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Original Article

Primary poorly differentiated monophasic synovial sarcoma of ileum mesenteries with pulmonary metastasis: a case report

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Abstract: Primary intra-abdominal synovial sarcomas are rare soft tissue malignancies. Herein, we present a case of poorly differentiated monophasic synovial sarcomain ileum mesenterieswith pulmonary metastasis. The patient was a 47-year-old female with a history of cough with variable expectoration and paroxysmal abdominal pain for two months. The tumor was located in ileum mesenteries and composed of monophasic spindle tumor cells with active mitosis and massive necrosis. Tumor cells were positive for vimentin and BCL-2 by immunohistochemistry staining and positive for SYT gene break apart by dual color break apart fluorescence in situ hybridization assay. The differential diagnosis includes gastrointestinal stromal tumour and other mesenchymal tumour.

Keywords: Synovial sarcoma, poorly differentiated, ileum mesenteries, pulmonary metastasis

Introduction

Synovial sarcomais a rare neoplasm that accounts for 5-10% of all soft-tissue tumors and occurs mainly near tendons/tendon sheaths and next to joint capsules. It rarely arises in head and neck, orbit, brain, esophagus, kidney, and prostate [1-6]. Primary intra-abdominal synovial sarcoma is very rare and mainly occurs in omentum [7-18]. The tumor has the specific chromosomal translocation t(X;18) (p11; q11) that leads to formation of a SS18-SSX fusion gene detected in more than 95% of cases [19]. Herein, we present a case of poorly differentiated monophasic synovial sarcoma in ileum mesenteries which was an unusual site and analyzed its clinicopathological features and the differential diagnosis.

Case presentation

A 47-year-old female presented cough with variable expectoration and paroxysmal abdominal painfor two months. She had no diarrhea, nausea, vomiting, and serious medical or surgical history. All laboratory examination including blood tests, biochemical tests, and urinalysis were within normal limits.

Abdominal computed tomography (CT) and magnetic resonance imaging (MRI) showed a giant well-defined ovalmass in the right abdomen. The masswas a cystic-solid and 16.5 cm×8.6 cm in maximum plane size. A small amount of pelvic fluid was visible (**Figure 1**). Chest CT revealed several round masses in the left upper and lower lungs. The largest mass was 5.5 cm×3.2 cm and well-circumscribed. F-18 fluorodeoxyglucose positron emission tomography-CT (18F FDG PET/CT) revealed an increased FDG uptake in the abdominal mass and the lung mass, with a maximum standardized uptake value of 13.0 (**Figure 1**).

The patient received operation for abdomen mass resection. The mass located in the ileum mesenteries and had an apparently well circumscribed with 19 cm×16 cm×9 cm in size. Grossly, the mass was well circumscribed and had a yellow-grayish color with necrotic and hemorrhagic areas on cut surface.

Microscopically, the tumor was composed of monomorphic spindle cells growing in tight fascicles. The spindle tumor cellswere uniform and had scant eosinophilic cytoplasm and round or ovoid nuclei with a finely dispersed chromatin

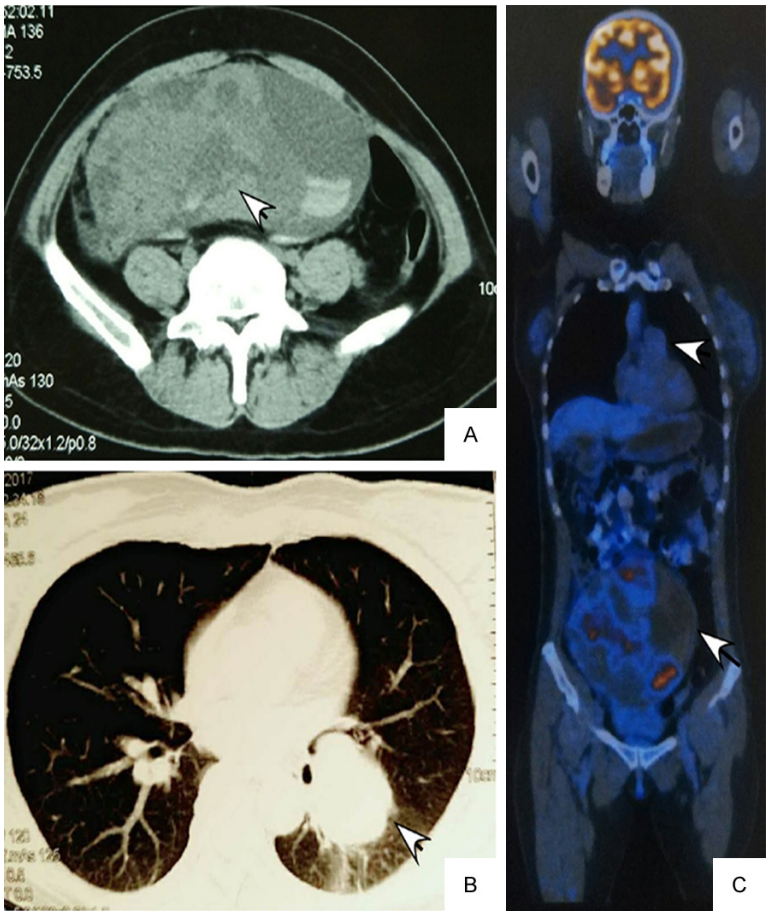

Figure 1. A. Abdominal computed tomography (CT) showed a giant well-defined oval mass (arrow) in the right abdomen. B. Chest CT revealed a round mass (arrow) in hilus of the left lung. C. PET-CT revealed increased FDG uptake in the abdominal mass (arrow) and the mass in hilus of the lung (arrow).

and inconspicuous nucleoli. The nuclear to cytoplasmic ratio was high. Mitoses were frequently found (>10 per 10 HPF). The tumor had a hemangiopericytoma-like pattern. There was massive necrosis and hemorrhage in the tumor. The tumor cells tended to concentrate around blood vessels. No epithelioid components were present. Immunohistochemically, the tumor cells were positive for vimentin and BCL-2, but negative for cytokeratin (AE1/AE3), CK5/6, EMA, desmin, SMA, S-100, WT-1, MC, CR, CD117, DOG1, ER, PR, p16, STAT6, myogenin, MyoD1, D2-40, and CD34 (which outlined only the blood vessel), and no INI-1 expression loss was found. Approximately 60% of tumor cells were positive for proliferation marker Ki-67. SYT gene break apart was identified in the tumor by fluorescence *in situ* hybridization (FISH) assay using SYT (18q11,2) Dual Color Break Apart Rearrangement Probe (**Figure 2**).

Based on clinical findings, histological features, immunophenotype, and FISH detection, we made a diagnosis of poorly differentiated monophasic synovial sarcoma in ileum mesenterieswith pulmonary metastasis.

Discussion

Synovial sarcoma may occur in any part of the body at any age. About 70% of synovial sarcomaarises in the deep soft tissue of the lower and upper extremities, especially in the lower limbs around the knee joint. Primary intra-abdominal synovial sarcoma is rare and mainly involves omentum [7, 9, 17]. We first report primary synovial sarcoma in ileum mesenteries, which is an unusual location.

Microscopically, synovial sarcoma has two main variants: monophasic and biphasic. Our case was composed of spindle tumor cells arranged in fascicleswith hemangiopericytoma-like pattern. Mitosis and massive necrosis were easily identified. Tumor cells were immunoreactive for vimentin and BCL2. SYT gene break apart was found in our case by FISH. The pathological diagnosis of poorly differentiated monophasic synovial sarcoma in ileum mesenteries was made. The differential diagnosis includes gastrointestinal stromal tumour (GIST), malignant solitary fibrous tumour (SFT), leiomyosarcoma, spindle cell rhabdomyosarcoma, and malignant peripheral nerve sheath tumour (MPNST). The tumor located in ileum mesenteries and was composed of spindle tumor cells. So GIST should be differentiated. However, GIST usually involves gastrointestinal wall although it rarely occurs in extra of gastrointestinal tract such as mesenteries. Moreover, tumor cells of GIST are usually immunoreactive for CD117, DOG1, and CD34. Our case was negative for these markers. Malignant SFT is composed of hypercelluar atypical spindle cells with active mitosis and/or focal necro-

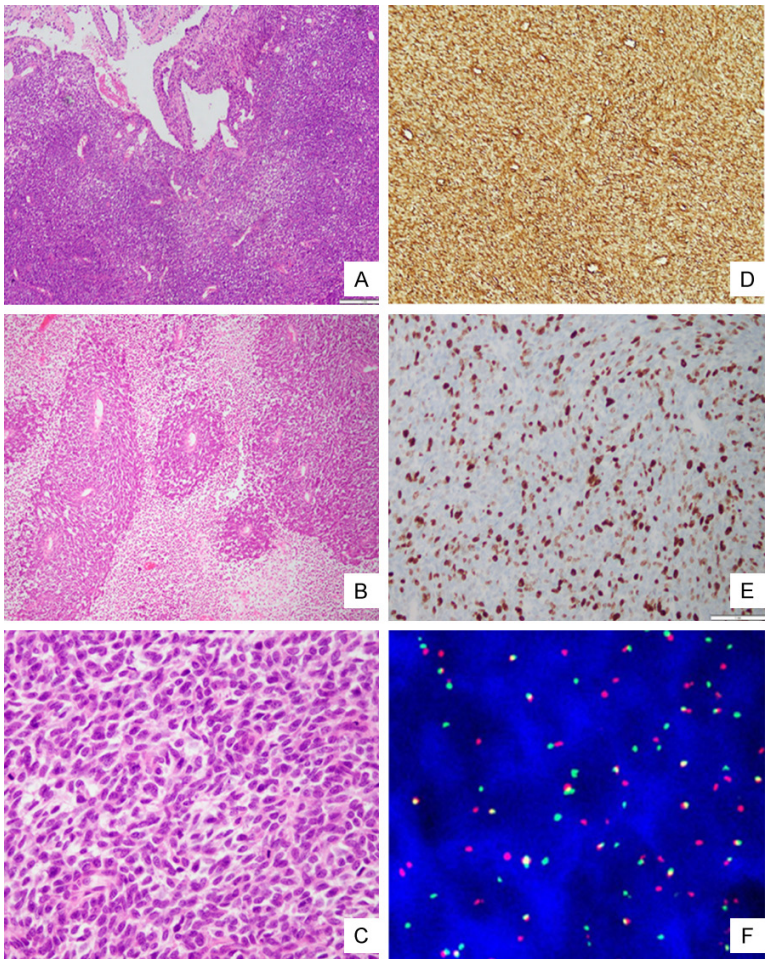

Figure 2. Microscopic features of monophasic synovial sarcoma. A. Tumor had hemangiopericytoma-like pattern. HE×100. B. The tumor cells tended to concentrate around blood vessels with massive necrosis. HE×200. C. Spindle tumor cells arranged in tight fascicles with active mitosis at high power magnification. HE×400. D, E. Tumor cells were immunoreactive for vimentin (D×200) and Ki-67 (E×400) in about 60% of tumor cells. Immunohistochemistry staining. F. SYT gene break apart was identified in tumor cells by dual color break-apart fluorescence in situ hybridization (FISH) assay. FISH×400.

sis and also has hemangiopericytoma-like pattern and is immunoreactive for BCL-2 and CD99. These features of malignant SFT are easily confused with poorly differentiated monophasic synovial sarcoma. However, malignant SFT is commonly immunoreactive for STAT6 and CD34 except for BCL-2 and CD99. Our case was negative for STAT6 and CD34 by immunohistochemistry staining. Leiomyosarcoma and spindle cell rhabdomyosarcoma are composed of spindle tumor cells. However, leiomyosarcoma is immunoreactive for SMA and desmin. Spindle cell rhabdomyosarcoma is immunoreactive for myogenin and MyoD1. The tumor cells tended to concentrate around blood vessels in our case, which is the feature of MPNST. However, MPNST usually arises from a peripheral nerve or from a pre-existing benign nerve tumour (usually neurofibroma) and is immunoreactive for S-100 protein and GFAP. Tumor cellsin our case were negative for S-100 protein. Most importantly, specific SYT gene break apart was found in our reported tumorby FISH. As our knowledge, GIST and the above mesenchymal tumour are lack of such specific chromosomal translocation.

Surgical resection and adjuvant chemotherapy are the preferred treatments for synovial sarcoma. But the prognosis of synovial sarcoma is usually poor. Histology and grading are important prognostic factors in synovial sarcoma. Sarcoma specific survival (OS) was 56.6% at 5 years and 46.9% at 10 years. Prognosis was worse in those patients monophasic synovial sarcoma as in those with grade 3 tumors [20]. Metastasis occurs most commonly in lungs and bone. The patient in our case had multiple pulmonary metastases by chest CT examination as primary synovial sarcoma in ileum mesenteries was confirmed. As for new therapeutic strategy, Yamda et al. have reported TAS-115 monotherapy may benefit synovial sarcoma patients whose tumour are dependent upon either c-MET or PDGFRα signaling by functioning as a multiple tyrosine kinase inhibitor to suppress c-MET as well as PDGFRα pathways [21].

Disclosure of conflict of interest

None.

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