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CASE REPORT

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A rare case report of omental synovial sarcoma complicated hemoperitoneum and literature review

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Abstract

Background Synovial sarcoma is a rare malignant soft tissue tumor originating mainly in the extremities and usually related to the joint capsule, tendon sheath, or synovial capsule. Only a few cases of synovial sarcoma arising in the abdomen, particularly the omentum, have been reported. We presented a case of omental synovial sarcoma and reviewed 7 cases of this disease.

Case presentation A 37-year-old man presented to the hospital with abdominal pain and distension for 2 months. A computed tomography scan revealed a massive heterogeneous low attenuation mass with amorphous solid components between the stomach and colon with suspected hemoperitoneum. The patient underwent surgery, and the pathological result demonstrated a greater omentum biphasic synovial sarcoma. Chemotherapy was administered with a good response. He has no signs of recurrence during 3 years of follow-up. Among 7 cases of omental synovial sarcoma, the mean age was 42, ranging from 16 to 66 years old with predominantly female (71.4%), tumor size from 9.5 cm to 20 cm. Biphasic synovial sarcoma accounted for 50%. The recurrence rate within one year is high (57.1%).

Conclusions Primary omental synovial sarcoma is uncommon and presents with nonspecific clinical symptoms, often leading to potential misdiagnosis with other conditions before surgery. They occur predominantly in females, mainly middle-aged, with a large mass size before presentation. Due to the high recurrence and mortality rate, it needs to be recognized at the early stage.

Keywords Synovial sarcoma, Malignant soft tissue tumor, Omentum

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Background

Synovial sarcoma (SS) is a mesenchymal tumor that typically arises from soft-tissue surrounding joints, bursa, and tendon sheaths [1]. The head, neck, and retroperitoneum account for 5–10% of synovial sarcomas [2, 3]. The clinical manifestation of intra-abdominal SS is nonspecific, and the symptoms vary depending on the tumor site [4, 5]. Imaging with magnetic resonance imaging (MRI) or computed tomography (CT) is helpful to characterize the tumor location, size, and relationship to surrounding structures and to guide the biopsy procedure to establish the diagnosis [6]. SS is a high-grade cancer with a 5-year survival rate of approximately 50–60% in adults [7]. The primary treatment for SS typically involves surgical resection followed by adjuvant chemotherapy [5].

There are only a few cases of primary intra-abdominal synovial sarcoma reported in the literature, particularly originating from omentum [8]. Due to the rarity, the absence of typical manifestation, and the unique imaging features of the disease, omental SS can be misdiagnosed preoperatively, as well as the ideal treatment approach for primary omental SS has not yet been clearly established. Therefore, we describe an uncommon case of primary omental synovial sarcoma complicated hemoperitoneum and reviewed 7 cases on this topic.

Case presentation

A 37-year-old man presented to our hospital with abdominal distension and mild abdominal pain in the left upper quadrant for 2 months. He denied any fevers, nausea, vomiting, diarrhea, and weight loss. His past medical history was only notable for gastritis. He was initially diagnosed with gastritis at the local hospital with mild erythema on the antrum of his stomach on esophagogastroduodenoscopy. He was treated with an empiric proton pump inhibitor; however, his symptoms were not relieved. Therefore, he went to our hospital. At initial evaluation, he was in no acute distress and had normal hemodynamics. The physical exam was remarkable for a palpable firm mass measuring approximately 10×15 cm in the left upper quadrant with distinct margins. His complete blood count and complete metabolic panels were normal. Both an upper endoscopy and colonoscopy showed no abnormalities. On CT scan, a multi-lobulated and heterogeneous mass measuring 17×18×9 cm with well-defined margins without bowel involvement occupied most of the left upper quadrant. The presence of medium hyperintense fluid suggested intra-abdominal hemorrhage. A preliminary diagnosis of a bleeding mesenteric gastrointestinal stromal tumor (GIST) or leiomyosarcoma was made, so an urgent exploratory laparotomy was performed (Fig. 1).

During surgery, a large fragile mass measuring 20×18×10 cm arising from the greater omentum with numerous proliferating blood vessels bleeding inside the tumor and the abdominal cavity was encountered. No peritoneal or liver metastases were discovered. An omentectomy en-bloc was done to take free margins. The macroscopical resulting specimen appeared as a soft, yellowish mass containing necrotic and cystic hemorrhagic materials (Fig. 2). Microscopic examination revealed hyperplastic spindle cell structures, hyperchromatic nuclei, and pseudoglandular structures. The cytology from the peritoneal washing and the resected tumor's margins were negative. Immunohistochemistry was positive for B-cell lymphoma 2 (BCL-2), vimentin, epithelial membrane antigen (EMA), cytokeratin (CK) 7, and CK AE1/3 and negative for CD99 and CD 117. The histological characteristics and immunohistochemistry confirmed the diagnosis of biphasic synovial sarcoma. This patient was discharged 10 days after surgery with a stable condition. 3 weeks later, the patient subsequently received 6 cycles of the AIM regimen (with doxorubicin, ifosfamide, and mesna). CT scans were performed every three months during the first year and once after two years for

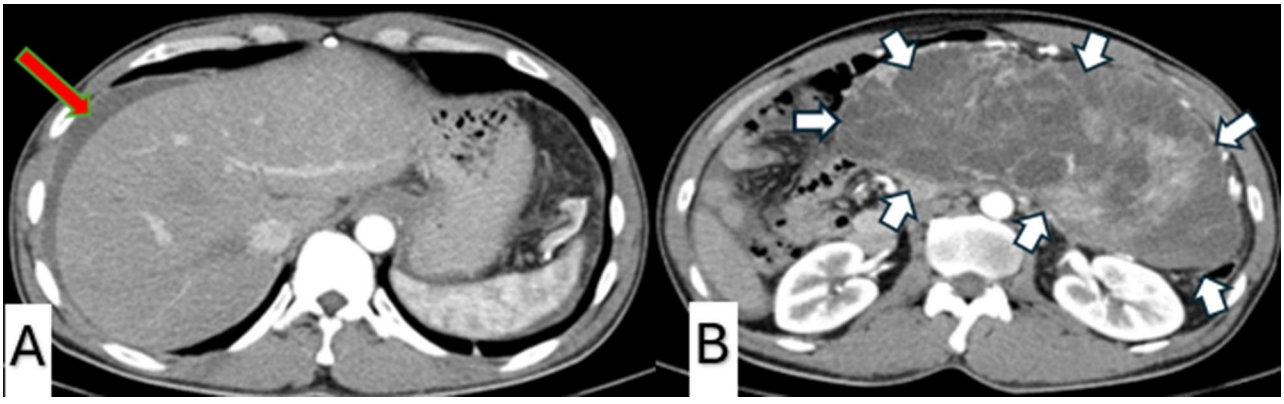

Fig. 1 Abdominal CT scan with contrast: (**A**) The free abdominal fluid near the mass and in the intraperitoneal cavity suggested intra-abdominal bleeding (red arrow); (**B**) CT with contrast showed a large, enhanced, heterogeneous, low attenuation mass in the omentum (white arrows)

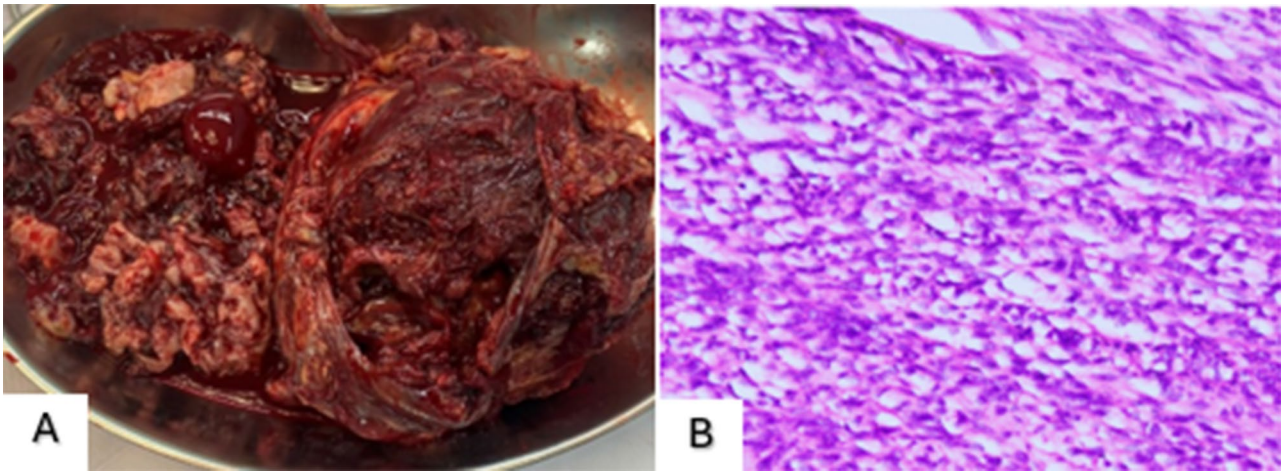

Fig. 2 Pathology of the tumor: (**A**) Image of the tumor with solid components, soft parts with bleeding sites; (**B**) Hematoxylin and eosin staining of the tumor, revealing diffused proliferation of small round and spindle-shaped tumor cells (HEx100)

Table 1 Case reports about primary omental synovial sarcoma

	References	Age (years)/ Sex	Chef complaints	Pathological result	Molecular test	Treatment	Outcome
1	Ko et al. [11]	37/female	Abdominal pain (10 cm mass)	Biphasic synovial sarcoma	N/A	Surgery: Removal of a tumor Chemotherapy	Time to progression: 6 months Survival time: at least 6 years
2	Hemmings et al. [12]	37/female pregnant	Abdominal pain (15 cm mass)	N/A	The (X;18) translocation	Surgery: removal of a tumor Chemotherapy	Time to progression: 9 months Survival time: 3 years
3	Wang et al. [13]	66/male	Abdominal distension, weight loss (20 cm mass)	Biphasic synovial sarcoma	N/A	Surgery: Removal of a tumor	Time to progression: 2 months Survival time: 2 months
4	Indranil et al. [14]	16/female	Abdominal pain, vomiting (9.5 cm mass)	Monophasic synovial sarcoma	The (X;18) translocation	Surgery: Removal of a tumor Chemotherapy	Time to progression: 13 months Survival time: 13 months
5	Iwahashi et al. [5]	51/female	Abdominal distension (12.5 cm mass)	Poorly differentiated synovial sarcoma	The fusion gene SYT-SSX was not found	Surgery: Removal of a tumor Chemotherapy Targeted therapy: Pazopanib	Time to progression: 7 months Survival time: N/A
6	Serrano et al. [15]	53/female	Abdominal pain, weight loss (19 cm mass)	Monophasic synovial sarcoma	Gene SS18 (SYT) at 18q11 was rearranged on 93% of the tumor cells	Surgery: Removal of a tumor Chemotherapy	Time to progression: N/A (> 12 months) Survival time: N/A
7	Our case	37/male	Abdominal pain, abdominal distension (18 cm)	Biphasic synovial sarcoma	N/A	Surgery: Removal of a tumor Chemotherapy	No progression until July 2024 (3 years from onset)

follow-up without any signs of recurrence. The patient has been well for three years from the onset.

Discussion

Soft tissue sarcomas (STS) originating from the mesoderm or ectoderm form a group of neoplasms with various primary sites and histological characteristics [9]. Soft tissue sarcomas account for 1% of adult malignancies [10]. Synovial sarcoma accounts for 10-15% of STS and is chiefly found in the extremities [3]. There are some cases of primary intra-abdominal synovial sarcoma reported in the literature, accounting for 3.7% of synovial sarcoma, particularly originating from omentum [8]. To the best of our knowledge, our case is the seventh synovial sarcoma arising primarily from the omentum. Among these 7 cases of omental synovial sarcoma shown in Table 1 [5, 11–15], there are 5 females (71.4%), from 16 to 66 years of age (median 37, mean 42), which was younger, but higher female proportion compared to Fisher et al. investigating intra-abdominal SS (median 46, mean 49, range

25–75 years old and 5 females/11 cases) [16]. There is one case who had a pregnancy with the omental synovial sarcoma [12]. Synovial sarcoma, particularly omental SS, predominantly affects young adults. The exact cause of SS remains unclear, and its risk factors are not well-defined likely: sex (male), age (15–50), exposure to chemical agents, radiation, chemotherapeutic drugs… However, certain genetic predispositions and molecular mechanisms like translocation t(X;18)(p11;q11) are critical in its pathogenesis [17–19]. In our case, the patient had no special risk factors other than age and gender. Unfortunately, the patient was not tested for genetic mutations.

Regarding the clinical presentation, unlike soft tissue sarcomas with painless mass, synovial sarcomas are usually painful [1, 3]. However, the symptoms are nonspecific and vary depending on the tumor site [5]. Other common symptoms include decreased appetite, weight loss, and vomiting [12]. As our results (Table 1), the most common clinical presentation is abdominal pain (5/7 cases, 71.4%), followed by abdominal distention (3/7 cases, 42.9%), weight loss (2/7 cases, 28.6%), and vomiting (1/7 cases, 14.3%). The maximal diameter of tumors varied in size from 9.5 to 20 cm (median 15 cm, mean 14.9 cm), which is quite smaller compared to Fisher et al. (median 15 cm, mean 21 cm) [16]. Due to the rarity, nonspecific symptoms, and atypical imaging findings of the disease, none of the cases were preoperatively diagnosed as synovial sarcomas. In our case, the differential diagnoses were a bleeding mesenteric gastrointestinal stromal tumor (GIST) or leiomyosarcoma. In some cases, the tumor was misdiagnosed as ovarian cancer, an ectopic pregnancy, or multicystic mesothelioma, particularly in female patients [5, 8, 12, 15]. Preoperative evaluation should include both a biopsy and imaging if there is no need for emergency surgery.

Radiology methods help determine the presence of a tumor, its size, its complications, and its invasions to the nearby organs. A slightly hyper-intense mass may be shown on T1 of MRI while hypo-dense findings on CT scan. Significant heterogeneity and enhancement strongly indicate SS on both MRI and CT scans [6]. In primarily anatomical synovial sarcoma other than the extremities like intra-abdomen, lung, etc., the definitive diagnosis still requires pathological methods [20, 21].

There are three subtypes of synovial sarcoma: monophasic type, comprised of spindle-shaped cells (up to 60% of SS); biphasic type, comprised of both epithelial and spindle cells (approximately 25% of all SS); and poorly differentiated type with rhabdoid features and a worse prognosis [12, 15]. Immunohistochemistry significantly contributes to diagnosing SS and excluding other potential diagnoses, depending on the location. In many cases, Vimentin, EMA, and BCL2 are positive, while h-caldesmon, SMA, CD34, and CD117 are negative indicators [16]. These findings are similar to our case. In addition, the detection of the translocation t(X;18) (p11;q11) can prove highly beneficial for the diagnosis of synovial sarcoma with poorly differentiated tumors [17]. GIST is the most common gastrointestinal mesenchymal tumor, which is typically considered the primary differential diagnosis for synovial sarcoma. Coexpression of CD34 and C-KIT were shown in GIST, whereas both markers are negative in synovial sarcoma. Table 1 also shows that biphasic type accounts for 3/6 cases (50%), followed by monophasic type (2/6 cases, 33.3%) and poorly differentiated type (1/6 cases, 16.7%). The chromosomal translocation t(X;18) is detected in 3/4 cases (75%), while one case has not detected this type of rearrangement, and the rest have not tested the molecular panel.

The standard treatment approach is resection of the entire tumor with negative margins for localized disease. Unplanned surgery increases the risk of local recurrence due to the association with positive margins [1]. This outcome is explained by the lack of complete preoperative staging [2]. Chemotherapy with an anthracycline-ifosfamide combination is recommended in high-risk localized tumors and advanced-stage patients [5, 17]. In our case report, there was a high risk of recurrence, including tumor size > 10 cm and urgent operation as intra-abdominal bleeding. Adjuvant chemotherapy with the AIM regimen was administered in six cycles because of its effectiveness in improving recurrent time and overall survival [22]. Despite the aggressive adjuvant chemotherapy, the risk of progression is high, ranging from 28 to 36% [23]. Molecular targeted agents like pazopanib are indicated postoperatively as second-line therapy after progression with chemotherapy [4, 5, 24]. Radiotherapy is indicated in patients with positive margins, a resection margin < 2 cm, or a histological high-grade (G2-3) tumor [3]. Table 1 demonstrates that 7/7 cases (100%) of patients underwent surgery, 6/7 cases (85.7%) for chemotherapy, and only one case was treated with the targeted therapy (pazopanib) because of the progression after 2 steps of chemotherapy. The recurrence rate within one year in patients with omental synovial sarcoma is 4/7 cases (57.1%), 2/7 cases showed no progression. Time to progression lasts at least 2 months. The survival time is not fully described in our results. In our case, no progression has been discovered after 3-year follow-up. Compared to the outcome with other rare intra-abdominal cases of SS, according to Fisher C et al., local recurrence and metastasized within the abdomen were 9/10 cases (90%) in the period of 4 to 48 months. The average survival time was only 17 months (8/10 cases died, with the survival from 4 to 36 months). Two patients remained alive with recurrent or metastatic disease at 43- and 48-months post-diagnosis, respectively [16]. There are a

few cases with good outcomes; however, the recurrence and mortality rates were still high.

Conclusion

Primary omental synovial sarcoma is quite rare, with a nonspecific clinical presentation; therefore, it can be preoperatively misdiagnosed with other diseases. The young doctor should place this disease on the differential list, which can help detect it in the early stage with better outcomes. We have described 7 cases of omental synovial sarcoma, which occurs in both sexes, mainly in middle age with a large size of mass before presentation. Treatment requires a multidisciplinary approach. Surgery is the cornerstone to remove the entire primary tumor. However, the recurrence rate of this disease is a high local extension of the disease.

Abbreviations

SS	Synovial sarcoma
CT	Computed tomography
MRI	Magnetic resonance imaging
EMA	Epithelial membrane antigen
GIST	Gastrointestinal stromal tumor

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None declared.

Author contributions
Conceptualization, design, methodology, and data analysis: NHT, DTH, and VTTA; investigation and writing-original draft: NHT, DTH, and VTTA, supervision and manuscript review: NVM, KMS. All authors reviewed the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
This is a case report that does not need ethics approval.

Informed consent
Written consent has been obtained from the patient.

Competing interests
The authors declare no competing interests.

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