Imaging Findings of Solitary Fibrous Tumors of the Gallbladder

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ABSTRACT

Solitary Fibrous Tumor (SFT) is a rare mesenchymal tumor with a higher incidence of benign than malignant, most common location in the pleura. Although this tumor has been found in other locations in the body such as the head and neck region, retroperitoneal space, and intra-abdominal omentum, SFT of the gallbladder remains extremely rare in the medical literature. In this article, we present the imaging characteristics of Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) of gallbladder SFT, thereby contributing to providing information in the study of this rare pathology.

KEYWORDS

solitary fibrous tumor; computed tomography; magnetic resonance imaging

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INTRODUCTION

Solitary Fibrous Tumor (SFT) is an uncommon mesenchymal tumor, formerly also known as Hemangiopericytoma, that develops mainly in the pleura, belonging to the group of fibroblastic neoplasms with intermediate behavior according to the World Health Organization (WHO) classification of bone and soft tissue tumors (1, 2). Although less common than the pleura, SFTs in other locations such as the thyroid, greater omentum, retroperitoneum, or pelvis have occasionally been recorded (3). On the other hand, the Solitary Fibrous Tumor of the Gallbladder (SFTG) is nearly unique in the medical literature. According to our knowledge, there are only two cases of SFTG reported in the literature and one case was reported as hemangiopericytoma which is the previous terminology of SFTG in 1983 (2, 4, 5). This neoplasm is typically insidious in clinical symptoms and imaging characteristics, which easily overlaps with other entities. Hence, definitive diagnoses in the majority of cases rely heavily on biopsy results[2]. In this article, the imaging features of SFTG on Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) are described with literature review.

CASE REPORT

A 41-year-old male patient with no medical history accidentally detected a mass in the right hypochondriac region three months before admission. He denied any pain, weight loss, or other clinical symptoms. A clinical examination of the abdomen revealed a hard and mobile area in the right hypochondrum. Further assessment of a Computer tomography (CT) scan on the chest and abdomen with contrast injection further detected a mass measuring 71 × 57mm with a uniform density before injection, lying at the level of the bottom of the gallbladder (arrow, Figure 1A) and extending downwards (star sign, Figurer 1A). The post-contrast image demonstrated the mass continuous with the base wall of the gallbladder (arrow mark, Figure 1B), extending downward with clear boundaries with surrounding structures (star mark, Figure 1B), and abundant vascular supply inside the mass (arrow Figure 1C). There were no other organ abnormalities on a chest and abdomen CT scan suggesting secondary lesions. Blood tests showed an increased white blood cell count of 16.28 G/L and normal red blood cell, as well as platelet counts with indexes of 4.62 T/L and 260 G/L, respectively. Blood biochemical tests showed normal total and direct bilirubin with indexes of 7umol/l and 2umol/l, respectively, and a normal GGT index of 32 U/I. On the hepatobiliary MRI, the T2-weighted (T2W) sequence showed a heterogeneous hypointense mass (Figure 2A arrow) with a well-defined border. The post-contrast image demonstrated heterogenously strong enhancement in the arterial phase (arrow, Figure 2B) and more contrast loading on the venous phase (arrow, Figure 2C). The mass located next to the D2 duodenum region. It showed no signs of invasion or compression of the extrahepatic bile ducts or liver parenchyma. There were no abnormal lymph nodes in the abdomen.

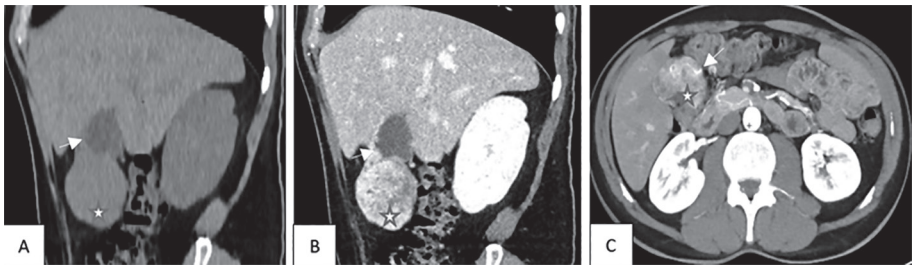

Fig 1 Image A Sagittal plane before contrast injection, tumor located at the base of the gallbladder (arrow) with relatively clear boundaries, uniform density, growing downwards (star sign). Image B Sagittal plane, portal vein phase, clearly observed tumor continuous with gallbladder base wall (arrow), strong and heterogenous enhancement (star sign). Image C Axial plane shows strong and irregular enhancement of the tumor artery (star sign); branch vessels are inside the tumor, which can be seen more clearly using the MIP reconstruction. (Maximum Intensity Projection).

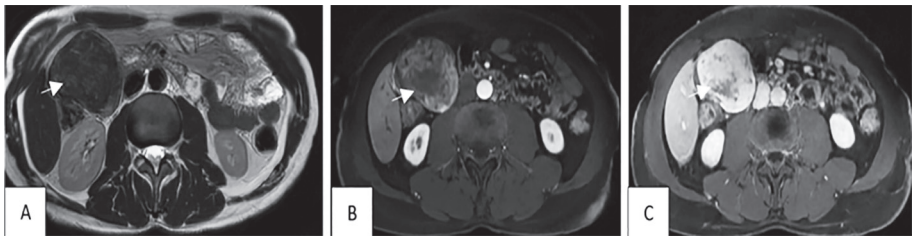

Fig 2 Image A Axial T2W sequence, the tumor has clear boundaries and a heterogeneous signal, with some hypointensity suggesting collagenous or fibrous stroma (arrow). Image B Axial T1FS arterial phase sequence, tumor enhances strongly but heterogeneously (arrow). In the image C Axial T1FS sequence in the venous phase, the tumor enhances strongly and more than in the arterial phase (arrow).

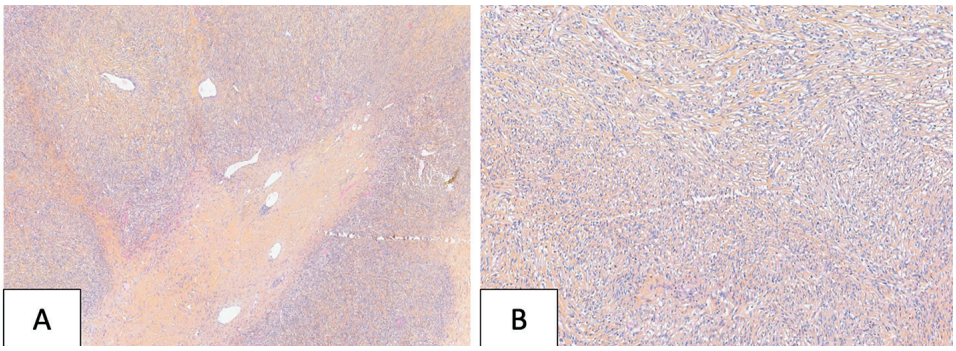

Fig 3 HPS staining. (A) HPS x250: alternation of cellular and hypocellular zone, hemangiopericytic vascularization. (B) HPS x100: monomorphic spindle cells, not atypical, low mitotic index. HPS = Hémalun, Phloxine, Safran.

After multidisciplinary consultation, the patient was treated with laparoscopic surgery to remove the gallbladder and tumor into a single mass. On postoperative pathology, microscopic tumor cell density ranged from moderate to high. Tumor cells are spindle-shaped, round-nucleated, and monomorphic, arranged in short bundles on a collagen-rich stromal base. Blood vessels behave as the hemangiopericytic type. There are areas of fibrosis, hyalinization, and poor cells. There are two divisions on ten fields with X400 magnification (field diameter is 0.62 mm). Immunohistochemical staining of anti-STAT6 antibody was strongly positive for nuclei of tumor cells. The histopathological results of the tumor concluded that it was an SFTG with a low risk of metastasis, according to Demicco risk stratification systems. The patient was stable after surgery and was discharged with regular follow-up examination.

DISCUSSION

SFT is a histologically characterized mesenchymal neoplasm originating from fibroblastic or myofibroblastic cells. It usually arises in the pleural cavity, with the first recognized case being described by Klemperer and Rabin in 1931. Until now, approximately 30–40% of SFT cases outside the pleural membrane have been reported in the medical documents (6–8). Although SFT can be found in multiple locations throughout the body, SFT in the abdominal cavity, and especially SFTG, is exceptionally rarely mentioned in medical texts. It is worth mentioning that the WHO has recently standardized the general terminology for SFT and Hemangiopericytoma using the term SFT (1–3). In general, SFT is considered a benign tumor with a rate of approximately 78–88% of cases. It typically does not significantly affect the patient's prognosis. However, there is still a rate of roughly 12–22% of SFT cases that progress into a malignant form, which can invade and metastasize. The recurrence rate of SFT is 6%, according to Decouvelaere et al. (7). Due to the malignant potential of SFT, mainly when the tumor size is > 10 cm, early diagnosis of SFT is crucial to excise the tumor and prevent its cancerous possibility. However, it still takes a huge of effort to give a definitive diagnosis of SFT preoperatively (7).

Clinically, SFT tumors outside the lung membrane commonly occur in middle-aged individuals, with an equal prevalence between males and females. They typically present a silent and asymptomatic development. However, when the tumor grows, it may manifest symptoms due to compression of nearby structures. Furthermore, less than 5% of SFT patients exhibit signs of hypoglycemia (6). Some clinical features that may occur in abdominal SFT include abdominal pain, palpable mass, and gastrointestinal or urinary symptoms due to the mass effect of the tumor (6). Two reported cases of benign include a 55-year-old female patient incidentally discovered during follow-up for breast fibroadenoma and an 83-year-old female patient admitted to the hospital due to recurrent upper abdominal pain for several months. On the other hand, the malignant case of Hemangiopericytoma of the gallbladder involved a 30-year-old female patient who was hospitalized for the development of a right upper quadrant abdominal mass over five months (2, 4, 5). In our report, a 41-year-old male patient was admitted to the hospital due to self-palpation of a painless mass located on the right lower chest wall, with no other accompanying symptoms. As a result, imaging diagnostic examinations are necessary to define the specific characteristics of SFT in general and SFTG in particular.

On ultrasound, typical SFT tumors often appear hypoechoic but occasionally with heterogeneous echoic structures corresponding to areas of myxoid degeneration within the lesion. Furthermore, despite SFT being a highly vascular tumor, color Doppler ultrasound typically does not demonstrate flow signals (8). On CT, SFT tumors frequently exhibit well-defined borders and heterogeneous density. They may occasionally demonstrate calcification, although it is rare. After contrast administration, the tumor displays highly vascular, but the central region may show decreased density, indicating necrosis or cystic degeneration within the tumor (6). In addition, determining the correlation of the tumor with adjacent structures, including signs of compression or invasion, and searching for any potential metastatic lesions, if present, is an advantage of CT in diagnosing and planning the treatment for patients. MRI with high soft tissue resolution, multiple sequences, and imaging planes defines the composition and nature of SFT better than CT and ultrasound. Nevertheless,

the features of SFT tumors on MRI can differ based on the amount of collagenous or fibrous stroma, vascularity, and cell density. On T1W images, the tumor typically exhibits intermediate signal intensity, while on T2W, the tumor shows heterogeneous low signal intensity corresponding to the collagenous or fibrous stroma component (Figure 2A). Image B T1FS Axial arterial phase pulse sequence, tumor enhances strongly but heterogeneously (arrow). Image C Axial T1FS pulse sequence in the venous phase, the tumor enhances strongly and more when compared to the arterial phase (arrow). After gadolinium injection, the tumor demonstrates strong and heterogeneous enhancement, with possible areas of non-enhancement in the core region. Additionally, the presence of tortuous blood vessels at the edge of the tumor is a visible indicator that can help confirm the diagnosis (Figure 1C) (6). Moreover, the dynamics of contrast enhancement in SFT tumors are also remarkable, exhibiting mild arterial phase enhancement, increased venous phase enhancement, and late enhancement, suggesting the existence of fibrous or collagenous stroma inside the tumor (Figure 2C) (6). Angiography is also worthy of vascular-rich neoplasms like SFT, with images showing increased vascularity and a feeding artery supplying the tumor. Dilated arterial branches within the tumor as well as the early visualization of veins (arteriovenous shunt) can also be observed (6).

Because SFTG is an uncommon lesion, it might be challenging to differentiate it from other gallbladder tumors clinically and imaging. This fact explains the unusual appearance of SFTG in the medical literature. Based on the anatomical location of the tumor in the gallbladder fundus, we identified two differential diagnoses that need to be considered in this case, including Gallbladder adenomyomatosis and Gallbladder Carcinoma. Gallbladder Adenomyomatosis is a benign lesion characterized by hypertrophy of the mucosal layer with invasion into the muscular layer, forming Rokitansky-Aschoff sinuses. It is manifested on ultrasound by comet-tail artifacts when cholesterol crystals are deposited inside. In addition, on MRI, the feature "pearl necklace sign" is notable in Gallbladder Adenomyomatosis, with increased signal intensity on T2W images, resembling the pearls on a necklace (9). On the other hand, contrast enhancement following injection is usually uneven in malignant tumors such as gallbladder carcinoma, showing an invading tendency to surrounding structures, especially the liver parenchyma, as well as the presence of metastatic lymph nodes (10).

Histopathological features are the gold standard for diagnosing SFT and distinguishing it from other types of tumors. On a histological level, SFT tumors typically grow from spindle cells and submesothelial stromal cells with a fibroblastic or myofibroblastic phenotype. However, the diagnosis can overlap with other conditions, such as mesothelioma and leiomyoma (11). The diagnosis is confirmed by immunohistochemistry, which shows that SFT tumor cells test negative for cytokeratins, S-100, and Desmin but positive for CD34, CD99, and BCL-2 (3, 11). It is important to mention that histopathological features and the Ki67 index are helpful in diagnosing malignant SFT (12).

Surgery is the primary treatment for SFT in general and SFTG in particular. Its goal is to reduce the tumor's chance of becoming malignant. Some factors positively associated with the malignancy potential of the tumor include a size greater than 10 cm and a high mitosis rate (> 4 per 10 high-power fields or > 4 mitoses per 2 mm^2) (11). Due to the SFT tumor's high vascular characteristic, performing preoperative embolization is encouraged to reduce the risk of bleeding during surgery (6). Up until now, there is no clear evidence of the benefits of treating SFT with radiotherapy or systemic therapies. However, systemic treatment should be considered for malignant cases with recurrent or metastatic lesions, even though at a low rate (11). Post-operative monitoring after surgical removal of SFT tumors is necessary, especially for malignant SFT tumors, due to the rate of local recurrence and metastasis (3). On surveillance of 81 SFT patients, author Winan found that the 5-year survival rate was 84%, with local recurrence and metastasis rates of 29% and 34%, respectively (11).

CONCLUSION

A solitary Fibrous Tumor is a mesenchymal tumor that is usually benign, while occasional malignant cases have been reported, mainly in the pleural membrane rather than other places. SFTs in the gallbladder are extremely rare. Early diagnosis and early surgical treatment are necessary to prevent the potential malignant progression of the tumor over time, thereby improving treatment effectiveness. Although the occurrence in the gall bladder is uncommon, SFT should be considered in some instances, especially when there are suggestive diagnostic features in imaging studies. On CT and MRI, some critical signs that indicate the diagnosis of SFT include well-defined borders, high-vascularity, and decreased signal on T2W imaging with more contrast enhancement in the arterial and late phases, suggesting the presence of fibrous or collagenous stroma within the tumor. In addition, the presence of tortuous dilated blood vessels in the periphery of the tumor is also a feature that helps confirm the diagnosis.

AVAILABILITY OF DATA AND MATERIALS

Data and materials used and/or analyzed during the current study are available from the corresponding author on reasonable request.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Our institution does not require ethical approval for reporting individual cases or case series. Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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