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Primary peritoneal spindle cell sarcoma presented with massive ascites: a case report

A. Bilici[1], K. Uygun[2], E. Altun[2], E. Karci[2], Y. Gurbuz[3], A. Semih Dogan[4]

[1]Department of Medical Oncology, Dr. Lutfi Kirdar Kartal Education and Research Hospital, Istanbul; [2]Department of Medical Oncology, [3]Department of Pathology, Kocaeli University, Medical Faculty, Izmit, Kocaeli; [4]Department of Nuclear Medicine, Anadolu Health Center, Gebze, Kocaeli, Turkey

Summary

Soft tissue sarcomas (STSs) are a heterogeneous group of rare neoplasms originating from the embryonic mesoderm and mesenchymal cells. Primary peritoneal sarcomas have been reported as leiomyosarcoma, liposarcoma and carcinosarcoma. However, primary peritoneal spindle cell sarcoma has never been reported in the literature. Herein, we report on a 38-year-old woman with massive ascites diagnosed as primary peritoneal spindle cell sarcoma. Following doxorubicin and ifosfamide chemotherapy, her symptoms improved and ascites regressed. Other primary peritoneal sarcomas rarely cause massive ascites. We suggest that in patients presenting with massive exudative ascites associated with malignancy, primary peritoneal sarcomas should be also considered in the differential diagnosis.

Key words: massive ascites, peritoneum, spindle cell sarcoma

Introduction

STSs are rare malignant tumors and constitute about 1% of all cancers in adults. They are thought to develop from mesenchymal cells residing in muscle, fat, and connective tissues [1-4]. Although malignant fibrous histiocytoma is the most common STS, liposarcoma is the most common STS encountered in the abdomen and retroperitoneum. Primary peritoneal leiomyosarcoma, malignant mixed müllerian tumor (MMMT) and primary mesenteric liposarcoma have been previously documented [5-7], whereas primary peritoneal spindle cell sarcoma has never been reported. Herein, we describe an unusual case of primary peritoneal spindle cell sarcoma with massive ascites.

Case presentation

A 38-year-old woman was admitted to the department of medical oncology, Kocaeli University hospital, in August 2007, with one-month history of abdominal swelling, early satiety, fatigue, anorexia and nausea. Her past medical history was not significant and she was not taking any medications. Her family history was non-contributory. Her physical examination was normal except massive ascites that was noted on percussion. The results of her initial laboratory examinations showed ALT 30 U/L (normal 5-37), AST 61 U/L (normal 5-37), lactate dehydrogenase 466 IU/L (normal 125-243), γ-glutamyltranspeptidase 74 U/L (normal 7-49), total protein 6.1 g/dL (normal 6.2-7.5), albumin 2.8 g/dL (normal 3.5-5.0), erythrocyte sedimentation rate (ESR) 45 mm/h, CA 125 142.3 U/ml (normal 0-35). Serology revealed HBsAg (+), anti-HBe(+), HBeAg (–), HBV DNA PCR (+), anti-HBs (–), anti-HBc IgG (–), anti-HCV, anti-HAV IgM (–) / IgG (–), anti-HIV, ANA, anti-smooth muscle antibody (ASMA) negative. Other laboratory values were within normal limits.

A diagnostic work up for all causes of ascites was initiated. Serology for viral hepatitis showed chronic hepatitis B infection (CHBI). Markers for autoimmune hepatitis were negative. The patient was firstly treated with lamivudine 100 mg/day for diagnosed of CHBI. Abdominal ultrasonography (US) revealed massive ascites, while no portal hypertension was detected on

Correspondence to: Ahmet Bilici, MD. Menderes Mah, 23.sok, Caglar Apt.No:16, D:1, Istanbul, Turkey. Tel: +90 532 528 04 86, Fax: +90 216 447 26 78, E-mail: ahmetknower@yahoo.com

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Doppler US. Ascitic fluid was exudate. Bacteria and acid-fast bacilli were not detected, and PCR for tuberculosis was negative. Fluid culture also turned out to be negative. Two repeat cytological examinations of the ascitic fluid showed findings suspicious for malignancy and proliferation of dense mesenchymal cells. CT scan of the abdomen was normal except ascites. Colonoscopy and gastroduodenoscopy were also normal. Then, positron emission tomography (PET) scan was performed which revealed omental implants and a mass, suggestive of malignancy (Figure 1). After exclusion of the most common possible causes of ascites, the condition was attributed to malignancy and exploratory laparotomy was carried out. In exploration, multiple implants measuring 0.5 cm in diameter were detected on peritoneal surfaces and multiple peritoneal biopsies were performed. Histopathological examination of peritoneal biopsy specimens revealed Ki-67 (+) and CD 117 (-) spindle cell sarcoma cells (Figures 2,3).

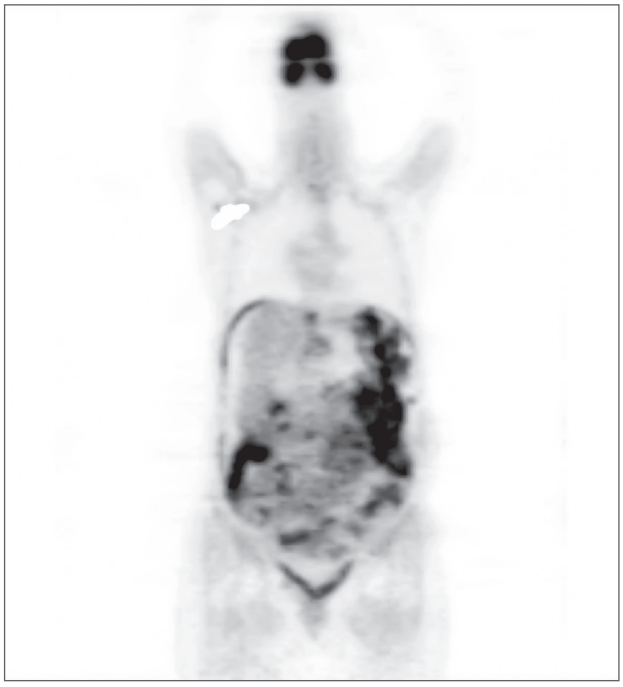

Figure 1. PET scan showing abnormally increased FDG localization in the omentum, which appeared as a soft tissue density mass, and peritoneal implants under the diaphragm and the abdominal wall.

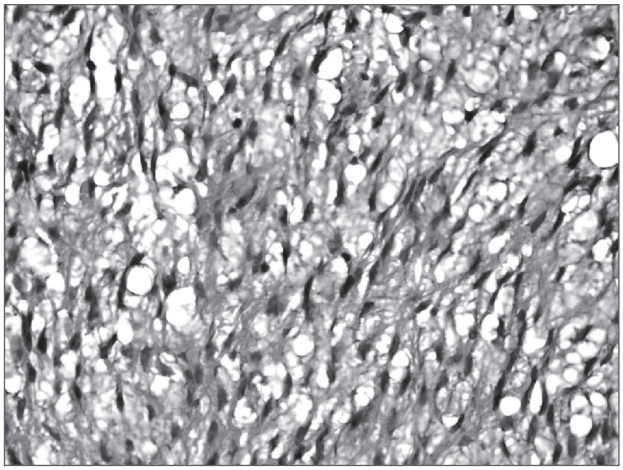

Figure 2. Malignant spindle-shaped tumor cells with long oval hyperchromatic nuclei (H&E ×400).

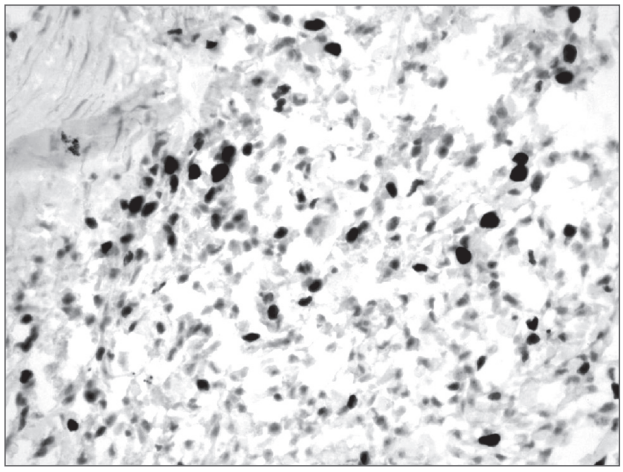

Figure 3. High nuclear Ki-67 immunoreactivity in tumor cells (immunohistochemistry Ki-67 ×400).

In the light of radiological and pathological findings, the patient was treated with combination of doxorubicin (75 mg/m^2, day 1) and ifosfamide (2 g/m^2, days 1-3) every 3 weeks. She tolerated treatment well and ascites decreased significantly after the first cycle of chemotherapy. After 6 chemotherapy courses, ascites and PET findings completely regressed. During an 8-month follow-up, she had no symptoms and both clinical and laboratory examinations were normal.

Discussion

Primary peritoneal malignancies are rare and frequently known as peritoneal carcinomatosis. They disseminate and implant tumor cells throughout the peritoneal cavity. The most common peritoneal tumor histology is carcinoma [8,9]. On the other hand, primary peritoneal malignant mesothelioma originates from the mesothelial cells, showing 3 histologic subtypes: epithelial, sarcomatoid and mixed [10-12].

Primary STSs located in the peritoneal surface are very rarely encountered in clinical oncology practice. They are unusual tumors arising from mesenchymal cells as other STSs. Primary peritoneal leiomyosarcoma [6], MMMT [7], and primary mesenteric liposarcoma [5] have been previously documented in the lit-

erature, although primary peritoneal spindle cell STS has not been reported.

Peritoneal malignancies are frequently presented with nonspecific symptoms, including abdominal distention and early satiety secondary to massive ascites, and rarely as abdominal mass [8,9]. Our patient also complained of abdominal swelling and early satiety.

This case constitutes an unusual presentation of primary peritoneal spindle cell sarcoma associated with massive ascites. In patients with malignancy-related ascites who have peritoneal implants and mass, primary peritoneal sarcomas should be considered in the differential diagnosis.

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