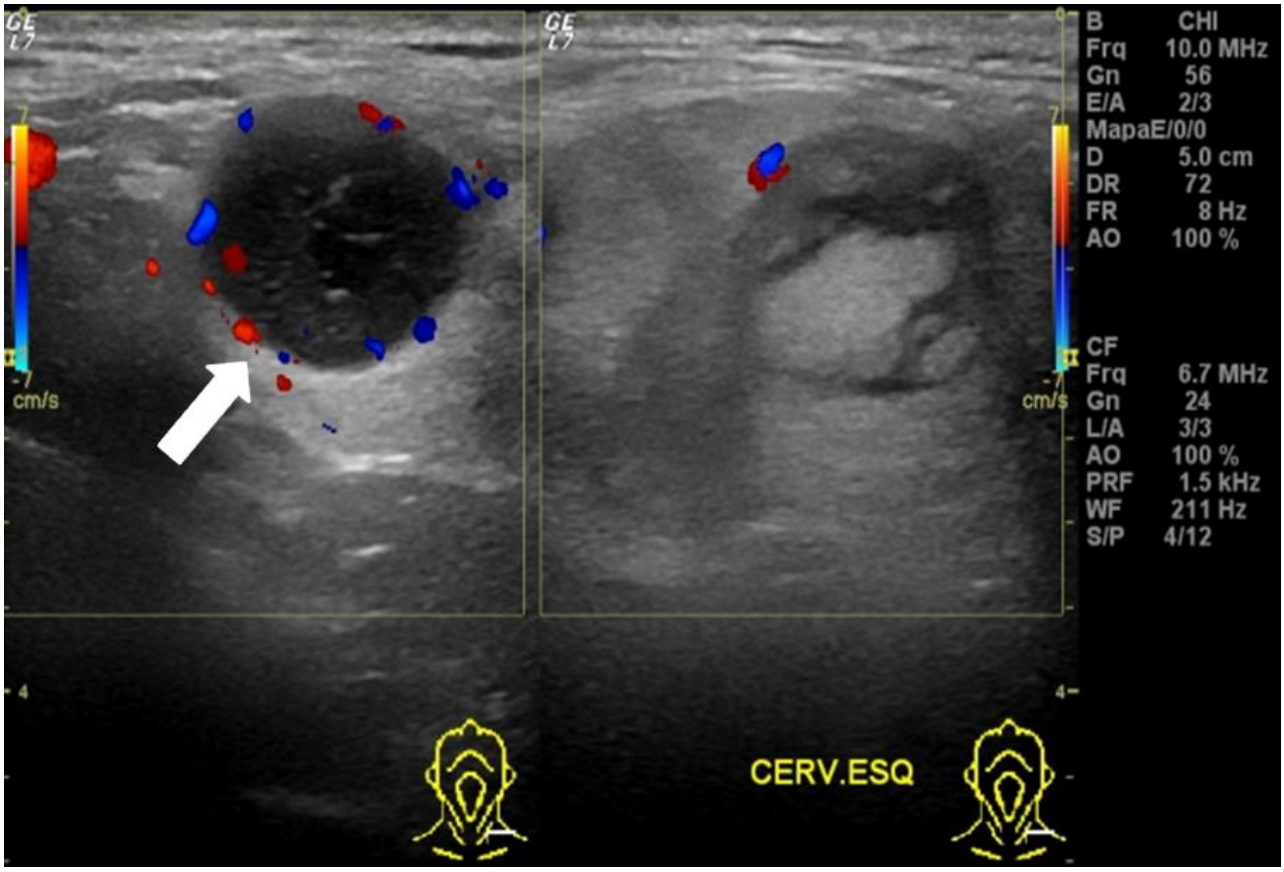

Figure 1: Malignant fat-forming solitary fibrous tumor in a 61-year-old man. Cervical ultrasound (linear transducer; frequency: 10.0 MHz) showing a heterogeneous lesion, predominantly hyperechoic, containing hypoechoic solid nodules with low vascularization on color Doppler (arrow).

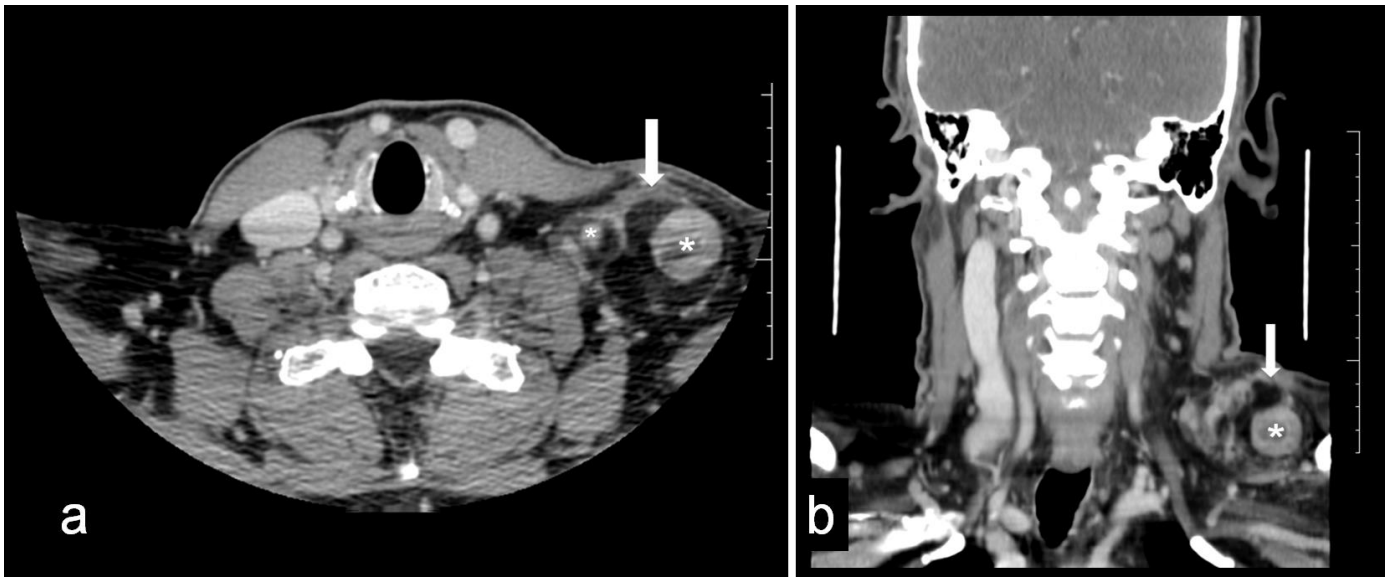

Figure 2: Malignant fat-forming solitary fibrous tumor in a 61-year-old man. Contrast-enhanced CT (multidetector helical CT; 16 channels; 350 mAs; 120 kVp; 1.25 mm slice thickness, 100 ml of intravenous iodinated contrast medium) in the transverse (a) and coronal (b) planes, demonstrating a lobulated and heterogeneous solid mass (arrows) located in the left supraclavicular fossa, containing enhancing solid nodules (asterisks), thin septations and scattered low-density areas suggestive of fat. The lesion measured 5.5 cm in its longest axis in the transverse plane.

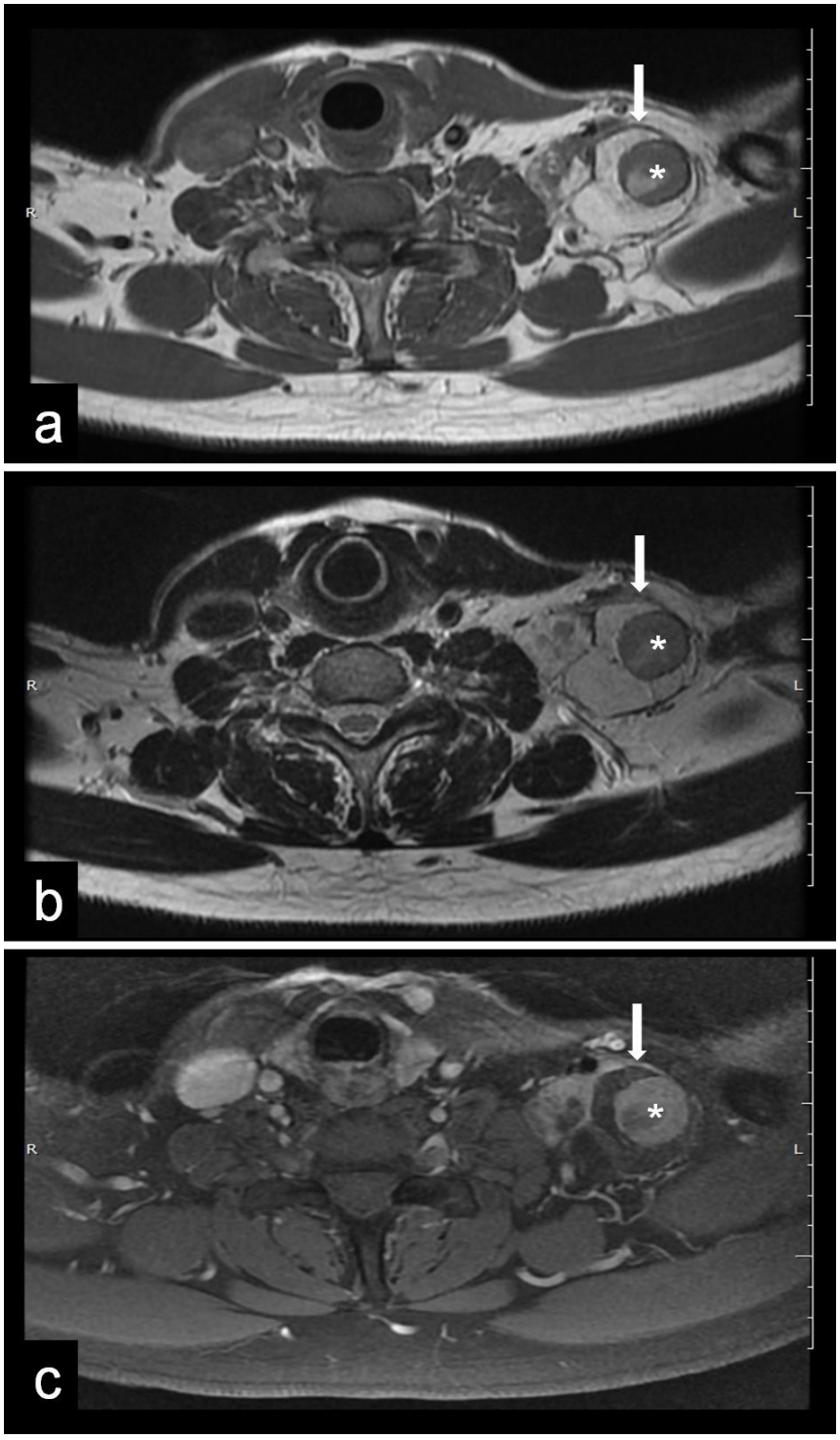

Figure 3: Malignant fat-forming solitary fibrous tumor in a 61-year-old man. MRI: Pre-contrast axial T1-weighted (a - 1.5 Tesla magnet, TR: 566; TE: 11) and axial T2-weighted (b - 1.5 Tesla magnet, TR: 3983; TE: 117) images showing the mass (arrow) with large areas of high signal intensity, identical to that of the subcutaneous adipose tissue. On the post-contrast T1-weighted images, with fat-suppression (c - 1.5 Tesla magnet, TR: 195; TE: 4; 15 ml of intravenous gadolinium contrast medium), the signal intensity decreases considerably in all these areas, confirming the predominantly fatty component. The nodules (asterisk) inside the lesion are isointense on the T1-weighted image (a), moderately hyperintense on the T2-weighted image (b) and they demonstrate moderate post contrast enhancement (c).

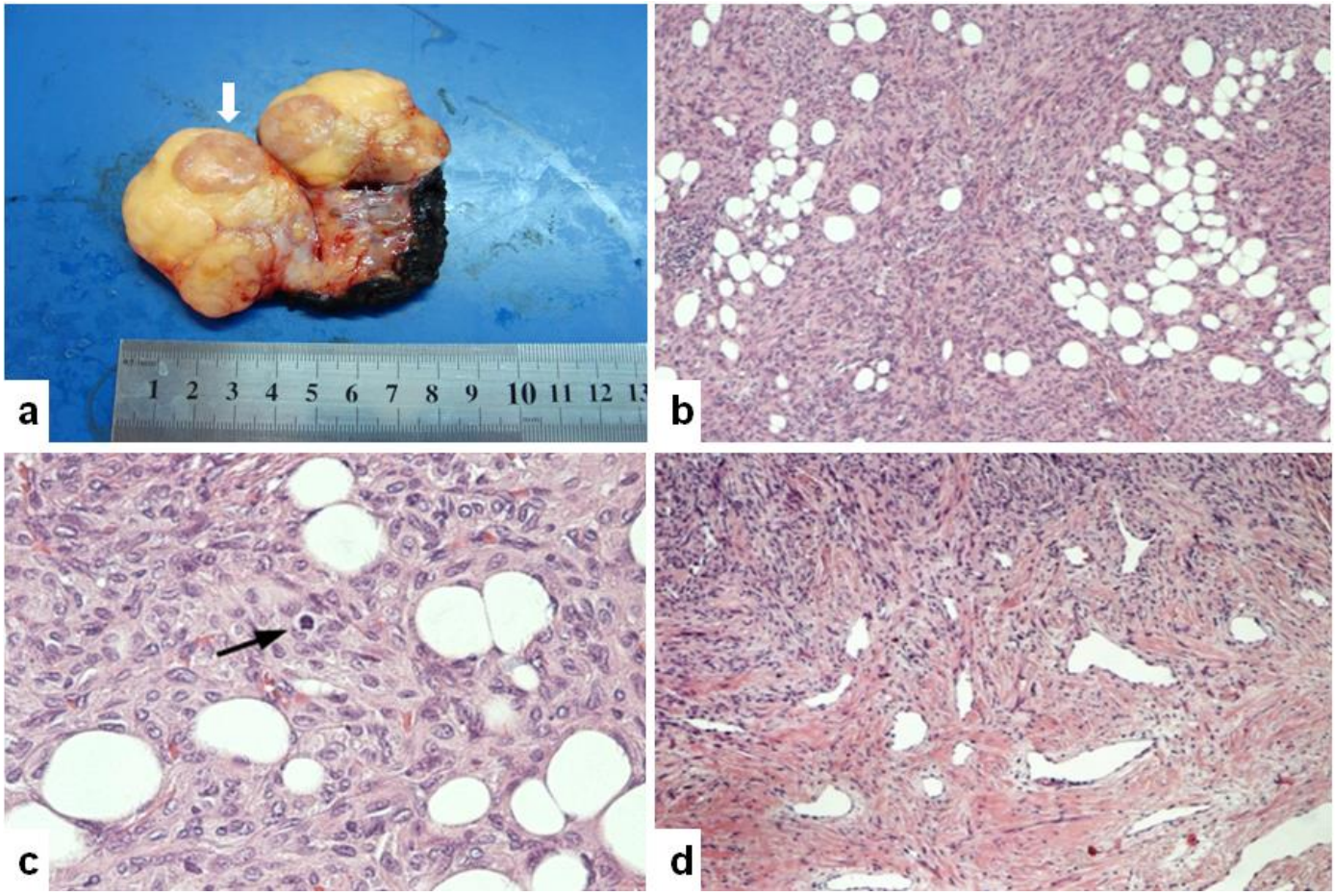

Figure 4: Malignant fat-forming solitary fibrous tumor in a 61-year-old man. Gross examination (a): the tumor showed a tan and whitish cut surface, with a firm appearance and yellowish fatty areas. Nodules of elastic consistency were present within the mass (white arrow). Histopathological findings (hematoxylin-eosin, original magnification of 100x in b and d, and 400x in c): the tumor demonstrates an intimate admixture of spindle cells and mature adipocytes (b). It shows pleomorphism and mitotic activity (black arrow in c). There are also areas with thick collagen strands and hemangiopericytoma-like vessels (d).

Incidence	Rare.
Gender ratio	1M : 1F
Age predilection	Middle-aged adults (typically between 40 and 60 yo).
Risk factors	None.
Treatment	Complete surgical excision of the tumor is the main treatment.
Prognosis	It usually shows benign histologic features and it more frequently has an indolent clinical course with good prognosis. 10-15% may show malignant morphologic features. Metastasis is very rare.
Findings on imaging	**US:** heterogeneous and predominantly hyperechoic lesion; with sound beam attenuation; usually associated to solid hypoechoic areas. **CT:** heterogeneous lesion, with solid enhancing regions and scattered low-attenuating adipose areas. **MRI:** adipose areas with high signal intensity on the T1-weighted images that decreases on the fat-suppression sequences. Solid areas are hypo, hyper or isointense on the T2-weighted images and hypo or isointense on the T1-weighted ones, usually with post-contrast enhancement.

Table 1: Summary table for fat-forming solitary fibrous tumor

Etiology	Age predilection	US	CT	MR
Fat-forming solitary fibrous tumor	Middle-aged adults (typically between 40 and 60 yo).	Heterogeneous and predominantly hyperechoic lesion; with sound beam attenuation; usually associated to solid hypoechoic areas.	Heterogeneous lesion, with solid enhancing regions and scattered low-attenuating adipose areas. Thin septations may be found. Calcification is rare.	Adipose areas with high signal intensity on the T1-weighted images that decreases on the fat-suppression sequences. Solid areas are hypo, hyper or isointense on T2-weighted images and hypo or isointense on the T1-weighted ones, usually with post-contrast enhancement. Thin septations may be found.
Liposarcoma	Middle-aged adults (typically between 40 and 60 yo).	Findings are similar to those observed in the fat-forming solitary fibrous tumor. Thick septations and cystic areas may be present. Calcification is rare.	Findings are similar to those observed in the fat-forming solitary fibrous tumor. Thick septations and cystic areas may be present. Calcification is rare.	Findings are similar to those observed in the fat-forming solitary fibrous tumor. Thick septations and cystic areas may be present. Calcification is rare.
Teratoma	Children and young adults (typically between 20 and 30 yo).	Findings may be very similar to those observed in the fat-forming solitary fibrous tumor. A cystic area is present in most of these tumors.	Heterogeneous mass; varying amount of fat and enhancing soft tissue components and multiple septations. Calcification is present in more than 50% of the tumors on CT examinations.	Heterogeneous mass; varying amount of fat and enhancing soft tissue components with multiple septations
Dermoid Cyst	Children and young adults (typically between 20 and 30 yo).	Homogeneous hyperechoic lesion; sound beam attenuation and fluid level may be present.	Homogeneous cystic lesion; thin wall; varying amount of fat and fluid, occasionally forming fat-fluid level.	Homogeneous cystic lesion; thin wall; varying amount of fat and fluid, occasionally forming fat-fluid level.

Table 2: Differential diagnoses table for fat-forming solitary fibrous tumor

ABBREVIATIONS

CT = Computed tomography
HPF = High-power fields
MRI = Magnetic resonance imaging
SFT = Solitary fibrous tumor
US = Ultrasonography

KEYWORDS

magnetic resonance imaging; MRI; helical computed tomography; ultrasonography; lipomatous hemangiopericytoma; malignant solitary fibrous tumor; fat-forming

Online access
This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/1336

Peer discussion
Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity
This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad
www.EduRad.org