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CASE REPORT

Serous adenocarcinoma of retroperitoneum: a case report

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Abstract Primary retroperitoneal serous adenocarcinoma (PRSA) is an extremely rare malignancy, with only seven cases having been previously reported. We report a case of PRSA in a 42-year-old woman treated with surgical resection and adjuvant chemotherapy. The histopathological findings of PRSA resemble those of ovarian serous carcinoma, which indicates that a combination of complete surgical resection with adjuvant chemotherapy may be the best treatment option for PRSA.

Keywords Serous adenocarcinoma · Retroperitoneal tumor

Introduction

Primary retroperitoneal tumors are rare, accounting for just 0.2–0.3% of all tumors. The histology of primary retroperitoneal tumors is generally fibrosarcoma, lymphoma, teratoma, etc. However, having epithelial neoplasm occur with retroperitoneum is rare, accounting for just that only 3.1% of all primary retroperitoneum tumors [1]. A primary retroperitoneal serous adenocarcinoma (PRSA) is an extremely rare malignancy, and is a subtype of the primary serous carcinoma of the peritoneum (PSCP), with only seven other cases previously reported. In this study, we report a 42-year-old woman with PRSA who underwent a surgical resection followed by adjuvant chemotherapy consisting of a combination of paclitaxel and carboplatin.

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Case report

A 42-year-old woman presented to our department with lower-left back pain. Her family history was not contributory. She had a previous history of a surgical resection of a left ovarian chocolate cyst 10 years ago. Her general appearance was thin and her body mass index (BMI) was 14.6 kg/m^2. Abnormal laboratory test values included only a CA (cancer antigen) 19-9 level of 177 IU/ml, along with a CA125 level of 10 IU/ml, which is within the normal limit. Ultrasonography revealed a cystic mass 6 cm in diameter located at the lower pole of the left kidney, where it was possible that there might be some association between the tumor and left low back pain.

A computed tomography (CT) scan showed the tumor had a mass of 62 × 55 × 55 mm and consisted of cystic lesion and solid component in the cranial side of the tumor. A magnetic resonance imaging (MRI) of the kidney indicated high signal intensity in the T2-weighted images and low signal intensity in the T1-weighted images (Fig. 1). The cystic mass extended into the retroperitoneum and was adjacent to the lower pole of the left kidney. These findings led to a preoperative diagnosis of a malignant retroperitoneal neoplasm.

Surgical resection of the retroperitoneal cystic mass was performed. There appeared to be no involvement of the tumor with ipsilateral kidney or iliopsoas muscle. The

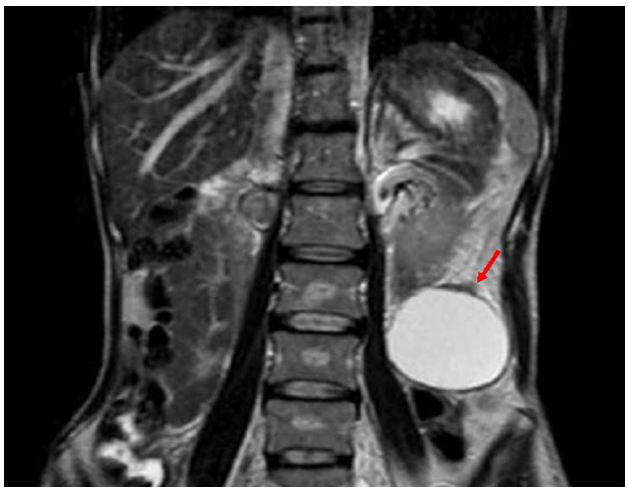

Fig. 1 Coronary T2-weighted MRI findings of retroperitoneal serous adenocarcinoma. The cystic mass was extending into the retroperitoneum and adjacent to the lower pole of the left kidney

tumor was markedly adhesive to the peritoneum and was suspected to have invaded the peritoneum, so, we performed en bloc resection including a part of the peritoneum.

The size of the resected tumor was 80 × 70 × 70 mm (Fig. 2). The resected tumor was cystic and had a well-encapsulated smooth external surface. The content of the cystic tumor was yellowish and serous fluid. The cut surface of the tumor showed a solid component in the cranial side of the tumor. No apparent hemorrhage or necrosis was present. The histopathological examination demonstrated cribriform and small alveolar proliferation of atypical columnar cells at the solid component of the resected tumor (Fig. 3). The internal cyst wall was covered by low polypoid atypical epithelium. The tumor cells were immunohistochemically positive for p53, CEA, CA125, cytokeratin (CK) 7, and Ber-EP4, while being negative for CK20. As there was no evidence of tumor tissue at the surgical margin, the tumor was defined as completely resected. On the basis of these findings, the resected tumor was histologically diagnosed as serous adenocarcinoma arising from retroperitoneum.

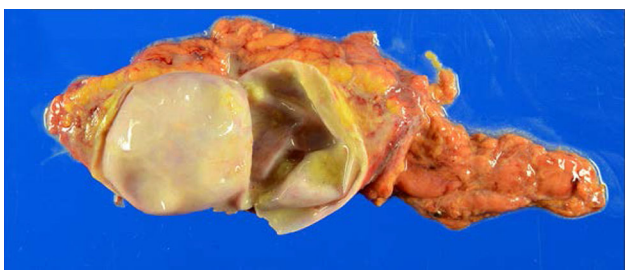

Fig. 2 Cut surface of the resected tumor showed monoocular cyst with a solid component

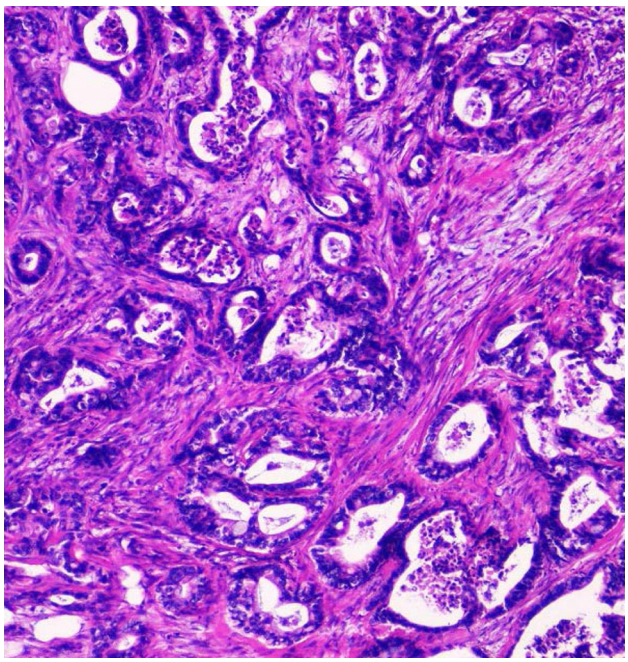

Fig. 3 Microscopic findings of the tumor revealed cribriform and small alveolar proliferation of atypical columnar cells

Serum level of CA19-9 decreased to 9 IU/ml, within the limit of normal after surgery. Because the pathological findings of the tumor were closely similar to serous adenocarcinoma of the ovary, adjuvant chemotherapy based on the regimen for ovarian cancer was performed (paclitaxel 150 mg/m^2 on day 1 plus carboplatin AUC 5 on day 1, every 3 weeks for 2 cycles). After adjuvant chemotherapy, the patient has been followed up for 5 months without clinical evidence of tumor recurrence in imaging study nor the elevation of serum CA19-9 level.

Discussion

Primary serous carcinoma of the peritoneum (PSCP) is rare while serous adenocarcinoma of the ovary is a common subtype of ovarian epithelial neoplasm. Swerdlow et al. were the first to describe serous adenocarcinoma that was found in a patient with peritoneal carcinomatosis with no known primary lesion [2]. The Gynecologic Oncology Group (GOG) developed a concise set of criteria for this diagnostic category as follows [3]: (1) both ovaries must be either physiologically normal in size or enlarged by a benign process. (2) Involvement in the extraovarian sites must be greater than involvement on the surface of either ovary. (3) Microscopically, the ovarian component must be one of the following: (a) nonexistent; (b) confined to the ovarian surface epithelium with no evidence of cortical invasion; (c) involving ovarian surface epithelium and

Table 1 Reports of retroperitoneal serous adenocarcinoma since 1983

	Age	Sex	Past hormonal therapy	Initial presentation	Size (mm)	Elevated tumor markers	Growth pattern	METASTASES	Surigical form	Elevated tumor markers of recurrence	Chemotherapy	Outcomes
Ulbright [4]	11	F	None	Weight gain	18 × 13 × 11	None	Solid	None	Partial resection of the tumor	Not mentioned	DTX + CTX + CDDP	NED 10 months
Caruncho [5]	49	F	None	Abdominal pain	90 × 60 × 50	CEA	Solid and cystic	None	Partial resection of the tumor	Not covered	None	Not mentioned
Kurosaki [6]	38	F	None	Abdominal distention	60	CEA	Cystic	None	Complete resection with partial nephrectomy	Not covered	None	NED 24 months
Fujiwara [7]	54	F	None	Abdominal distention	None	CA125	Invasive	Periaortic lymph nodes	None	Not covered	CTX + Carbo	DOD 24 months
Kaku [8]	44	F	None	Elevated serum CA19-9	60 × 35 × 30	CA125 CA19-9	Cystic	None	Complete resection with a partial resection of the psoas muscle	CA125 CA19-9	None	AWD 23 months
Iura [9]	66	F	Estrogen stimulation (10 years)	Abdominal pain	200 × 95 × 85	CA125 CA19-9	Solid and cystic	None	Complete resection with a partial resection of the ileocecum	Normal	PTX + Carbo	AWD 32 months
Arichi [10]	75	F	None	Physical examination	38 × 47 × 50	CA125	Cystic	None	Complete resection with a partial resection of the diaphragm	Not covered	DTX + Carbo	NED 6 months
Present case	42	F	Estrogen stimulation (6 years)	Left back pain	55 × 62 × 55	CA19-9	Cystic	None	Complete resection of the tumor	Not covered	PTX + Carbo	NED 3 months

NED no evidence of disease, *DOD* died of disease, *AWD* alive with disease

underlying cortical stroma but with any given tumor size less than 5 × 5 mm, or (d) tumor less than 5 × 5 mm within ovarian substance associated with or without surface disease. (4) The histological and cytological characteristics of the tumor must be predominantly of the serous type that is similar or identical to ovarian serous papillary adenocarcinoma, any grade.

PRSA, which is a subtype of PSCP, has been reported in only seven cases (Table 1) [4–10]. The clinical features and histopathological findings of PRSA resemble those of serous adenocarcinoma of the ovary. All seven of the previously reported cases were women. The details of the tumor origin in PRSA remain unclear. Some possibilities of tumor origin include coelomic metaplasia, extra-ovarian endometriosis, supernumerary ovary, teratoma, and enterogenic cyst (enteric duplication cyst or enteric cyst). However, coelomic metaplasia is the most widely accepted as tumor origin [11–13]. The secondary Müllerian duct hypothesis may also be applicable to the development of PRSA.

Previous studies have demonstrated four cases of PRSA completely resected including adjacent organs. The gross appearance of PRSA was cystic in three cases, while a combination of solid components with cystic lesion made up the final case. These findings suggest that PRSA tends to have a cystic pattern and localized growth in the retroperitoneum. The surgical option of a total hysterectomy and bilateral salpingo-oophorectomy is useful to rule out the possibility of the metastasis from primary gynecological malignant lesions. On the other hand, it is important to consider the possibility of excessive invasion in the case with PRSA. There has been a recent report of a subset of serous carcinoma in the ovary that was thought to originate from the fallopian tube [14]. In the present case, such possibility could not be ruled out because of the lack of hysterectomy and bilateral salpingo-oophorectomy.

Adjuvant chemotherapy was considered because PRSA pathologically resembles serous adenocarcinoma of the ovary. Kaku et al. have suggested that a combination chemotherapy consisting of docetaxel with carboplatin may be a good option for primary PRSA as the first-line chemotherapy based on the histologic similarity between retroperitoneal epithelial tumors and ovarian epithelial carcinomas [8]. The histopathological findings of the positive surgical margins, local tumor infiltration, and locoregional lymph node involvement have been reported as risk factors [8]. In the cases with these findings, immediate chemotherapy might be recommended postoperatively.

The prognosis of PRSA remains unclear due to the rarity of reported cases. Among the seven cases previously reported, one patient died of the PRSA 24 months after initial presentation and two showed a local recurrence within 2 years of surgical treatment.

We treated a 42-year-old woman with PRSA using treatment plan that included both a surgical complete resection and adjuvant chemotherapy, although a definite therapeutic strategy for PRSA has not been established. Accumulation of evidences from further reports is required to clarify the biological characteristics and optimal management of PRSA.

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