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Case Report

Solitary primary peritoneal carcinoma arising from the omentum

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ABSTRACT

Objective: To report a case of isolated omental peritoneal carcinoma without peritoneal carcinomatosis.
Case report: A 60-year-old female with abdominal distention was found to have a pelvic mass. Under the impression of ovarian cancer, laparotomy was performed only to show one isolated mass over omentum. Serial examination and pathology study including immunochemical staining indicated primary peritoneal serous carcinoma.
Conclusion: Isolated omental peritoneal carcinoma without peritoneal carcinomatosis and ascites is rare, and whether this represented a unique entity with different chemotherapy response and treatment outcome from the disseminated form of primary peritoneal carcinoma needs to be reviewed in the future.

Introduction

Primary serous peritoneal carcinoma (PSPC) is a malignancy that originates from the peritoneum with typical features of diffuse peritoneal involvement and ascites. Despite the common clinical features shared by PSPC and epithelial ovarian carcinoma, the incidence of PSPC is considerably lower than that of epithelial ovarian carcinoma (6.78 cases per million vs. 120.5 cases per million, respectively) [1]. However, the true incidence of PSPC is unknown owing to inconsistent definitions and diagnostic criteria from retrospective data. The most common presenting symptoms of PSPC are abdominal distension (59.1%) and abdominal pain (19.0%), and ascites are present in 63.6% of cases [2]. Almost all cases of PSPC involve disseminated disease at diagnosis [3]; only a few cases of solitary PSPC have been reported.

We report a case of PSPC mimicking a solitary pelvic tumor with an unusual pattern of spread. To the best of our knowledge, primary peritoneal carcinoma developing on the omentum as a single tumor without peritoneal carcinomatosis has not previously been reported. At present, limited data are available with regard to the optimal treatment strategy and prognosis of localized primary peritoneal carcinoma. Despite its rarity, isolated omental peritoneal carcinoma should be included in the differential diagnosis of pelvic tumors. This case could provide a new paradigm for isolated peritoneal carcinomas.

Case presentation

A 60-year-old woman, gravida 5 parity 2, came to our hospital because of a recent diagnosis of a pelvic tumor. She had been well until 6 months earlier when low abdominal discomfort developed, followed by abdominal distention. One week prior to this consultation, she noted a mass over her right lower abdomen. An evaluation at a local clinic led to the diagnosis of a pelvic tumor, and she was referred to our hospital.

Menopause had occurred at the age of 51 years without hormone replacement therapy. There was no abnormal bleeding or change in bowel or bladder habits, and results of general examinations were normal. On gynecologic examination, her uterus was found to be of normal size, although a firm and movable adnexal mass about fetal head in size was noted without motion tenderness. Transvaginal and transabdominal ultrasonography showed a 9.1 cm × 8.9 cm right adnexal solid tumor with an irregular border and heterogeneous content. Data taken from her chest X-ray, electrocardiogram, complete blood count, serum electrolyte measurements, and liver and renal-function tests were within normal limits. Tumor markers were checked with a mild elevation of serum

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CA-125 level to 56.9 U/mL (normal range <35 U/mL) and a normal level of carcinoembryonic antigen (1.54 ng/mL; normal range <2.5 ng/mL). Computed tomography (CT; Fig. 1) of the abdomen and pelvis showed a large pelvic mass of about 12 cm, extending to the right lower abdomen with a cystic component and heterogenous enhancement. Diffuse peritoneal thickening and omental infiltration suggested right ovarian cancer with cancerous peritonitis. There were no ascites or definite lymphadenopathy.

On laparotomy, a solitary solid mass arising from the omentum was identified (Fig. 2A and B). The tumor was whitish in color with some central yellow necrosis. The uterus and bilateral adnexa were intact without tumor involvement. A thorough inspection of the abdomen and pelvic cavity revealed no gross tumor seeding or metastases. Intraoperatively, a frozen section of the tumor showed high-grade carcinoma. Omentectomy, total abdominal hysterectomy, bilateral salpingo-oophorectomy, and pelvic and para-aortic lymph-node dissection were then performed, with optimal debulking surgery without gross residual tumor. A pathological examination revealed extensive necrosis (Fig. 3A) of the tumor that was composed of pleomorphic and hyperchromatic cells with brisk mitosis (Fig. 3B). The frequent sarcomatoid features of tumor spindling and bizarre giant cells were also noted. Immunohistochemically, these neoplastic cells showed diffusely and strongly positive for p53 (Fig. 4A), p16 (Fig. 4B), and PAX-8. However, peritoneal malignant mesothelioma markers including cytokeratin 5/6 (Fig. 4C) or calretinin (Fig. 4D) were negatively stained. No neoplastic cells were seen in bilateral ovaries, and no *in situ* adenocarcinoma was identified at bilateral fallopian tubes or uterus. Both cytological and histopathological examinations of peritoneal washings and regional lymph nodes yielded negative results for malignant cells or metastatic cancer cells. A high-grade primary peritoneal serous carcinoma, FIGO (International Federation of Gynocology and Obstetrics) stage IIIC (pT3N0M0), was thus diagnosed. Chemotherapy with paclitaxel (175 mg/m^2) and carboplatin (area under the curve = 6) every 3 weeks was arranged for six cycles.

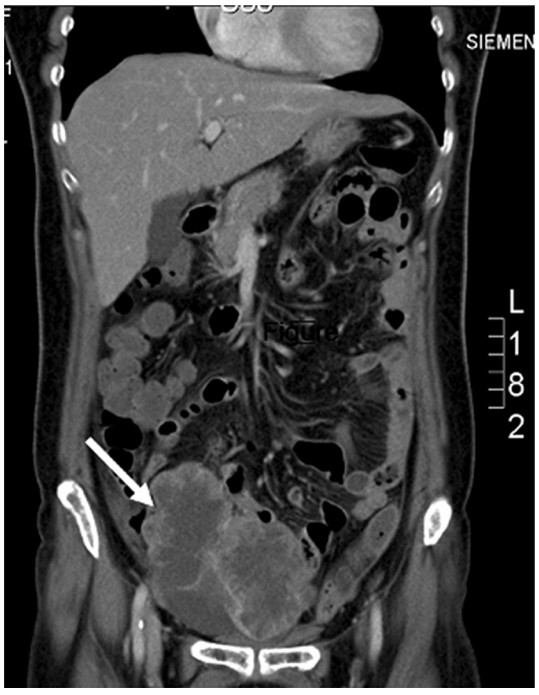

Fig. 1. Computed tomographic scan showed one 10-cm hypervascular, heterogeneous mass (arrow) in the right lower pelvic cavity.

The serum CA-125 level normalized after the surgery, and there was no evidence of recurrence after 6 months of follow-up.

Discussion

PSPC, first described in 1959 [4], is a malignancy arising from extraovarian mesothelial cells with müllerian potential, and different nomenclatures have been used including serous surface papillary carcinoma, primary peritoneal carcinoma, extraovarian pelvis serous carcinoma, and psammomacarcinoma [5]. A diagnosis of primary peritoneal carcinoma is based on the Gynecologic Oncology Group criteria: (1) both ovaries are normal in size or enlarged by a benign process; (2) extraovarian involvement is greater than ovarian involvement; (3) absence of deep-seated invasive ovarian carcinoma or cortical invasion with tumors that measure less than 5 × 5 mm^2; and (4) the tumor is of a serous cell type [6]. Our case fit these criteria as the omental mass showed serous carcinoma without other organ involvement.

The most common presenting symptoms of PSPC are abdominal distension, abdominal pain, and poor appetite. The differential diagnosis of a solitary pelvic mass includes pedunculated uterine fibroid and ovarian tumors. Typical fibroid tumors are well-defined masses of low-signal intensity compared to the myometrium on magnetic resonance imaging T2-weighted images. Pelvic examinations may give a clue to the diagnosis; however, occasionally the correct diagnosis is not made until the surgery. The typical CT features of PSPC are a large volume of ascites, mesenteric or omental mass, peritoneal thickening, and lack of an obvious ovarian tumor. However, the CT findings were misleading in our case because there were no ascites or classic "omental cake" appearance except for one pelvic mass.

PSPC is clinically and histologically difficult to differentiate from stage III or IV ovarian serous papillary carcinoma (OSPC). A preoperative diagnosis is unlikely to be based solely on image findings; nonetheless, exploratory laparotomy can offer a diagnosis and staging evaluation. However, PSPC with distant metastasis at pleura and supraclavicular lymph node but without peritoneal carcinomatosis had been reported in two cases [7,8]. Kim et al [9] reported the first case of the localized form of PSPC, which was a single primary tumor originating from the peritoneal lining of the sigmoid colon. Our case is unique in its presentation of an isolated omental mass without peritoneal spread. To the best of our knowledge, there have not been any reported cases of solitary primary peritoneal carcinoma arising from the omentum.

Pretreatment CA-125 level is an effective tumor marker for diagnosing, correlating with clinical status, and reflecting disease dissemination. Preoperative CA-125 values have been reported to be significantly elevated in 90.5% of PSPC patients with all stages, and in 95% of patients with stages II–IV [2]. Roh et al [10] reported that 21 out of 22 PSPC patients had elevated preoperative CA-125 values with a median value of 706 U/mL (range, 35–7647 U/mL). CA-125 level can also be used for the evaluation of treatment response and disease recurrence [11]. The solitary PSPC patient reported by Kim et al [9] also showed a slightly elevated serum CA-125 level of 44.6 U/mL. In our case, the preoperative serum CA-125 level was also slightly elevated at 56.9 U/mL, which dropped to 13.4 U/mL after six cycles of adjuvant chemotherapy. CA-125 could therefore have been used as a surrogate marker for this patient.

The treatment and prognosis of PSPC are similar to OSPC with platinum-based chemotherapy as prescribed in our patient. Ayhan et al [12] reported 32 PSPC and 43 OSPC patients who received adjuvant cisplatin (carboplatin) and paclitaxel after cytoreductive surgery. No significant differences were noted with respect to clinical or surgical response, progression-free survival (14 months for both groups), or overall survival (30 months for PSPC and 28