

## Original Article

# Primary intra-abdominal synovial sarcoma: a case report

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Received January 3, 2017; Accepted March 24, 2017; Epub April 1, 2017; Published April 15, 2017

**Abstract:** Background: The primary intra-abdominal synovial sarcoma is rare while the mortality is relative high. There is currently no effective treatment except surgery for this type of sarcoma. Case presentation: We report a 66-year-old female patient with intra-abdominal synovial sarcoma. A huge mass lesion was found in the abdomen. Computed tomographic (CT) scanning and pathological examination cannot determine the origin of tumor. The histologic examination of detected biphasic pattern comprised epithelial and spindled cells in mitotic figures, which was consistent with synovial sarcoma. However the origin of the tumor is uncertain. Conclusion: Intra-abdominal synovial sarcoma has unique characteristics that need to be further investigated in order to understand the underlying molecular mechanism, pattern of development and metastasis, and thereby establish effective treatment strategy.

**Keywords:** Intra-abdominal synovial sarcoma, origin, immunohistochemistry, primary

## Introduction

Synovial sarcoma (SS) is a malignant mesenchymal neoplasm with variable epithelial differentiation that mainly occurs adjacent to joint capsules' bursal and tendon sheaths [1]. It is the fourth most common type of soft-tissue sarcoma, following malignant fibrous histiocytoma, liposarcoma, and rhabdomyosarcoma. Only a small proportion of the cases occur in several unusual locations, such as head and neck region, mediastinum, nerves, blood vessels, abdominal wall, skin, prostate, kidney [2], heart [3], gastrointestinal tract [4-6], larynx and hypopharynx [7], and liver [8]. Intra-abdominal synovial sarcoma is rare, and only a few cases have been reported in the literature. Primary intra-abdominal SS has been reported as a rare case in several studies [9-12]. A rare case of intra-abdominal SS was described in the present report. The pathologic examination and clinical characteristics are briefly discussed.

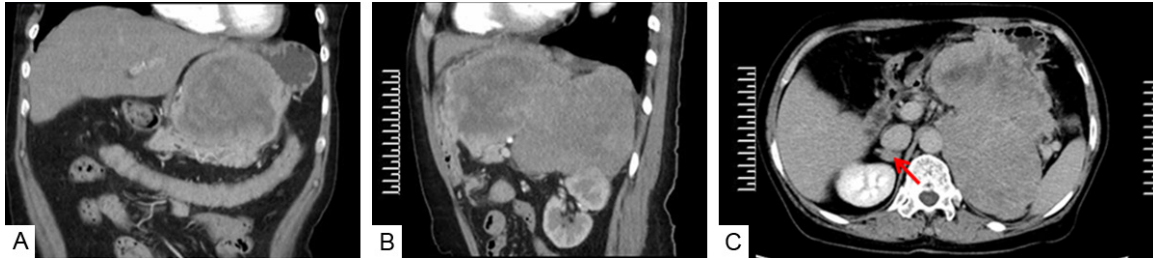
## Case presentation

A 66-year-old female patient was complained of occasional and aspecific abdominal disten-

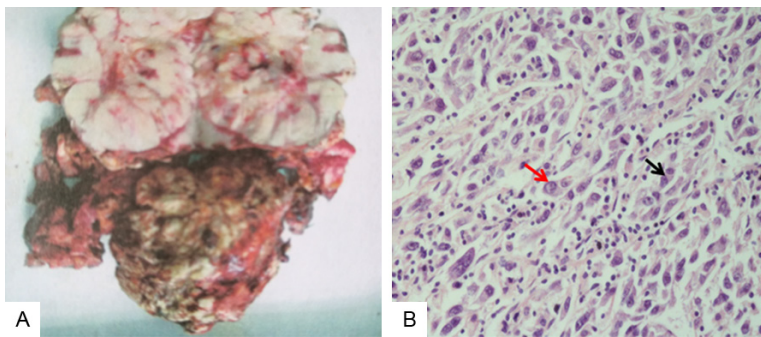
tion for two months. A history of cholecystectomy for gallbladder stone and hypertension was noted. The examination was negative except for an immobile mass at 11×16 cm arising from the left upper abdominal cavity. Computed tomographic (CT) showed an irregular solid mass in the left upper abdominal cavity (**Figure 1**). The upper bound was located between the left lobe of the liver and stomach fundus, while the lower bound was located in the upper left of the kidney. Another nodule was near to the right adrenal at about 1.8×1.3 cm and was found with well-defined boundary. It was diagnosed as gastrointestinal stromal tumor with highly suspected left adrenal tumor upon imaging. In addition, the right side of the adrenal nodule was considered to be metastasis. The laboratory findings included aldosterone (recumbent position), 223.68 pg/mL (normal range 30-160 pg/mL); aldosterone (standing position), 241.82 pg/mL (normal range 70-300 pg/mL); cortisol (8:00 am), 853.10 nmol (normal range 171-536); cortisol (4:00 pm), 642.50 nmol; and cortisol (0:00 am), 252.2 nmol.

In the surgery, the tumor was found to invade the left colon mesentery and posterior gastric

## Primary intra-abdominal synovial sarcoma



**Figure 1.** CT scan showed a huge irregular solid and inhomogeneous enhancement mass in the left upper abdominal cavity. A. The upper bound was located between the left lobe of the liver and stomach fundus. B. The lower bound was located in the upper left kidney, and the largest cross-sectional area was about 16.5×11 square centimeters. C. A nodule (red arrow) located adjacent to the right adrenal with well-defined boundary. The size was about 1.8×1.3 cm.



**Figure 2.** A. The gross specimen comprised a solid gray red mass at about 20×14×14 cm in size, exhibiting a coated surface and nodular appearance with gray section. B. Histologic examination shows a biphasic pattern comprised epithelial cell (red arrow) and spindled cells (black arrow) in mitotic figures (11/10 high-powered fields) together with lymphocyte infiltration.

body. The tumor extended to the medial margin of abdominal aorta, diaphragm, costophrenic angle, splenic portal vessels. The shape of tumor was irregular and the surface was covered by capsule. The maximum diameter of the tumor was 20 cm. According to operative exploration, a total gastrectomy + distal pancreatectomy + splenectomy + peritoneal tumor resection and reconstruction of digestive tract were performed.

The gross specimen comprised a solid gray red mass at about 20×14×14 cm in size. It exhibited a coated surface and nodular appearance with gray section (**Figure 2A**). Histologic examination showed a biphasic pattern comprised epithelial and spindled cells in mitotic figures (11/10 high-powered fields) together with lymphocyte infiltration (**Figure 2B**). Immunohistochemical staining demonstrated that the spindle cells were positive for epithelial membrane antigen (EMA), cytokeratin (CK), vimen-

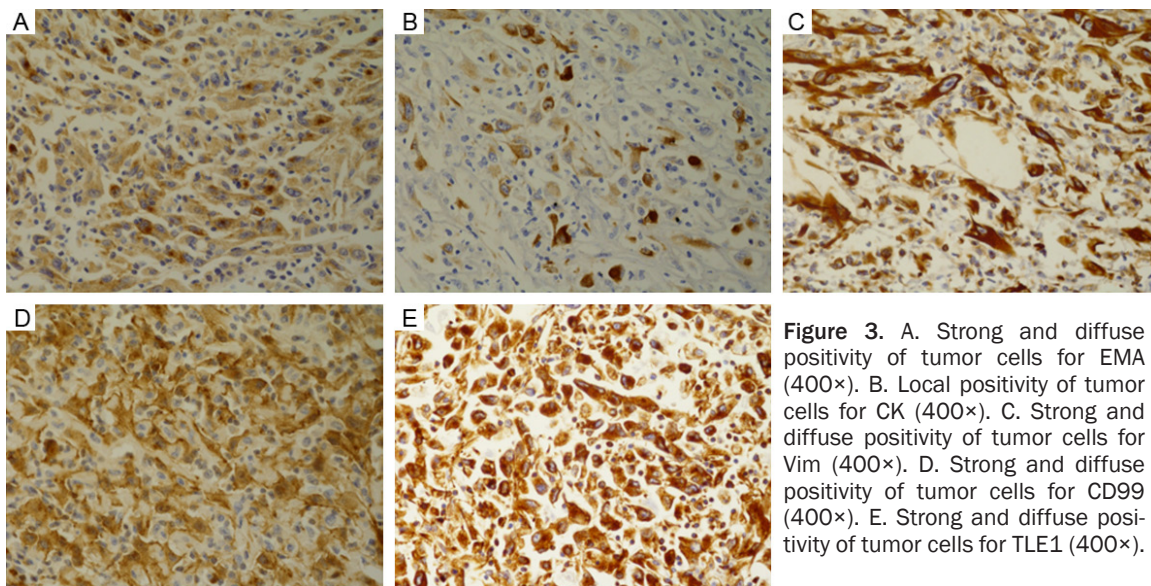
tin, cluster of differentiation 99 (CD99), and transducin-like enhancer protein 1 (TLE1) (**Figure 3**). Moreover, the cells were negative for CD117, Dog-1, CD34, S-100, desmin, and bcl-2 (**Figure 4**). These findings help the diagnosis as synovial sarcoma invading the serosa of stomach, deep muscular layer and pancreatic capsule. The lymph nodes around gastric wall and the greater and lesser curvatures were also invaded (1/1, 1/3).

The patient was diagnosed as intra-abdominal synovial sarcoma with uncertain tumor origin. After operation, the patient was hospitalized for 10 days and then discharged. However, 1 month later, she was readmitted to our hospital due to severe abdominal distension and pain. Abdominal CT indicated recurrence of multiple nodular lesions over the abdominal cavity and wall, the right adrenal nodules were considered to be metastasis. The patient died 10 days later due to poor and worsening condition.

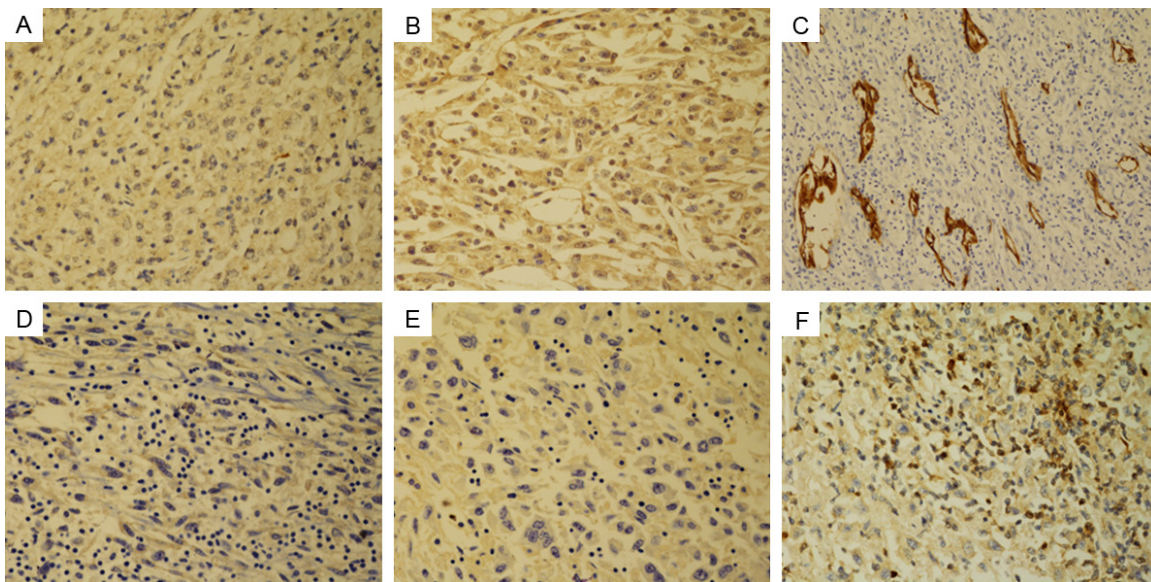
### Discussion

SS is a type of mesenchymal spindle cell tumor, which shows a certain degree of epithelial differentiation (including glandular components) and chromosomal abnormalities t(x;18) (p11.2,q11.2). According to the number and differentiation degree of immature tumor cells, epithelioid cells, and spindle cells in the tumor, SS could be classified into four subgroups, including biphasic SS (BSS), monophasic epi-

## Primary intra-abdominal synovial sarcoma



**Figure 3.** A. Strong and diffuse positivity of tumor cells for EMA (400×). B. Local positivity of tumor cells for CK (400×). C. Strong and diffuse positivity of tumor cells for Vim (400×). D. Strong and diffuse positivity of tumor cells for CD99 (400×). E. Strong and diffuse positivity of tumor cells for TLE1 (400×).



**Figure 4.** A. Immunonegative of tumor cells for CD117 (400×). B. Immunonegative of tumor cells for Dog-1 (400×). C. Immunonegative of tumor cells for CD34 (400×). D. Immunonegative of tumor cells for S-100 (400×). E. Immunonegative of tumor cells for desmin (400×). F. Immunonegative of tumor cells for bcl-2 (400×).

thelial SS (MESS), monophasic fibrous SS (MFSS), and poorly differentiated SS (PDSS) [13].

At first the present case was mistaken for gastrointestinal stromal tumor or adrenal tumors. The pathologic examination was carried out to confirm the location of the tumor. Primary intra-abdominal SS is rare with only several case reports. Except for a subset of monophasic tumor, most of synovial sarcoma shows posi-

tive immunohistochemical staining with EMA, CK, vimentin, CD99, and TLE1. Synovial sarcoma is immune negative for CD34, desmin, and SMA [14]. CK7 and EMA can be used as important indicators for the differential diagnosis of synovial sarcoma [15]. Gastrointestinal stromal tumor (GIST) is one of the most common tumors that needs to be distinguished from intra-abdominal SS. CD117 (c-Kit) has been proved to be a reliable and sensitive biomarker for GIST. Dog-1 is mainly used in the diagnosis of

Kit-negative gastric epithelioid GISTs [16]. In addition, CD34-positivity is found in the great majority of gastric spindle cell GISTs [17], whereas it is excluded from SS [18]. We present the tumor which was immunopositive for EMA, CK, vimentin, CD99 and TLE1, whereas immunonegative for CD117, Dog-1, and CD34 thus ruling out the possibility of GIST. In addition, it was also needed to identify adrenal tumor, malignant sarcomatoid mesothelioma, leiomyosarcoma, solitary fibrous tumor, lymphoma, dendritic cell tumor, and malignant peripheral nerve sheath tumor (MPNST). Normal intraoperative adrenal gland helped us to exclude adrenal tumor. Negative staining for CR, WT-1, CD34, LCA, CD20, Pax-5, CD30, ALK, CD45RO, CD3, CD21, CD23, S-100, CD1a assisted us for further differential diagnosis. However, the origin of tumor was uncertain.

SS is an aggressive tumor since more than half of the reported SS recurs locally within 2 years. Metastasis commonly occurs in the lung, and less to the lymph node and bone. All patients with primary retroperitoneal synovial sarcoma died (at about 7-24 months) with local recurrence or extension, but none of the patients metastasized outside the abdomen. In addition, metastatic SS limited in the abdomen was associated with positive desmin, which may mimic myogenic tumor. The prognostic factors include age of the patient, mitotic activities, and margin-free resection. However, the impact of factors on prognosis such as the histological subtype and the variants of SSX gene involving the translocation was still under debate.

Surgery is still the main method for the treatment of intra-abdominal synovial sarcoma. Due to late diagnosis and no adequate effective adjuvant treatment, the prognosis of intra-abdominal synovial sarcoma remains poor. A tumor size larger >5 cm with a presence of necrosis and a high mitotic rate are thought to be poor prognostic factors [19]. In this case, local recurrence in the abdominal cavity and wall developed 1 month after complete resection of the primary tumor. However, the origin of the tumor still cannot be confirmed.

### Conclusion

SS is a high-grade malignancy with highly metastatic potential; efficient and timely diagnosis of SS may impact treatment.

### Disclosure of conflict of interest

None.

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## Primary intra-abdominal synovial sarcoma

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