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Primary peritoneal serous carcinoma, an extremely rare malignancy: A case report and review of the literature

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Abstract. Primary peritoneal serous carcinoma (PPSC) is an extremely rare malignancy that was first described in 1959. This type of cancer arises from the peritoneal epithelium and is similar to serous ovarian carcinoma. A diagnosis of PPSC is typically made based on the Gynecology Oncology Group criteria; however, a correct differential diagnosis of PPSC is difficult preoperatively. The current study describes the case of a 66 year-old female patient presenting with abdomen distension. A computed tomography (CT) scan revealed abundant ascites in the abdominal cavity and omental infiltration. The results of positron emission tomography/CT showed hot uptake in the greater omentum. Furthermore, preoperative serum cancer antigen-125 levels were 1,032 U/ml. Upon surgical exploration, a whitish mass and nodule were found in the greater omentum. Therefore, omentectomy was performed. Pathological examination of the resected specimen revealed a diagnosis of PPSC. PPSC is extremely rare with few cases cited in the current literature. The present study describes a rare case of PPSC with a review of the literature.

Introduction

Primary peritoneal serous carcinoma (PPSC) is an extremely rare malignancy that was first described in 1959 (1). The estimated incidence of PPSC in the United States is 6.78 cases per 1,000,000 individuals (2). This type of cancer arises from the peritoneal epithelium and is similar to serous ovarian carcinoma. A diagnosis of PPSC is typically made based on the Gynecology Oncology Group criteria (3). However, a correct differential diagnosis of PPSC is difficult preoperatively. The median overall survival time of PPSC is 11-17 months (4) and the optimal treatments for PPSC are surgical resection and platinum based chemotherapy (5).

The current study presents the case of a 66 year-old female patient diagnosed with PPSC mimicking omental metastasis and peritoneal carcinomatosis from the gastrointestinal tract or ovarian malignancy. PPSC is extremely rare with few cases cited in the current literature. The present study describes a rare case of PPSC with a review of the literature. Written informed consent was obtained from the patient.

Case report

In September 2014, a 66-year-old woman presented to Yeungnam University College of Medicine (Daegu, Korea) with abdomen distension for 3 weeks. Otherwise, the patient was in good health with intermittent abdominal discomfort. Findings upon physical examination were unremarkable. Furthermore, the initial laboratory tests were all within normal ranges. Chest and abdominal radiographs were also unremarkable. The patient underwent a contrast-enhanced abdominal computed tomography (CT) scan (SOMATOM Definition AS; Siemens AG, Munich, Germany) to evaluate the cause of the abdominal distension. The CT scan revealed abundant ascites in the abdominal cavity and omental infiltration (Fig. 1). Other abdominal organs, including the liver, spleen, kidney and both ovaries, appeared normal upon CT. Ascitic cytology was then performed; evaluation of the cytological characteristics of the ascites suggested the presence of malignant cells.

18F-fluorodeoxyglucose positron emission tomography (PET)/CT (Discovery DVCT; GE Healthcare Bio-Sciences, Pittsburgh, PA, USA) was then performed to evaluate the primary origin of the malignancy. The result of PET/CT revealed hot uptake in the greater omentum (Fig. 2). Other abdominal organs, including the stomach, colon, pancreas, uterus and both ovaries, were within the normal range of uptake. Furthermore, the patient's preoperative serum cancer antigen-125 (CA-125) levels were elevated to 1,032 U/ml (normal range, 0-35 U/ml).

Based on the aforementioned results, a diagnosis of primary peritoneal malignancy was determined. Surgery was identified as the most appropriate treatment strategy, therefore, surgical exploration was performed in December 2014. Upon surgical exploration, ~1,000 cm^3 ascites, a whitish mass (measuring 16x10x2.4 cm) and a nodule were found in the greater omentum (Fig. 3). The uterus and both adnexa were

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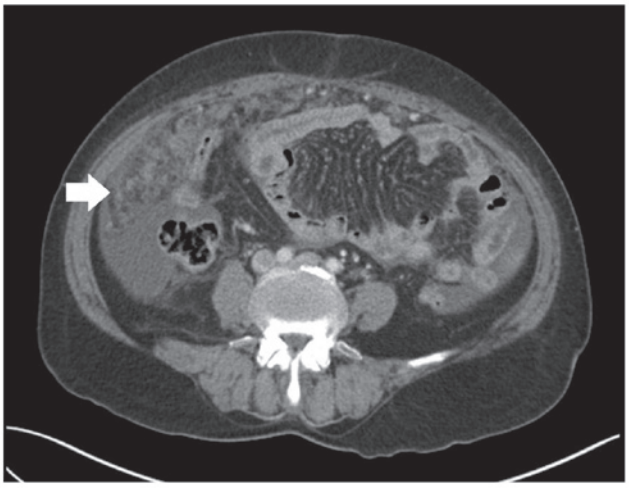

Figure 1. Computed tomography scan revealing abundant ascites and omental infiltration. The white arrow indicates omental infiltration.

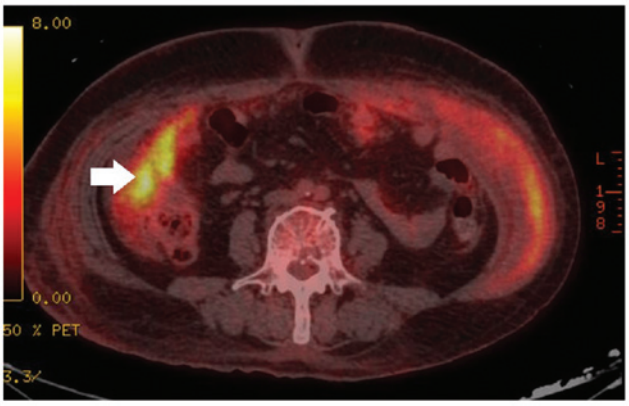

Figure 2. Positron emission tomography/computed tomography findings. The white arrow indicates hot uptake in greater omentum.

unremarkable without tumor involvement. The stomach, liver, small bowel, large bowel and pancreas were also unremarkable. Inspection of the abdominal cavity revealed a rice-sized nodule in the mesentery, peritoneum and pelvic cavity. Thus, omentectomy was performed. Resected tissue was fixed in 10% buffered-formalin, paraffin-embedded and cut into 4 μm sections, and subsequently stained with hematoxylin and eosin. For immunohistochemical analysis, the specimens were incubated with monoclonal mouse anti-human cytokeratin 7 (cat. no. M7018; 1:200; Dako, Glostrup, Denmark), monoclonal mouse anti-human WT-1 (cat. no. 348M-94; 1:100; Cell Marque™; Sigma-Aldrich, St. Louis, MO, USA) and monoclonal mouse anti-human CA-125 (cat. no. M3519; 1:40; Dako) antibodies. The results revealed positivity for cytokeratin-7, WT-1 and CA-125. Pathological examination of the resected specimen further indicated a diagnosis of PPSC. In the current case, both ovaries were of normal size with no observed abnormalities, and the main tumor was identified in the omentum. Therefore, according to the Gynecology Oncology Group criteria (3), surgical and pathological findings, a diagnosis of PPSC was determined. The patient was treated with regular chemotherapy [7 cycles of 175 mg/m^2 paclitaxel and carboplatin (AUC, 4-7.5) every 3 weeks], which was initiated in

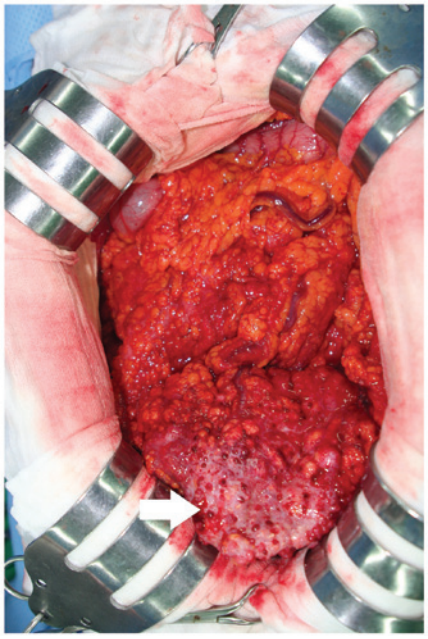

Figure 3. Intraoperative finding. The white arrow indicates whitish mass in the greater omentum.

December 2014, and was followed up every month. The patient had an uneventful postoperative course, and is currently alive and well.

Discussion

Peritoneal malignancies may be classified as primary or secondary, depending on the site of origin of the cancer. Primary peritoneal malignancies, which include malignant mesothelioma, serous carcinoma and sarcomas, are rare (6). Among these primary peritoneal malignancies, PPSC, is an extremely rare subtype that was first described in 1959 (1). In the United States, the incidence of PPSC is 6.78 cases per 1,000,000 individuals (2). However, the worldwide incidence rate is unknown and publications concerning PPSC are only case reports or case series.

The common presenting symptoms of PPSC are abdominal distension, abdominal pain and discomfort. Therefore, symptoms of PPSC are similar to those of peritoneal carcinomatosis. Furthermore, the majority patients described in case reports to date were diagnosed in the advanced stages of the disease (7,8). Traditionally, a diagnosis of PPSC is based on the Gynecology Oncology Group criteria, as follows: i) The ovaries are either absent or normal in size; ii) the involvement of the extraovarian sites is greater than the involvement of the surface of either ovary; iii) the absence of a deep-seated invasive ovarian carcinoma or invasive disease in the ovarian cortical stroma with tumors that measuring <5x5 mm^2; and iv) histopathological and cytological characteristics of the tumors similar to those for epithelial ovarian cancer (3). Preoperatively, a correct diagnosis of PPSC is difficult. The current case was preoperatively diagnosed with omental metastasis and peritoneal carcinomatosis from the gastrointestinal tract or ovarian malignancy. However, PET/CT is beneficial due to its ability to define the extent of metabolically active disease

and its ability to detect distant metastasis. Therefore, PET/CT is beneficial for the differentiation of PPSC from ovarian cancer (9).

In postoperative immunohistochemical examinations, PPSC is typically positive for cytokeratin-7, CA-125, estrogen receptor and Wilms' tumor-1 (WT-1) (5). In the current case, cytokeratin-7, WT-1 and CA-125 were positive.

The optimal treatment of PPSC is surgical resection and chemotherapy, with platinum-based chemotherapy used most commonly (10). Intraperitoneal chemotherapy has recently demonstrated a survival benefit in patients with PPSC when compared to those treated with surgery alone or surgery in combination with systemic chemotherapy (11).

Although it is difficult to generalize, as some published survival data were from small size studies, the median survival time of patients with PPSC is 11-17 months (4), which is similar to the present case with a current survival time of 15 months.

In conclusion, the current study presented a case of PPSC treated with debulking surgery. Although PPSC is considered extremely rare, its recognition is important for accurate evaluation and management. Due to its significant rarity, multicenter studies are required to further understand prognosis and identify an effective treatment approach.

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