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Case Report

Imaging features of primary peritoneal serous carcinoma: A case report

Ji Soo Oh, M.D.ª, Beum Jin Kim, M.D.ª, Myoung Jin Ju, M.D.^b, Eun Ae Yoo, M.D.^{a,*}

^a Department of Radiology, Presbyterian Medical Center, 365, Seowon-ro, Wansan-gu, Jeonju-si, Jeollabuk-do, Republic of Korea

^b Department of Pathology, Presbyterian Medical Center, Jeonju, Republic of Korea

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ABSTRACT

Primary peritoneal serous carcinoma (PPSC) is an epithelial tumor that arises from the peritoneum. On histopathologic analysis, it resembles a malignant ovarian surface epithelial stromal tumor. A 62-year-old woman visited the emergency room with low abdominal pain. Peritoneal carcinomatosis from ovarian cancer was initially suspected and underwent radical hysterectomy, both salpingo-oophorectomy and omentectomy. Both ovaries and uterus were intact, and ultimate diagnosis was PPSC. PPSC is a rare disease that is difficult to diagnose prior to surgery. We suspected peritoneal carcinomatosis due to the rarity of primary tumor originating from the peritoneum and overlooked PPSC, but when the primary site is not clear, we advise to consider the possibility of PPSC. Median survival time is 11-17 months with poor prognosis, and thus early diagnosis, treatment is important, and further imaging studies are warranted.

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Introduction

Primary peritoneal serous carcinoma (PPSC) is an epithelial tumor that arises from the peritoneum. On histopathologic analysis, it resembles a malignant ovarian surface epithelial stromal tumor. PPSC is very rare, and the median survival time is 11-17 months with poor prognosis. Therefore, early diagnosis and treatment is important, and further imaging studies are warranted. We herein report a case of PPSC confirmed by imaging, surgery, and pathologic examination.

Case report

A 62-year-old woman visited the emergency room at 9 am on January 27, 2019. She presented with low abdominal pain that started 1 month ago. She had undergone abdominal and pelvis CT at a local hospital several days ago and was suspected as having peritoneal carcinomatosis and TB peritonitis. There was a history of cholecystectomy in the past.

In laboratory tests, both Quantiferon-TB (TB Ag-Nil: -0.03, Nil: 0.10, and Mitogen-Nil: 52.80) and sexually transmitted disease tests showed negative findings. CA 125 was increased to

* Corresponding author.

E-mail address: silver-824@hanmail.net (E.A. Yoo).

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Fig. 1 - Primary peritoneal serous carcinoma (PPSC) of the omentum in a 62-year-old woman.

Precontrast computed tomography (CT) image reveals dense soft tissue mass measuring 7.7 \times 2.5 cm and lobulated contour in the pelvic cavity omentum (A). Soft tissue mass was well enhanced, and there was also mild peritoneal infiltration (B). On the coronal image, a mass is shown that infiltrates the omentum and shows homogenous enhancement (C).



Fig. 2 - No soft tissue mass was observed in the pelvic cavity omentum when compared with CT images taken in April 2016.

59.4 U/mL (normal, 0.0-35.0 U/mL), and she was classified as belong to the ovarian cancer high-risk group.

Contrast-enhanced abdominopelvic computed tomography was taken and revealed a dense soft tissue mass measuring 7.7×2.5 cm with lobulated contour in the pelvic cavity omentum (Fig. 1A), and the mass was well enhancing (Fig. 1B). In addition, mild peritoneal infiltration (Fig. 1B, C) and minimal amounts of ascites were noted. All of these lesions were found to be new when compared to CT images from April 2016 (Fig. 2A-C).

18F-fluorodeoxyglucose positron emission tomography (PET/CT) showed a nodular mass lesion with FDG hot uptake (Fig. 3A-C) and a nodular lesion with oval-shaped FDG uptake presumed to be the right ovarian or seeding nodule (Fig. 3D-F). However, these lesions were not clearly distinguished due to overlapping bowels. Focal FDG uptake was also observed at the site of the uterine cervix or lower body (Fig. 3G-I), suggesting malignancy, and there was no evidence of any other significant FDG uptake in the entire body.



Fig. 3 – Positron emission tomography-computed tomography (PET/CT) findings. The nodular mass lesion shows FDG hot uptake (A-C). Oval-shaped FDG uptake lesion presumed to be the right ovarian or seeding nodule (D-F). Focal FDG uptake was also observed at the site of the uterine cervix or body (G-I).

Omental biopsy was performed and 2 pieces of pinkish soft tissues measuring $1.3 \times 1.3 \times 0.5$ cm were obtained. Immunohistochemical staining revealed WT1 (focal +), CDX2 (–), CK20 (–), CK7 (+), ER (focal +), and PR (focal +), consistent with serous carcinoma (low grade). Considering the PET/CT and biopsy results, the possibility of metastasis from right ovarian cancer was thought to be high and the patient underwent surgery.

The patient underwent radical hysterectomy, both salpingo-oophorectomy and omentectomy. Both ovaries and uterus were intact. Several peritoneal seeding nodules were identified, and metastasis to the left fallopian tube was confirmed.

The size of the surgically excised omental tissue was $35 \times 18 \text{ cm}$ (Fig. 4A), and conglomerated masses were observed on cross section. The largest mass was $8 \times 8 \text{ cm}$ and showed solid white gray tissue (Fig. 4B). In hematoxylin and eosin (H & E) stains, cells around the omental fat showed hypercellularity suggesting malignancy at 40 magnification (Fig. 4C) and 200 magnification (Fig. 4D), which was ultimately confirmed as PPSC.

The total weight of the uterus and the organs attached to each side was 54 g, and the size of the uterus was $7 \times 3.5 \times 2.5$ cm. The uterine cervix was nonspecific and the endometrium was atrophied with overall bleeding. No specific findings in the myometrium were noted. The fimbria portion of the left fallopian tube was attached with a grayish white mass which measured 2×1.5 cm in size. The other organs showed no specific findings. Retrospectively, the region that was considered as a right ovarian or seeding nodule in PET/CT was thought to be an FDG uptake by omental seeding nodule. The area thought to be uterine cervix was found to be an FDG uptake due to adenomyosis of the adjacent lower body of uterus. In addition, there was tumor cell infiltration in the left fallopian tube; however, there was no FDG uptake on PET/CT, presumably due to its tiny size.

The primary lesion was omentum; the patient was diagnosed as having PPSC, and chemotherapy was performed as the first-line therapy with taxol and carboplatin. Follow-up CT at 3 months after operation showed no evidence of remnant or recurring soft tissue mass in lower abdominal and pelvic cavities (Fig. 5).

Discussion

PPSC is an epithelial tumor that arises from the peritoneum. On histopathologic analysis, it resembles a malignant ovarian surface epithelial stromal tumor [1]. PPSC almost always occurs in postmenopausal women (mean age, 56-62 years) [2,3].

In the United States, the incidence of PPSC is 6.78 cases per 1,000,000 individuals [4]. It is known that the risk of PPSC is high in patients with germinal mutations of BRACA-1 and BRACA-2 [5].

The common clinical symptoms of PPSC are abdominal distension, abdominal pain, and discomfort.



Fig. 4 – The patient underwent radical hysterectomy and both salpingo-oophorectomy and omentectomy. On gross examination, several masses were observed on the cross-section (A). The largest mass was 8 \times 8 cm and showed solid white gray tissue (B). On microscopic examination (hematoxylin and eosin, 40 \times), hypercellular cells were visualized, suggesting malignancy around the omental fat vacuole (C). Cells were diagnosed as peritoneal serous carcinoma (hematoxylin and eosin, 200 \times) (D) and were ultimately confirmed as primary peritoneal serous carcinoma.



Fig. 5 – Follow-up CT at 3 months after operation. There is no evidence of remnant or recurred soft tissue mass.

These symptoms are similar to those of peritoneal carcinomatosis. Therefore, it is difficult to diagnose PPSC prior to surgery. The diagnostic criteria of PPSC as described by the Gynecology Oncology Group include (1) ovaries must be normal size or enlarged as a result of the benign process, (2) extraovarian involvement must be greater than the surface involvement of either ovary, (3) ovarian involvement must be absent, confined to the ovarian surface epithelium without stromal invasion, or involve the cortical stroma with a maximum tumor dimension of less than $5 \times 5 \text{ mm}^2$, and (4) histopathologic and cytologic characteristics of the tumors should be similar to those of epithelial ovarian cancer. These criteria aid in differentiation from primary serous carcinoma of ovary [6].

Ascites, peritoneal nodules or thickening, and omental nodules or masses are the most common imaging features. The peritoneal recesses of the upper abdomen, particularly the subphrenic spaces, should be carefully evaluated for the presence of the disease because these areas are major sites of lymphatic clearance of the peritoneum. Peritoneal and omental nodules and masses are enhanced by intravenous contrast material during CT and MR imaging. Calcifications within peritoneal and omental nodules represent psammoma bodies histopathologically [7].

PET/CT is useful for assessing tumor origin, extent, and distant metastasis. However, in our case, ovarian and seeding nodules could not be distinguished completely on PET/CT.

PPSC is known to be positive for cytokeratin-7, CA-125, estrogen receptor, and Wilms' tumor-1 (WT-1) on immunohistochemical staining [8]. In our case, estrogen receptor, WT-1, CA-125 positivity was noted.

Treatment of PPSC involves surgical resection and platinum-based chemotherapy. Intraperitoneal chemotherapy reportedly has survival benefit over surgery alone or surgery and systemic chemotherapy [6].

The median survival time of PPSC is 11-17 months, which means poor prognosis [9]. Therefore, early diagnosis and treatment is important, and further imaging studies should be performed.

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