Case Report

Primary malignant mixed mullerian tumor of the peritoneum: a case report

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Abstract: Malignant mixed mullerian tumor (MMMT) occurs rarely extragenital sites and generally originates from female genital tracts such as the uterus. Here, we report a patient with a primary MMMT of the peritoneum in a 64-year-old woman, who experienced a discontinuous epigastric pain and an abdominal distention for 3 months. An ultrasound-guided fine needle aspiration biopsy (FNA) was executed and indicated a highly malignant soft tissue sarcoma. And after that, the patient received a tumor resection (cytoreductive surgery) and postoperative chemotherapy. The postoperative immunohistochemical analysis revealed a MMMT in peritoneum. Up to now, the patient has survived for 5 months.

Keywords: Malignant mixed mullerian tumor (MMMT); primary peritoneal carcinosarcoma

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Introduction

MMMT, also referred to as the malignant mixed mesodermal tumor and carcinosarcoma (1,2), is a highly aggressive biphasic neoplasm comprised coexisting elements of cancer and sarcoma. Mostly deriving from the genital tract in postmenopausal elderly women, such as the uterus or the ovary, it also can be found in neck and head, peritoneum, biliary tract and gastrointestinal tract (3). Primary peritoneal MMMT is extremely rare, and commonly occurs in the pelvic peritoneum. Furthermore, it has also been arisen in the serosal surface of the colon, rectal peritoneum, retroperitoneum, cul-de-sac, diaphragm peritoneum, anterolateral abdominal peritoneum, and omentum (4,5). Since the first reported by Ober and Black in 1955, only 41 cases have been described in the well documented literatures (6). In this paper, we present a peculiar case of primary peritoneal MMMT.

Case presentation

A 64-year-old woman was admitted to our department (First Affiliated Hospital of Zhejiang University, China) in April 2018 due to a discontinuous epigastric pain and an abdominal distention for 3 months. On physical examination, a palpable mass was touched in the left and upper quadrant. Initial complete blood count, liver, and kidney function tests were normal. Her serum CA 125 was 486.7 U/mL. The abdominal computed tomography (CT) scan demonstrated a 12×19 cm tumor with equal or low density, with multiple retroperitoneal enlarged lymph nodes, and the tumor's margin in the artery phase obviously enhancing and delayed enhancing in the portal phase, uterus and adnexal region revealing no abnormality. The abdominal magnetic resonance imaging (MRI) revealed a giant tumor in the posterior peritoneum with iso-hypointense on T1WI, hyperintense on T2WI, uneven hyperintense or hypointense on diffusion weighted imaging (DWI), and progressive uneven enhancement in enhancement period (*Figure 1*). And then, the patient received an ultrasound-guided FNA of abdominal tumor in the left middle abdomen, and the initial pathological analysis indicated a highly malignant soft tissue sarcoma. According to pathological results and iconography's characteristics, the patient was diagnosed as retroperitoneal sarcoma.

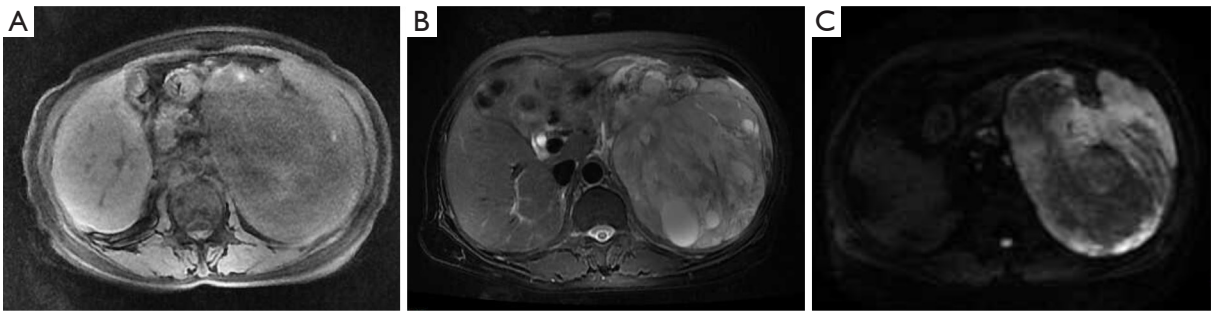

Figure 1 The splenic magnetic resonance imaging (MRI) revealed a giant focus of the posterior peritoneum showing iso-hypointense on T1WI (A), hyperintense on T2WI (B), uneven hyperintense or hypointense on DWI and progressive uneven enhancement in enhancement period (C). DWI, diffusion weighted imaging.

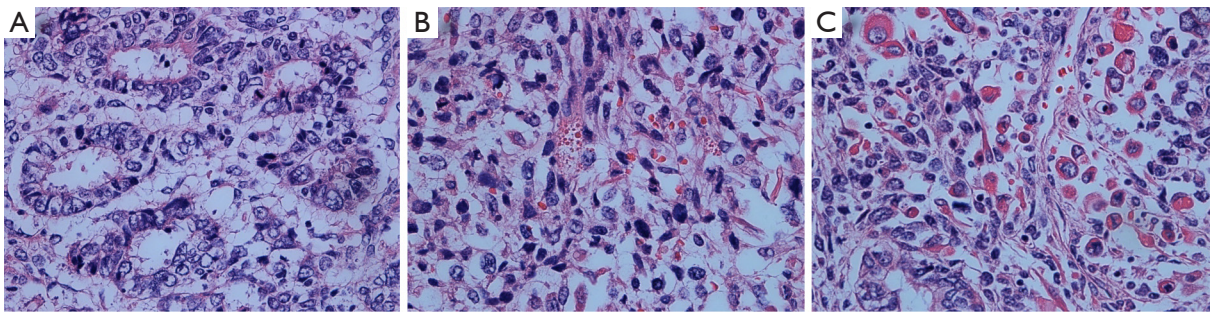

Figure 2 Histopathology of tumor showed biphasic differentiation, and epithelioid components with tumor cells of marked heteromorphism arranged in streak glands with structures (A) (magnification, ×400). Stromal components were diffuse arrangement, infiltrative growth and spindle shaped with pleomorphic nuclei (B) (magnification, ×400), rhabdomyosarcoma differentiation in some region (C) (magnification, ×400). Immunohistochemistry staining showed positive expression for CK, CK7, SMA, WT1, PAX-8, Ki-67 (75%) and negative expression for CD117, CD30, CK20, S-100, P53.

Treatment procedures

Retroperitoneal tumor resection with terminal segmental pancreatectomy and splenectomy were planned. On the operation, A 21 cm × 18 cm × 13 cm medium-texture tumor was found in the abdominal cavity, which has an undistinguishable boundary with the spleen. The tumor along with terminal segmental pancreas and spleen was resected. Histopathology of tumor showed biphasic differentiation, and epithelioid components with tumor cells of marked heteromorphism arranged in streak glands with structures. Stromal components were diffuse arrangement, infiltrative growth and spindle shaped with pleomorphic nuclei, rhabdomyosarcoma differentiation in some region, the pathological and immunohistochemical analysis suggestive of malignant mixed mullerian tumor (MMMT) with positive expression of CK7, CK, SMA, WT1, PAX-8, Ki-67 (75%) and negative expression of CD117, CD30, CK20, S-100, P53 (*Figure 2*). Postoperatively, the patient received three cycles chemotherapy of taxinol combined with carboplatin, the patient obtained partial response and the abdominal symptoms were obviously improved. Up to now, the patient has survived for 5 months.

Discussion

The terminology ‘mixed Müllerian tumor’ was made up of mesenchymal and epithelial elements of Müllerian origin. It generally classified into adenomyomas, adenofibromas, adenosarcomas, and carcinosarcomas (MMMT) according to the grade malignancy of the stromal and epithelial components (7). Tumors showing no differentiated sarcomatous elements have conventionally been known as homologous carcinosarcomas (8). Nevertheless, tumors

consisted of carcinomatous elements in sarcomatous stroma with recognizable heterologous components, such as cartilage or muscles, went by the name of heterologous carcinosarcomas (8). The peritoneal carcinosarcoma's pathogenesis has not been clarified at present. However, the most popular theory, known as the combination or conversion theory, is that both elements derived from a common, strange precursor or probably that the sarcomatous component composes of metaplastic transformation (9).

It is possible that carcinomatous and sarcomatous components are originated from an alike source, in view of the both components showing similar mutations (8). Rajanbabu *et al*. reported 36 cases with primary peritoneal MMMT, most of which originated in the pelvic peritoneum and uterine serosa, followed by the serous surface of the Colon, rectum, and other peritoneum (10). Consequently, it most often arises from genitourinary tract whereas could occur in any organ. MMMT, especially in peritoneum, is extremely rare with a poor prognosis and rapidly fatal course, regardless of tumor stage (11). El Hachem *et al*. (12) reported a case of the longest disease-free survival that was treated with a complete radical surgical treatment and adjuvant chemotherapy with 6 cycles of carboplatin and paclitaxel, and this patient was followed up to 113 months.

At present, the treatment of primary peritoneal MMMT is guided by experience of prior patients and treatment recommendations for uterine sarcoma, owing to limited data on its management and treatment. In terms of MMMT treatment, surgical tumor resection is the most effective method, and we should try to reach the status of no evidence of tumor by the surgeon's unaided eye. Systemic adjuvant chemotherapy is essential for all patients after operation. Ifosfamide and cisplatin are usually the preferred chemotherapy regimen, and the reaction rate of ifosfamide is 25% and that of cisplatin as a single drug for uterine carcinosarcoma is 17% (13). The effect of radiotherapy is controversial for MMMT. There is no clear conclusion as to whether to undergo radiotherapy (11).

The patient reported by us underwent tumor resection firstly and received postoperative chemotherapy with 4 cycles of cisplatin combined with ifosfamide. Up to now, the patient has survived for 5 months and has been receiving current chemotherapy. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/tcr.2019.05.07). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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