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Case Report

An infrequent case of retroperitoneal synovial sarcoma ☆

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ABSTRACT

Synovial sarcoma (SS) is an uncommon malignant tumor, ranking third in prevalence within the soft tissue sarcomas group. The vast majority of synovial sarcomas are present in the extremities, with only 15% developing in the retroperitoneal space. Retroperitoneal synovial sarcoma (RSS) is an infrequent case of SS, with only about 20 cases reported in the literature. Diagnosing RSS before treatment remains challenging because of its nonspecific clinical symptoms. The disease is often detected at a later stage, leading to additional damage to other organs as well as complicated and ineffective treatment. Consequently, the 5-year survival rate is only 20%-29%. This report introduces a case of RSS in a 19-year-old male patient with imaging characteristics on computed tomography (CT) and magnetic resonance (MR).

Introduction

Synovial sarcoma (SS) is a malignant tumor that accounts for about 10% of soft tissue sarcomas, primarily found in young people with an average age of 32 years old [1]. Despite its name, SS does not develop inside the joint capsule. This neoplasm tends to originate outside the joint capsule, around large joints, especially the knee joint, which are easily misdiagnosed as benign lesions and lead to delayed planning optimal treatment [2,3]. RSS is very rare, accounting for only 1% of SS and the primary treatment option is surgery. However, until

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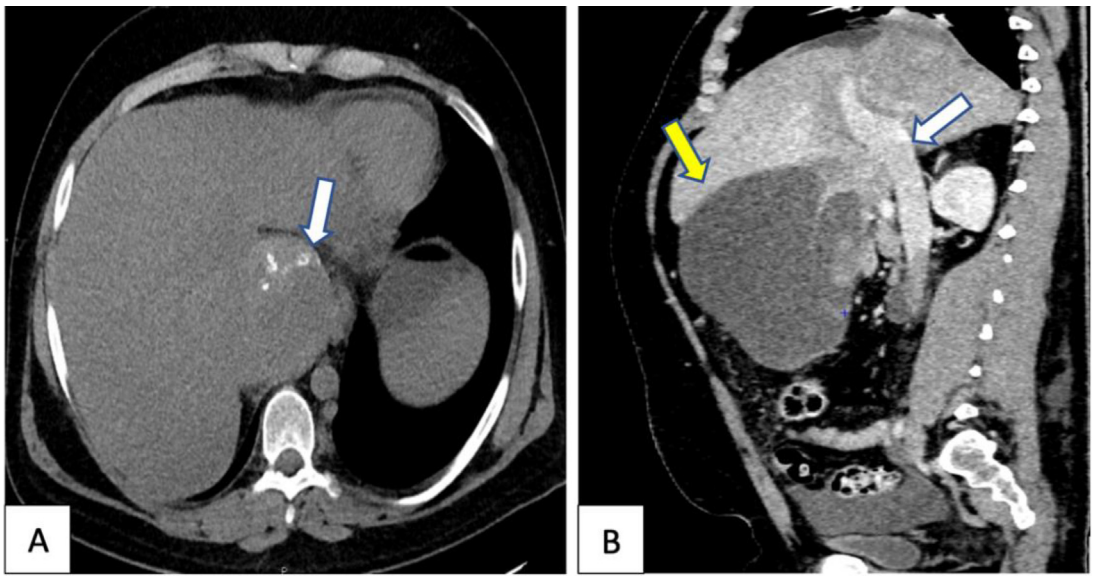

Fig. 1 – Pre-injection axial plane computed tomography (A) shows naturally hyperdense calcifications within the tumor (white arrow, image A). The venous phase sagittal plane (B) shows that the mass contacted the inferior vena cava but does not cause stenosis of the vessel lumen as well as thrombosis, the fluid-like hypodense part of the tumor compresses on the liver parenchyma but shows no signs of invasion (yellow arrow, B).

now, this disease has been associated with poor prognosis [1]. Diagnosing RSS before treatment often faces many challenges because of the diversity of retroperitoneal tumors in general, the morphological overlap between sarcoma types as well as taking the diagnostic biopsy of the lesions in the retroperitoneal space is not always easy in any case [4]. Even though a final diagnosis of RSS is mainly based on histopathological results, imaging diagnosis is vital in navigating towards a definitive diagnosis with signs such as calcification, bleeding, and fluid-fluid level. Furthermore, treatment planning also depends on the tumor's relationship to imaging with other structures in complex anatomical spaces in the retroperitoneum [5]. This report presents signs on computed tomography (CT) and magnetic resonance (MR) that help make an RSS diagnosis.

Case report

A 19-year-old male patient with no previous medical history in his personal or family history, symptoms of digestive disorders appeared many months before admission to the hospital, including loss of appetite, nausea, and vomiting. These symptoms progressed significantly 2 months before admission with marked weight loss. The patient felt dull chest and abdominal pain, mainly tightness and heaviness in the back, and dyspnea during exertion. A 3-month previous abdominal ultrasound was unremarkable. Pain relievers and anti-inflammatory drugs were used by himself, but the symptoms did not improve. After hospitalization, he underwent an emergency CT scan of the chest and abdomen. On the CT scan, a large heterogenous retroperitoneal mass measuring 240 × 195 × 143 mm was detected at the diaphragm level. This entity extended along the spine's edge, with intratumoral coarse calcification (arrow Fig. 1A), no fat density inside, and vividly heterogeneous enhancement. The mass compressed but did not invade the liver (yellow arrow Fig. 1B), contacted but did not cause narrowing of the inferior vena cava, and no thrombosis was seen (white arrow Fig. 1B). There was no sign of secondary lesions and a moderate level of abdominal fluid.

The patient had an abdominal MR imaging, which revealed a heterogeneous retroperitoneal mass close to the spine. This neoplasm includes a solid part of the mass, which strongly enhances the postcontrast image (yellow arrow in Fig. 2B). On the T2 weighted imaging (T2WI), the mass had 3 different types of signal, including the fluid part with a high signal, the bleeding part with a low signal that formed the blood-fluid level (white arrow in Fig. 2A), and the intermediate signal of soft tissue corresponding to the enhancing part in Fig. 2B. In addition, no damage to the spine or spinal canal was seen. Blood tests revealed no abnormalities. The patient underwent a biopsy under CT guidance (Fig. 3), which showed synovial sarcoma monophasic grade II, according to FNCLCC (Fédération Nationale des Centres de Lutte Contre le Cancer) (Figs. 4 and 5). After multi-specialty consultation, the tumor was not suitable for surgical removal. He was treated with Ifosfamide and Doxorubicin, analgesics, and antiemetics. However, the cancer responded poorly to treatment and increased in size rapidly. At the time of writing, after 6 months of treatment, the tumor size has increased to 293 × 244 × 205 mm. The patient is still undergoing chemotherapy treatment and is taking symptomatic relief medications such as painkillers and antiemetics.

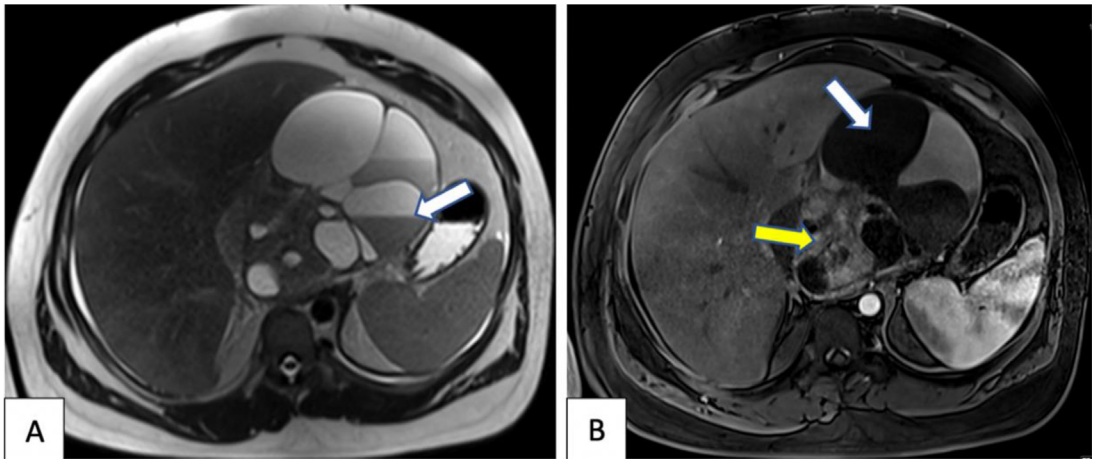

Fig. 2 – MR imaging with axial T2WI (A) and Axial T1 fatsat, post Gadolinium injection, arterial phase (B). (A) shows the tumor includes hyperintense part of the fluid and hypointense part due to bleeding, forming the fluid- blood sign (arrow), in addition the intermediate signal of soft tissue corresponding to the strong and heterogeneous enhancing part (yellow arrow, B). (B) shows the non-enhanced hypointense fluid part after Gadolinium injection (white arrow). No lesion of the spine as well as the spinal canal was seen.

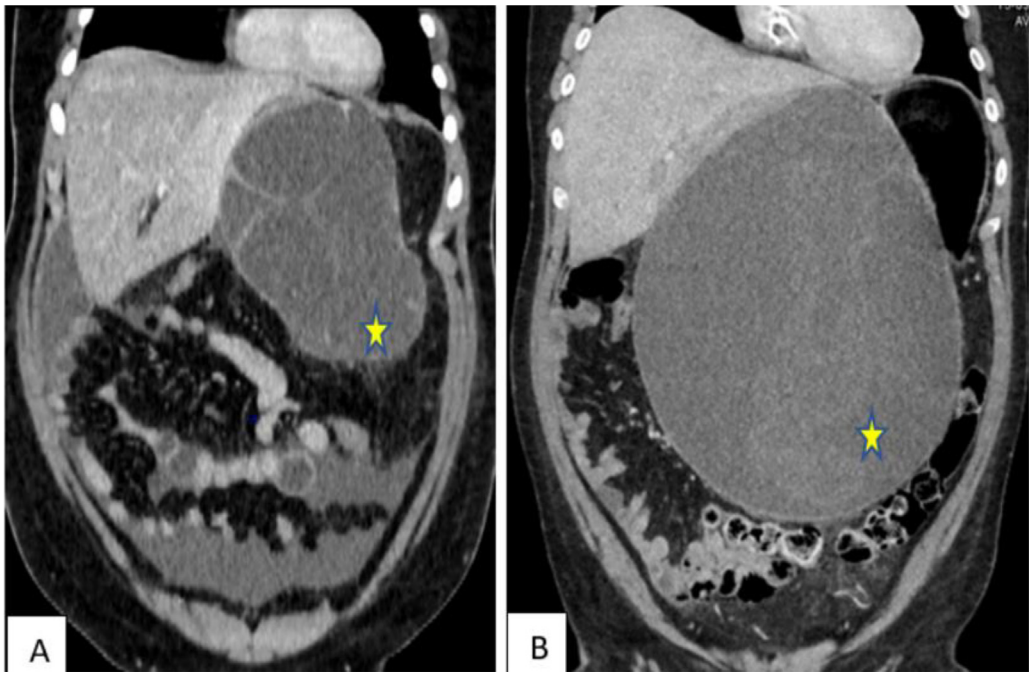

Fig. 3 – Coronal plane CT at the time of diagnosis (A) and 6 month later (B) demonstrating the tumor increases in size rapidly, growing downwards (star sign, B), this is a sign that the tumor responds poorly to treatment, poor prognosis.

Discussion

Retroperitoneal synovial sarcomas (RSS) were first described in 1954 by Pack and Tabah. In our medical literature search, only 18 cases of RSS were reported in the retroperitoneum with no difference in gender ratio [6]. Clinical diagnosis of RSS, especially in the early stages, is challenging due to the anatomical characteristics of the retroperitoneal space, which is deep and wide, allowing the tumor to grow without causing noticeable symptoms. Clinical manifestations are often vague and non-specific. They can appear many years before diagnosis [7,8] and are similar to other diseases, such as abdominal or lumbar pain, weight loss, anemia blood, and palpable abdominal mass. RSS can also be detected in the context of gastrointestinal and urinary symptoms, including nausea, vomiting, and urinary tract obstruction due to tumor pressure on surrounding structures [9]. Therefore, the immediate clinical diagnosis of RSS still requires supportive methods, including diagnostic imaging.

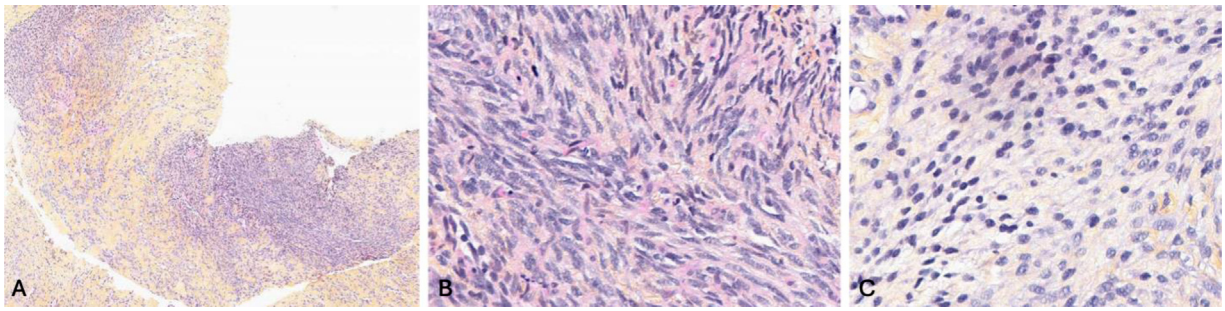

Fig. 4 – HPS staining. (A) proliferation of small spindle cells arranged in fascicles forming hypocellular zone on a fibrohyaline background and hypercellular zone (magnification x40). (B) Hypercellular zone: : monomorphic spindle cells with oval or elongated nuclei. No mitosis, no necrosis (magnification x400). (C) Hypocellular zone (magnification x400). HPS = Hémalun, Phloxine, Safran.

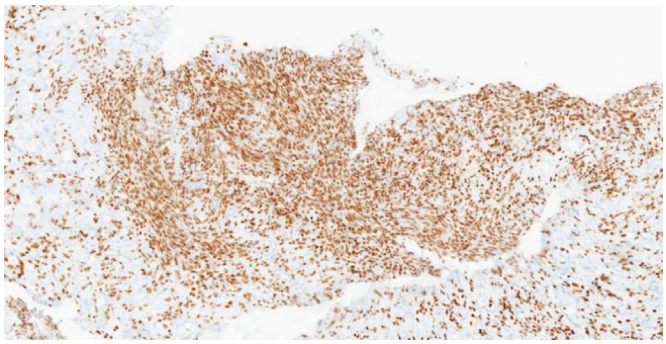

Fig. 5 – IHC staining: SS18—nuclear staining of all tumor cell (magnificationx100). IHC, immunohistochemical.

Ultrasound often has a limited role in examining retroperitoneal lesions, including RSS. CT and MRI play an essential role in determining tumor characteristics such as morphology, size, calcification, and hemorrhage within the tumor. Moreover, the relationship of the tumor with retroperitoneal structures or distant metastasis also helps differentiate RSS from other tumors and surgical planning. CT is especially useful in assessing calcifications, hemorrhage, and the degree of bone and spinal canal invasion of SSs and superior in complex anatomical spaces such as the retroperitoneum. However, calcification in SS occurs in 27%-41% of cases, in which calcification at the lesion's periphery is a diagnostic sign, while bone invasion around the tumor is found in about 25% [1,2,10]. In our case, we detected a tumor in the retroperitoneal space with a hyperdense area of calcification in the upper outer part of the tumor (Fig. 1A) without hemorrhage and bone lesions surrounding the tumor. With high resolution and multiple examination planes, MRI is considered the optimal imaging method to evaluate RSS to determine tumor characteristics and the degree of invasion of RSS with retroperitoneal structures. This modality favors assessing large blood vessels and vital organs such as the kidneys, pancreas, adrenal glands, spinal bones, and spinal canal. On T1WI, SS typically appears as a multilobular mass with heterogeneous signal due to hemorrhage, calcification, and tissue, which has a similar signal to adjacent muscle. On T2WI, the triple sign described by Jone et al. occurs in 35%-57% of SS, with three different signal types, including the intermediate signal part of solid tissue, the hyperintense part of fluid necrosis, and the hypointense part of calcification or regressive bleeding within the tumor [2]. In our case, in addition to the triad sign, we also found another sign: the fluid-blood level inside the tumor (Fig. 2A) forming a "Bowl of grapes" appearance, occurring in about 50% of SS's cases [2].

Because diagnostic signs of RSS are not 100% specific, distinguishing RSS from other tumors is necessary before planning treatment [6]. The differential diagnosis of RSS in the retroperitoneum may include liposarcoma, undifferentiated pleomorphic sarcoma, and leiomyosarcoma. Not detecting intratumoral fat on CT and MRI for liposarcoma is a vital sign distinguishing SS from liposarcoma [7]. Unlike liposarcoma, accurately differentiating undifferentiated pleomorphic sarcoma and leiomyosarcoma from RSS based on only imaging is sometimes tricky because of overlapping images between these tumors. Therefore, combining epidemiological factors and histopathological features, if necessary, along with imaging diagnosis, may help distinguish RSS from other tumors [7].

Regarding histopathology, SS is classified into monophasic, biphasic, and poorly differentiated, of which monophasic is the most common type [4]. The FNCLCC (French Federation Nationale des Centres de Lutte Contre le Cancer) system is widely accepted in grading lesions based on the combined scoring of 3 parameters: tumor differentiation, mitotic count, and tumor necrosis [2]. Regarding immunocytochemistry, SS18-SSX fusion antibody is sensitive and specific for SS,

while being negative with CD34 helps distinguish synovial sarcoma from other tumors such as solitary fibrous tumors, malignant peripheral nerve sheath tumors, leiomyosarcoma [4].

The treatment method for RSS is mainly surgical, ideally removing the entire tumor; in addition, removing surrounding structures related to RSS is also advisable [9,11]. However, surgical removal of the RSS as a whole is not always feasible due to the tumor's late stage, large size, high risk of bleeding, and complications due to the overlap of the cancer with other structures of retroperitoneum [12]. Local recurrence or metastasis after surgery is up to 80% for RSS, and 25% of RSS patients have metastatic lesions at the time of discovery, mainly in the lungs, further complicating the treatment process [6]. Chemotherapy is still controversial in terms of effectiveness. At the same time, radiotherapy is not recommended to treat RSS. In some cases, it is used with the desire to reduce tumor size before surgery [9].

Conclusion

RSS is a rare malignant tumor and presents difficulties in diagnosis and treatment. When suspecting tumors in the retroperitoneum in young patients, careful clinical examination and detection of positive signs on CT and MR, including peripheral calcifications, triple signs, and the level of blood-fluid inside the tumor as well as negative signs such as no intratumoral fat, are important factors towards the diagnosis of RSS.

Patient consent

Informed consent for patient information to be published in this article was obtained.

Author's contributions

Ho XT and Trinh AT contributed equally to this article as co-first authors. Ho XT, Trinh AT, and Nguyen MD: Case file retrieval and case summary preparation. Ho XT, Trinh AT, and Nguyen MD: preparation of manuscript and editing. All authors read and approved the final manuscript.

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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