Primary Intra-abdominal Synovial Sarcoma

Yzu-Jen Wang[1]*, Shi-Chi Wen[1], Shang-Tao Chien[2], Jin-Wen Sheu[3], Chao-Wen Hsuea[1], Nan-Hsiung Feng[4]

[1]Division of Gastroenterology, Department of Medicine, and Departments of [2]Pathology, [3]General Surgery, and [4]Medicine, Kaohsiung Military General Hospital, Kaohsiung, Taiwan, R.O.C.

We report a case of primary intra-abdominal synovial sarcoma of the omentum in a 66-year-old man hospitalized for intermittent abdominal fullness for 1–2 months and tenesmus for 2 weeks. The patient had a palpable mass that was solid, hard and with well-defined thickness within his abdomen. A huge heterogeneous mass lesion over the middle abdomen that started from S2, S3 of the liver to the transverse colon was shown on abdominal computed tomography. The major cell types of the tissue mass were confirmed to be spindle and epithelial cells, which was consistent with biphasic synovial sarcoma according to pathologic and immunohistochemical findings. [*J Chin Med Assoc* 2006;69(10): 492–495]

Key Words: intra-abdominal, synovial sarcoma

Introduction

The term synovial sarcoma refers to morphology that resembles developing synovium,[1] and it is a malignant mesenchymal neoplasm that mainly occurs in the vicinity of joint capsules' bursal and tendon sheaths.[2,3] It usually develops among young adults, and the para-articular regions are the most diagnosed sites.[4,5] According to previous reports,[2,5–7] only 5–10% of synovial sarcomas are in the head and neck, mediastinum, abdominal wall, esophagus and retroperitoneum. To date, synovial sarcomas have also been found to occur in several unusual locations, including skin, blood vessels, nerves, mediastinum, pleural cavity, prostate and kidney.[8–14] Primary intra-abdominal synovial sarcoma has been mentioned as a rare case in several reports.[5,15–17] We describe an unusual case of primary intra-abdominal synovial sarcoma that was discovered in the omentum, and its pathologic and clinical features are briefly discussed.

Case Report

A 66-year-old man was admitted to Kaohsiung Military General Hospital due to intermittent onset of abdominal fullness for 2 months and tenesmus for 2 weeks. A history of hypertension and alcohol consumption for more than 30 years was noted. On physical examination, a palpable mass measuring about 10×15 cm in the left abdomen, with hard constitution and definite margin, was detected. The pulse rate and blood pressure were 82/min and 140/82 mmHg, respectively. Complete blood cell count showed: hemoglobin, 9.7 g/dL; mean corpuscular volume, 89.2 fL; white blood cell count, 15.3×10^3 cells/μL; polymorphonuclear leukocytes, 69.4%; lactate dehydrogenase, 356 U/L.

Computed tomography (CT) showed a huge heterogeneous lower attenuation mass with an amorphous solid component and a thin capsule; it was situated between the stomach and colon. In addition, the lesion was over the middle abdomen, extending from S2 and S3 of the liver to the transverse colon (Figure 1A). Body fluid near the mass and in the intraperitoneal cavity was presumably hemoperitoneum due to tumor bleeding (Figure 1B). The associated symptoms in this patient were body weight loss (5 kg within 2 months), lack of appetite, reduced stool passage and increased urination frequency.

A huge fragile mass ($20 \times 20 \times 10$ cm) with rupture of the capsule occupying the upper abdomen with compression of the stomach was found intraoperatively. The tumor mass was bulging out from the anterior wall and body of the stomach: the lesser curvature part

*Correspondence to: Dr Yzu-Jen Wang, Division of Gastroenterology, Department of Medicine, Kaohsiung Military General Hospital, 2, Chung-Cheng 1st Road, Kaohsiung 802, Taiwan, R.O.C.
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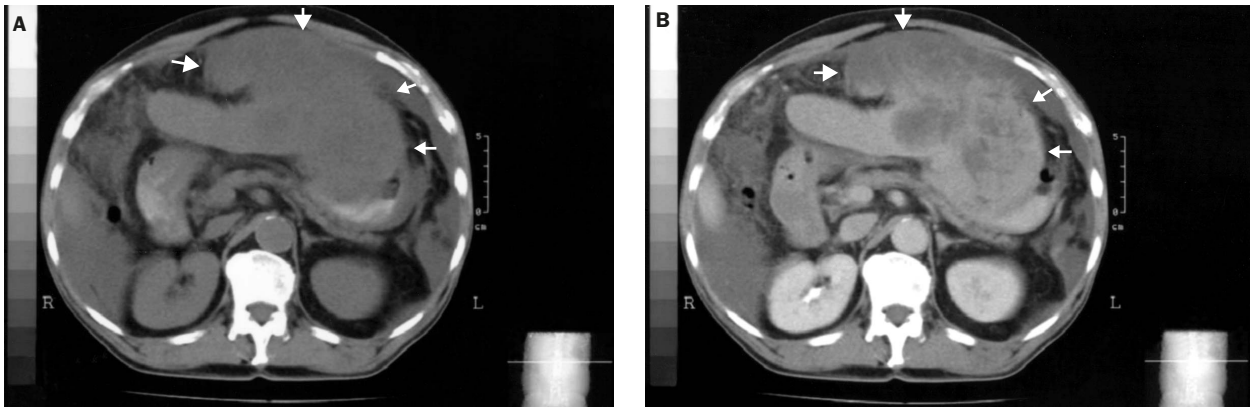

Figure 1. Synovial sarcoma: (A) noncontrast computed tomography (CT) shows a huge, low attenuation mass in the omentum (arrows); (B) postcontrast CT shows an enhanced, heterogeneous lesion involving the inferior liver and anterior stomach (arrows).

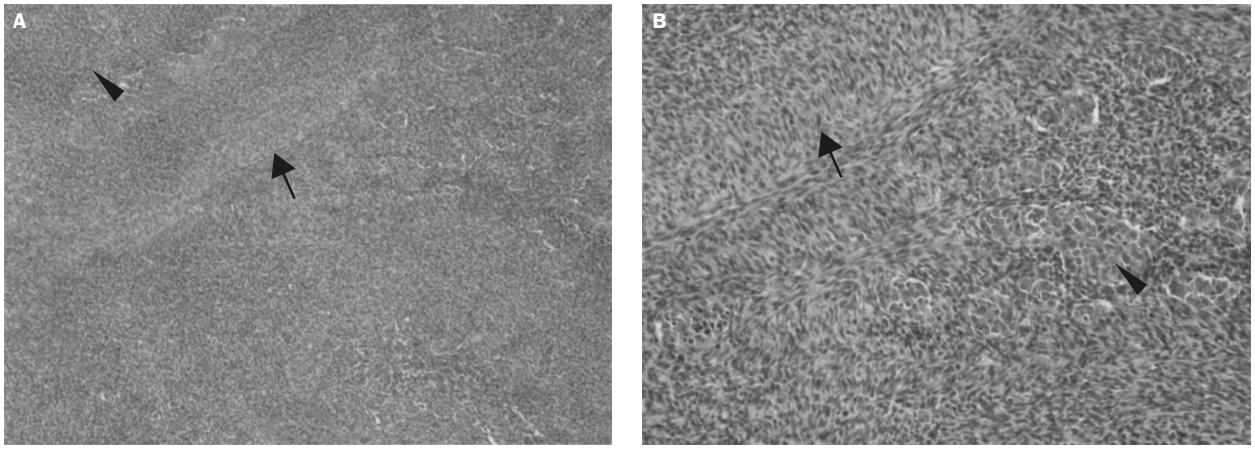

Figure 2. Histology shows that the tumor has a biphasic pattern. The sarcomatous component demonstrates fascicular growth of spindle cells featuring a herring-bone pattern (black arrows). The epithelial component is in rounds nests of epithelial cells with ovoid nuclei and abundant cytoplasm (arrowheads). (A) 40×; (B) 100×.

directly invaded into the lateral segment of the liver and downward to extend below the umbilicus without invasion of the colon and mesocolon. A nodular lesion about 1 cm in diameter seeding over the surface on the left lateral segment of the liver and multiple nodular lesions seeding over the surface of the left diaphragm were cleared. Moreover, liver cirrhosis was also diagnosed. Left lateral segmentectomy of the liver, subtotal gastrectomy with Billroth's II anastomosis and extensive resection of the left diaphragm tumor were performed.

The gross specimen comprised resected stomach, omentum and liver. Histologic examination demonstrated a biphasic pattern with proliferative sarcomatous cells arranged in abortive glands or rounded nests among fascicular growth of spindle cells featuring a herring-bone pattern (Figure 2). Immunohistochemical staining showed that the major cell types were spindle and epithelial cells; furthermore, the tumor cells also possessed histologic similarity of epithelial membrane antigen (EMA) and cytokeratin. The spindle cells were positive for bcl-2, negative for smooth muscle actin (SMA), S-100, CD117, hepatocyte antigen and calretinin. These findings helped us to exclude leiomyosarcoma, malignant peripheral nerve sheath tumor (MPNST, malignant schwannoma), gastrointestinal stromal tumor (GIST), hepatic cellular carcinoma and mesothelioma. The diagnosis of synovial sarcoma was confirmed and documented.

After operation, the patient was hospitalized for 10 days and then discharged. However, 2 months later, he was readmitted to our emergency room due to consciousness disturbance and severe abdominal distension and pain. Abdominal CT found recurrence of multiple nodular lesions over the peritoneal area, perirectal area, left lateral region of the urinary bladder and mesenteric area. In addition, multiple lymphadenopathies over the right external iliac lymph chain and massive ascites were found. The patient died 10 days later due to poor and worsening condition.

Discussion

Intra-abdominal synovial sarcoma is rarely found. Immunohistochemistry is useful for diagnosing and distinguishing synovial sarcoma from other malignancies. Most synovial sarcomas are focally positive for cytokeratin and EMA. It has recently been suggested that EMA, cytokeratin AE1/AE3, and E-cadherin, in combination with CD34 negativity, are the most useful and sensitive protein biomarkers for diagnosing monophasic fibrous pattern, and also good for diagnosing the scantily differentiated synovial sarcoma.[18] In addition, bcl-2 and vimentin were reported as being diffusely expressed in spindle cells of synovial sarcoma. In our case, immunostaining was positive for EMA, cytokeratin AE1/AE3, bcl-2 and vimentin, negative for S-100 protein, and there was a lack of ultrastructural features of Schwann's cells, which led us to exclude MPNST. The negative results for SMA, CD117, hepatocyte and calretinin led us to exclude leiomyosarcoma, GIST, hepatic cell carcinoma and mesothelioma. This case was compatible with biphasic synovial sarcoma according to light-field microscopic and immunohistochemical findings.

To the best of our knowledge, this is the first reported case of synovial sarcoma in the omentum. Synovial sarcomas are aggressive tumors. Up to 50% of synovial sarcomas recur locally within 2 years. Metastases occur mainly to the lungs, and less commonly to the lymph nodes and bones. According to previous reports,[19,20] all patients with primary retroperitoneal synovial sarcoma died (at intervals of 7–24 months) with local recurrence or extension, but none metastasized outside the abdomen. It has been shown that the translocation t(X;18)(p11.2;q11.2) is a characteristic chromosome aberration in more than 90% of synovial sarcoma.[21] Monophasic tumors have either SSX1 (about 60%) or SSX2 in their rearrangement, but the majority of biphasic tumors (defined as having distinct gland formation with lumna) have the SSX1 rearrangement.[22] In this patient, the formalin-fixed paraffin-embedded tissue was submitted for molecular analysis using reverse transcription–polymerase chain reaction to determine the specific genetic aberration of this synovial sarcoma. Unfortunately, the molecular evidence was not confirmed due to RNA decay during the experimental procedures.

Carcinogenesis seems to be a multistage process, wherein the primary causal event of synovial sarcoma is presumably the chromosomal translocation. The monophasic and undifferentiated synovial sarcoma might be difficult to distinguish from other tumors, such as fibrosarcomas, malignant schwannomas, malignant fibrous histiocytomas and, in rare instances, carcinomas such as adenocarcinomas.[23] In this case, local recurrence in the retroperitoneal region and bladder with massive ascites developed 2 months after complete resection of the primary tumor. Descending aggression was noted and supposed to be the main reason causing death in this patient. In conclusion, synovial sarcoma is a high-grade malignancy with highly metastatic potential; correct and early diagnosis of synovial sarcoma may impact treatment.

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