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Case Report

CT and MR imaging of a huge retroperitoneal synovial sarcoma: A case report ☆

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ABSTRACT

Retroperitoneal synovial sarcoma is extremely rare, with only a few cases reported in the literature. The diagnosis is often made late, due to anatomical considerations, with common symptoms including low back pain and weight loss. Imaging is critical for diagnosis, often revealing a heterogenous mass with the "triple sign" and calcifications. The main differential diagnoses include liposarcoma, undifferentiated pleomorphic sarcoma and leiomyosarcoma. Surgical resection remains the treatment of choice, though it is not always feasible, which contributes to the poor prognosis. The presented case is a retroperitoneal synovial sarcoma in a 29 years old man with findings from ultrasound, computed tomography and magnetic resonance imaging.

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Introduction

Synovial sarcoma are malignant tumors arising from mesenchymal soft tissue, and accounts for 5% to 10% of all soft tissue sarcomas. It typically affects young people, and usually arise from extremities, especially the lower limb around the knee, which remains the most prevalent site [1]. Still others localizations might be involved, and retroperitoneal synovial sarcoma remains one of the rarest, accounting for only 1% of synovial sarcomas [2]. Given the large number of retroperitoneal tumors and the morphological overlap between them, it may be difficult to reach the final diagnosis [3]. Therefore, imaging is critical for ruling out differential diagnoses. Radiology also plays a role in management and follow-up, as the ability to determine the relationship between the tumor with nearby structures in the retroperitoneum and to promptly recognize local progression strongly impacts the survival rate of patients [4]. However, the prognosis remains poor with limited treatment options [5]. Computed tomography (CT) and magnetic resonance imaging (MRI) features of retroperitoneal synovial sarcoma (RSS) in a young man are reported in this article.

Case report

A 29-year-old man with no prior medical history arrived at the emergency department with severe back pain that has been escalating for a month. This issue did not improve

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with pain relief medication. This symptom did not improve with painkillers. An history of weight loss has also been reported. A physical examination revealed a large palpable left sided abdominal mass. Common laboratory blood and urine investigations were within normal range. Thus, an abdominal ultrasound has been performed, showing a large heterogenous soft tissue lesion involving all the left side of the abdomen (Fig. 1A). Significant doppler vascularity has also been found (Fig. 1B). Subsequently, the patient had an urgent CT scan of the abdomen, which revealed a huge left sided retroperitoneal mass with heterogeneous density, moderately and heterogeneously enhanced after contrast injection, containing necrotic-appearing fluid areas as well as multiple calcifications, with no detectable fatty component, measuring 190 × 133 × 227 mm. This process extends into the ipsilateral paravertebral soft tissues, from L2 to S4 (Fig. 2A), with lysis of the left transverse processes of L4 and L5 (Fig. 2B), slightly encroaching on the L4-L5 foramen. Anteriorly and laterally, it comes into contact with the anterior abdominal wall and pushes forward the left rectus and transversus abdominis muscles, without breaching the adjacent wall (Fig. 2B). It displaces the small intestine and colon medially (Fig. 2C). Posteriorly, it infiltrates the spinal and longissimus thoracis muscles as well as the iliocostalis lumborum muscle (Figs. 2B and C). It also infiltrates the left iliacus muscle, which is pushed forward. Medially, it pushes the left psoas major muscle inward and forward, and displaces the abdominal aorta, the left common, internal, and external iliac arteries to the right side. Superiorly, it reaches the lower pole of the left kidney.

Finally, an MRI has been performed, revealing a retroperitoneal mass with 3 different types of signal on T2 weighted sequence. The high signal represents the fluid part, the intermediate signal the soft tissue and the low signal the bleeding (Fig. 3A). High signal on DWI sequence with low ADC can also be noted (Figs. 3B and C). Heterogenous enhancement has been demonstrated following injection, aiding in the differentiation between the solid part of the tumor, marked by intense enhancement, and the rest of the tumor (Fig. 3D). A biopsy under CT guidance has been performed, and the ensuing histopathology demonstrated biphasic synovial sarcoma grade II, according to FNCLCC (National Federation of Cancer Centers in French). Given the clinical and radiological findings, the patient was not a candidate for surgical removal, and the treatment was based on chemotherapy and analgesics.

Discussion

Synovial sarcoma is an unusual type of soft tissue sarcoma and retroperitoneum is the most uncommon affected region, accounting for less than 1% of all cases of primary synovial sarcoma [6,7]. People of all ages can be affected, but more than half of the cases are found in adolescents and young adults [3]. Given the anatomical structure of the retroperitoneal space, RSS can remain asymptomatic until they increase in size and exert a compressive effect on neighboring organs. Thus, the diagnosis is often made late, which explains the large size of the tumor at time of diagnosis. Low back and abdominal pain remain the most typical symptoms, as seen in our case [7]. Gastro intestinal and urinary symptoms, as well as anemia and loss of weight, can also occur [5].

Imaging is critical for making a diagnosis. Although the role of ultrasound is limited in assessing retroperitoneal lesions, CT and MRI play a crucial role in determining the nature of the tumor, its relationship with adjacent structures, and the presence of metastasis. These elements are essential for differentiating it from other tumors and for surgical planning [2]. On CT, RSS commonly show a density that is lower or similar to that of muscle. Necrosis and hemorrhage are usually seen, and adjacent bone erosion and destruction may also be found [8]. According to Ho Xuan Tuan, bone destruction is seen in only 25% of cases, which confirms the rarity of our case [2]. Marginal stippled or punctate calcification are significant for the diagnosis. In our case, numerous calcifications along the upper edge of the tumor have been detected (Fig. 2A). However, extensive diffuse calcification and ossification are rarer, and are seen in only 1.4% of cases [8]. On MRI, a nonspecific heterogenous mass is usually seen, with signal intensity relatively equal to that of skeletal muscle on T1 weighted imaging, and higher than that of subcutaneous fat on T2 weighted imaging [9]. The T1 weighted sequence delineate the relationship of the tumor with neighboring structures, while the T2 weighted sequence demonstrate potential muscular invasion [5]. In 1993, jones described “the triple sign” to characterize the signal heterogeneity encountered with synovial sarcomas. This sign is explained by a mixture of solid elements, fibrotic or calcified regions and hemorrhage or necrosis. Our case showed the “triple sign” on T2 weighted sequence (Fig. 3A). In addition, intervening septa and fluid level forming a bowl of grapes appearance have also been reported. However, the calcifications usually seen on CT may not be identified on MRI,

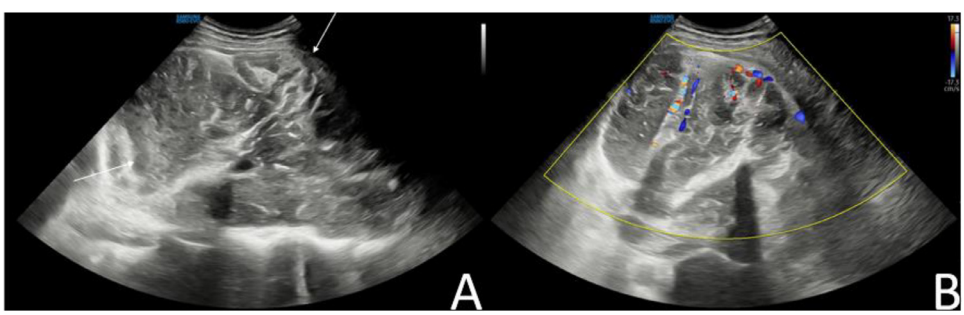

Fig. 1 – US images show a large heterogenous soft tissue mass (A, white arrow). Doppler images show tumor vascularity (B).

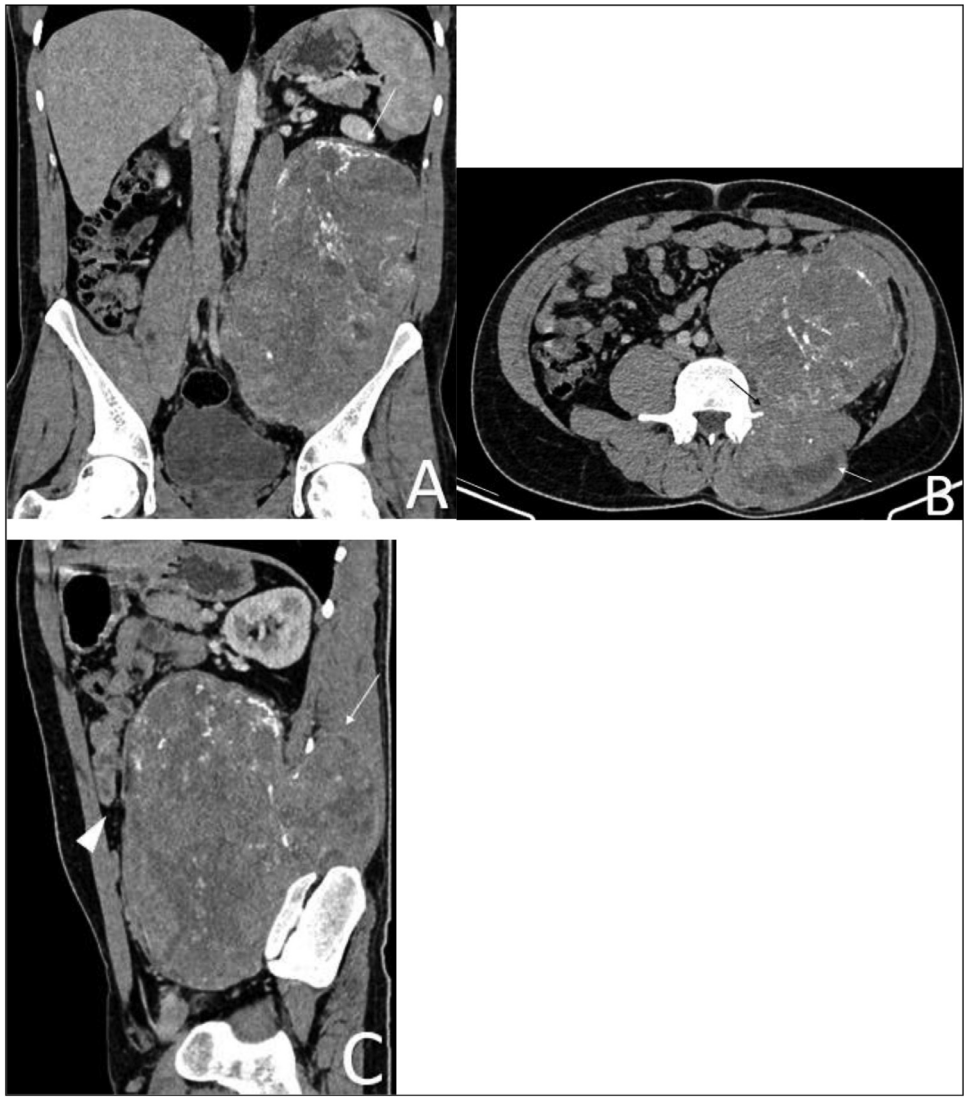

Fig. 2 – CT scan images: (A) The coronal plane shows a large heterogenous mass extending from L2 to S4, with numerous calcifications attached to the upper edge of the mass (white arrow). (B) The axial plane shows invasion of iliocostalis lumborum and longissimus thoracis muscles (white arrow); Lysis of the left transverse process of L4 is also visible (black arrow). (C) The sagittal plane shows invasion of the back muscles (white arrow), with the bowels pushed forward (white arrow head).

although larger ones may be seen as area of low signal intensity on all weighted sequences [9].

Since imaging features of RSS, as "the triple sign", are not pathognomonic, other differential diagnoses must be ruled out before planning treatment. Liposarcoma, undifferentiated pleomorphic sarcoma and leiomyosarcoma represent the main differential diagnoses. CT and MRI can easily distinguish RSS from liposarcoma by ruling out the presence of fat within the tumor. Due to the overlapping imaging features between undifferentiated pleomorphic sarcoma, leiomyosarcoma and RSS, it might be difficult to differentiate them based on imaging only. Epidemiologic and histopathologic features, along with imaging, are necessary to make a distinction [10].

Aggressive surgical resection, along with lymphadenectomy and resection of neighboring affected structures, remains the treatment of choice for RSS. There is some controversy regarding the efficacy of chemotherapy in managing this tumor. Recent reports have demonstrated some benefit of neoadjuvant chemotherapy, especially for large and high-grade tumors, by reducing the tumor size and subsequently simplifying surgery. The Implementation of radiation therapy alone, whether before or after surgery, has shown no difference in survival rates. It is used in limited cases involving large tumors, despite some potential complications. The addition of neoadjuvant chemotherapy to radiation therapy may be beneficial, according to some reports. Furthermore, the survival rate may improve following the resection of pulmonary and hepatic metastases [5]. However, the tumor's large size of most retroperitoneal sarcoma make surgery difficult and sometimes unfeasible [11]. As a consequence, the survival

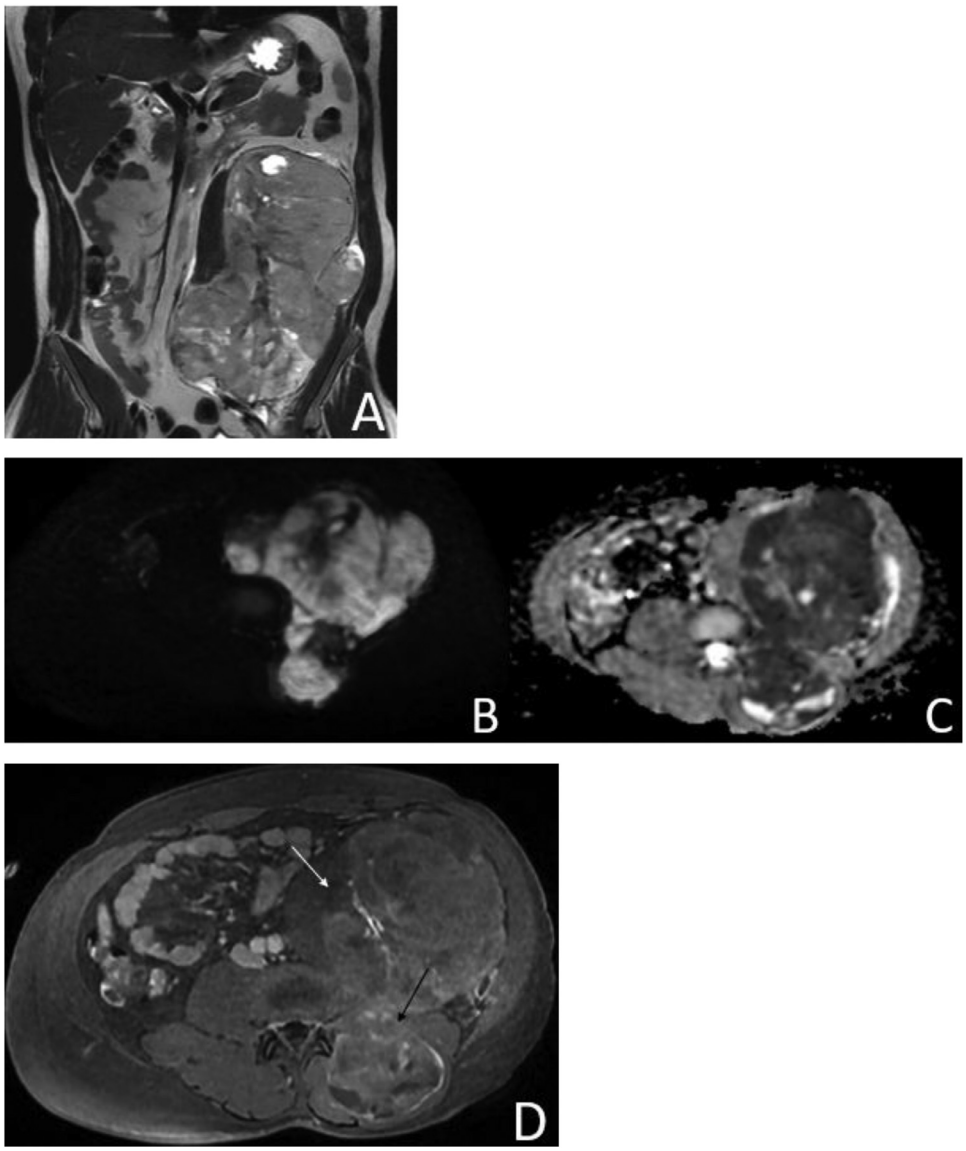

Fig. 3 – MRI images: (A) T2 coronal weighted imaging reveals a heterogenous mass with 3 different types of signal intensity also known as "triple sign". (B and C) Axial DWI and ADC show the largest part of the tumor with marked diffusion restriction and a low ADC value. (D) Axial T1 fat-sat postgadolinium injection demonstrates an area with bright enhancement (black arrow) representing the solid part of the tumor, and a nonenhanced hypointense area representing the fluid part of the mass (white arrow).

rate is quite poor, with only 20-29% of patients surviving at 5 years [5].

Conclusion

Retroperitoneal synovial sarcoma is rare tumor, mostly found in young people. The diagnosis may be difficult and is often made late, due to anatomical consideration. CT and MRI are essential for identifying the nature of the tumor, its relationship with adjacent structures and the presence of metastases, which determine the further management of the tumor. Given the difficulty of performing a complete resection, the prognostic remains very poor.

Patient consent

Informed consent for publication was obtained from patient.

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