A Case of Carcinosarcoma of the Abdominal Wall

Hajime Taniguchi¹, Mitsuhiro Nokubi² and Fumio Konishi¹
¹Department of Surgery, Saitama Medical Center, Jichi Medical University
²Department of Pathology, Saitama Medical Center, Jichi Medical University

Abstract
This report details a rare case of carcinosarcoma of the abdominal wall. A 62-year-old female patient with no prior history of abdominal surgery was referred to our hospital. She experienced general malaise and had a palpable mass of 8 cm at the lower right quadrant of her abdominal cavity since a month prior to admission. Imaging studies revealed invasion of a large tumor into the abdominal wall. The patient was operated to resect the tumor and to determine future treatment possibilities. Operative findings suggested development of a primary lesion from the abdominal wall, and histological findings revealed a carcinosarcoma. The origin of this carcinosarcoma was unclear, and hence, immunohistochemical studies were performed. Based on surgical findings and CD10 positivity, the tumor was diagnosed as primary peritoneal malignant müllerian mixed tumor (MMMT). The patient underwent adjuvant chemotherapy, but succumbed to the disease on postoperative day 34. Previous literatures and the present case suggest that primary peritoneal MMMT is rare neoplasm, and it is highly malignant and aggressive with a poor patient prognosis.

Key words: carcinosarcoma, primary peritoneal MMMT

Introduction
Carcinosarcoma is a rare malignant tumor, comprising of both carcinoma and sarcoma components. It has been detected in many organs such as the esophagus, breast, lung, liver, fallopian tube, uterus, and ovary. However, carcinosarcomas of the abdominal wall are very rare. This case had a primary peritoneal carcinosarcoma with no history of former abdominal surgery or malignancies.

Case Report
A 62-year-old female patient with no former history of surgery or malignancies, was referred to our department. The patient’s chief complaints included general malaise and an abdominal mass at the right lower quadrant since one month prior to admission. A relatively hard, smooth, fixed tumor with slight tenderness was detected. Laboratory data were within normal limits. Enhanced CT revealed a 7-cm, heterogeneous, round tumor at the right lower abdominal quadrant. Invasion into the abdominal wall was suspected, but that into the intestinal tract was not observed (Fig. 1). Feeding vessels were observed from both the superior mesenteric and right external iliac arteries. Colonoscopy revealed an extrinsic compression at the cecum, ileocecal valve, and sigmoid colon. However, no abnormalities were observed in the mucosa (Fig. 2). PET-CT was also performed. SUV was elevated to 7.8 in the tumor, and was 5.9 in the left obturator lymph node (Fig. 3). Taking these results into account, we suspected that this tumor originated from the mesentery. Extra intestinal GISTs, malignant lymphomas, abdominal desmoids, and desmoplastic small round cell tumors were considered as differential diagnosis. Biopsy was not performed to avoid possibilities of tumor seeding. An operation was scheduled to
Primary peritoneal malignant müllerian mixed tumor

determine future treatment possibilities.

The patient was operated under general anesthesia. Ascites and peritoneal dissemination were present. Considering the size, rapid tumor growth, and overall diagnoses, we decided to resect the tumor. The tumor was primarily localized in the abdominal wall, and involved the right iliac vessels and right abdominal wall. It was separated entirely from the kidney, uterus, and ovary, and did not invade the colon or ureter. We resected the tumor with the abdominal wall; however, a residual tumor remained along the right external iliac vein and bladder. Operative findings suggested development of the primary lesion from the abdominal wall.

Macroscopic pathological findings, revealed expanding tumor growth beneath the peritoneum. Half the tumor was connected to the abdominal wall, while the other half extended towards the abdominal cavity (Fig. 4). The resected specimen revealed carcinoma component only at tumor edges, and was comprised of hob nail as well as clear cells. Carcinoma cells were also detected in ascites. The sarcoma component comprised of collagen fibers (Fig. 5). Immunohistochemical studies revealed positive NSE in both components, keratin only in the carcinoma component, and vimentin as well as CD10 were expressed in the sarcoma component (Table 1). Alveolar growth of clear cells and hob nail cells with papillary growth were observed from the edge facing the peritoneum. Considering both tumor components, the patient was diagnosed with a carcinosar-
Fig. 5 Carcinoma comprising of clear cells (indicated by a black arrow) and hob-nail cells (indicated by a white arrow). (H&E stain, 400×)

Discussion
Carcinosarcoma comprises of both carcinoma and sarcoma elements. Three theories have been proposed to explain the pathogenesis of carcinosarcoma: the collision tumor theory, combination tumor theory (true carcinosarcoma), and composition tumor theory (carcinosarcoma). Recent gene-based studies have favored the combination tumor theory, while the collision and composition tumor theories have no evidential proof. It was also classified into 2 groups, sarcomatoid carcinoma or carcinosarcoma, from the degree of contribution, but the combination theory is now favored over either. Immunohistochemical staining aids in elucidating the likely origin of carcinosarcoma.

In this case, the tumor was composed of both carcinomatous and sarcomatous components. Hence, the patient was diagnosed with carcinosarcoma. An extensive search of medical literature revealed instances of this tumor in many different organs, such as the liver, gallbladder, peritoneum, rectosigmoid colon, kidney, fallopian tube, ovary, uterus, urinary tract, prostate, and urinary bladder. Carcinosarcomas arising from the genitourinary tract are relatively common, but those arising from the peritoneum are very rare. To biochemically evaluate the carcinosarcoma, immunohistochemical staining was
Primary peritoneal malignant müllerian mixed tumor

performed. The tumor stained positive for CD10, and was therefore diagnosed as MMTT. CD10 is positive in endometrial stromal sarcomas and MMTT\(^{20}\). MMTTs are highly aggressive biphasic neoplasms composed of carcinomatous and sarcomatous components\(^{20}\). Most tumors in the female genital tract arise from the endometrium\(^{20}\), but cases of tumors in the ovaries, falloplian tubes, cervix, and vagina are also encountered although with lesser frequency\(^{20-29}\). Extragenital primary peritoneal MMTTs are rare, and few cases have been reported in literature\(^{1,22}\). Extragenital MMTTs are most likely independent primary neoplasms. The female peritoneum is known as the secondary müllerian system because it arises from the same embryologic origin. Hence, any müllerian neoplasm could arise from the secondary müllerian system, and present as a primary peritoneal tumor\(^{22,31}\). Our patient was suspected to have a primary peritoneal MMTT, based on imaging studies and the surgical findings. Although extragenital MMTTs are often associated with endometriosis, no evidence of endometriosis was found in this case. Up to 37.5% of peritoneal MMTTs are associated with gynecological tumors, but not in this instance\(^{3}\).

22 cases with primary MMTT of the peritoneum have been reported\(^{20}\). Among them, 18 patients were well documented and were able to amend analysis. All were elderly postmenopausal women. The mean age was 66.3 years (range 40-84 year-old). The most effective treatment was surgical excision. Adjuvant radiotherapy and various combinations of chemotherapy yielded inconsistent results but, most of the patients died within 1 year regardless of the tumor stage.

In summary, we examined a rare case of primary peritoneal MMTT, which is a highly malignant, aggressive neoplasm behavior with a poor patient prognosis.

References


