Report of a case: Retroperitoneal mucinous cystadenocarcinoma with rapid progression

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A 62-year-old woman with a complaint of abdominal discomfort was admitted to our hospital. Her right lower abdomen was mildly swollen without pain and tenderness. Abdominal CT showed multiple cystic lesions in the retroperitoneum (Fig. 1A). Those cysts displaced the right kidney to the median of the abdomen (Fig. 1B). No solid component was found inside the cysts. Abdominal MRI showed 4 cysts, 9 × 10 cm, 7.5 × 9.5 cm, 4.5 × 6.5 cm, and 2.5 × 3 cm in size, respectively (Fig. 1C). Two cranial cysts were low in T1 weighted images and high in T2 weighted images, suggesting two different components existed (Fig. 1D). The smaller cysts were prospected to contain some blood. The internal intensity of those cysts was homogeneous, and there was no mural nodularity. Blood examination showed no particular abnormality, and serum CEA and AFP were within normal range.

1. Introduction

Retroperitoneal mucinous cystic neoplasms are very rare, and little is known about the etiology of the disease. Malignant forms of these, primary retroperitoneal mucinous cystadenocarcinoma (PRMC), are exceedingly rare, with only 63 cases, including our case, having been described in the literature to date [1–7]. Here, we report a case of a large multicyastic PRMC which took an unexpectedly aggressive clinical course despite finding only a relatively focal area of adenocarcinoma.

2. Case report

A 62-year-old woman with a complaint of abdominal discomfort was referred to our hospital. Her right lower abdomen was mildly swollen without pain and tenderness. Abdominal CT showed multiple cystic lesions in the retroperitoneum (Fig. 1A). Those cysts displaced the right kidney to the median of the abdomen (Fig. 1B). No solid component was found inside the cysts. Abdominal MRI showed 4 cysts, 9 × 10 cm, 7.5 × 9.5 cm, 4.5 × 6.5 cm, and 2.5 × 3 cm in size, respectively (Fig. 1C). Two cranial cysts were low in T1 weighted images and high in T2 weighted images, suggesting two different components existed (Fig. 1D). The smaller cysts were prospected to contain some blood. The internal intensity of those cysts was homogeneous, and there was no mural nodularity. Blood examination showed no particular abnormality, and serum CEA and AFP were within normal range.

She underwent operation under the diagnosis of retroperitoneal cystic neoplasms. Preoperative aspiration of the cysts was not attempted to avoid iatrogenic dissemination. Transabdominal resection of the cysts was performed (Fig. 2). The cysts could mostly be dissected off from retroperitoneal tissue without
Fig. 1. (A) Abdominal CT showed multiple retroperitoneal cystic lesions (arrowhead). (B) The cystic lesions displaced the right kidney to the median of the patient’s body (arrowhead). (C) Abdominal MRI (T2 weighted image) showed homogenous cysts (arrowhead). (D) Abdominal MRI (T1 weighted image) showed different intensities of cystic lesions (arrowhead).

difficulty except for a firm adhesion at the caudal tendon of the right psoas muscle, suggestive of the origination of the cysts. Her appendix and ovary appeared normal. Two cranial cysts contained approximately 700 ml of transparent colorless mucinous fluid, and other caudal cysts contained approximately 400 ml of transparent black mucinous fluid, which turned out to be slight contamination of blood. Those fluids were mucinous, but not as phlegmatic as pseudomyxoma peritonei. Cytological examination for those fluids revealed no malignant cells.

Fig. 2. Operative finding: cystic lesion (arrow head) displaced the ascending colon (black dot).

The cyst wall was lined with epithelial cells (Fig. 3A). However, most of the epithelial liner was either attenuated or denuded, and the cyst wall showed areas of erosions and patchy hemorrhage. Immunohistochemical studies were strongly positive for CA19-9, and negative for CEA. Ovarian-type stroma was seen in the cyst wall, however, immunohistochemical staining with estrogen receptor and progesterone receptor antibodies was negative. There was focal involvement by adenocarcinoma in the caudal smaller cyst, approximately 2 × 2 mm in size, 1 mm in depth, and the resection margins were negative for carcinoma (Fig. 3B). The post operative course was uneventful, and the patient had been under watchful observation.

Eight months later, the patient complained of back pain, and CT revealed osteoclastic lumbar bone metastasis at L4 (Fig. 4). Serum CA19-9 was elevated at 160U/ml. 40 Gy of irradiation to the lumbar bone was performed immediately, and chemotherapy with S-1, an oral fluoropyrimidine, and docetaxel (oral S-1 of 80 mg/m² for the first two weeks in 3 weeks and injection of docetaxel of 60 mg/m² on day 1 in 3 weeks) were administered. However, the disease never responded to chemotherapy, and rapidly spread to the retroperitoneum (Fig. 5). Eventually, the patient died of the disease 15 months after surgery.

3. Discussion

Retroperitoneal cysts are so rare that the accurate incidence is not available. There was a study that estimated the incidence of retroperitoneal cysts in addition to mesenteric cysts to be 10 for every one million hospital admissions, and the incidence of malignancy was 3% [8].

Retroperitoneal cystic masses are divided into neoplastic lesions (cystic lymphangioma, mucinous cystadenoma, cystic teratoma, cystic mesothelioma, mullerian cyst, epidermoid cyst, tailgut cyst, bronchogenic cyst, cystic change in solid neoplasms, pseudomyxoma retroperitonei, perianal mucinous carcinoma, and ancient
Fig. 3. (A) Hematoxylin and eosin (H & E) stained sections reveal a cyst wall focally lined with mucinous epithelial cells and showing partial erosion and patchy hemorrhage. (B) A small focus of moderately to poorly differentiated adenocarcinoma arising in a background of mild chronic inflammation in the caudal smaller cyst seen on this H & E stained section.

Fig. 4. Abdominal CT, taken 8 months after the surgery, showed osteoclastic lumbar bone metastasis at L4.

Fig. 5. (A) Abdominal CT, taken 15 months after the surgery, showed massive osteoclastic bone metastasis at L4. (B) In addition to progression of osteoclastic bone metastasis, massive local recurrence of retroperitoneal mucinous cystadenocarcinoma had developed.

Several hypotheses have been postulated regarding the pathogenesis of retroperitoneal mucinous cystadenocarcinoma: (1) heterotopic ovarian tissue [14], (2) monodermal variant of teratomas [15], (3) embryonal urological remnants [16], (4) intestinal duplication [17], and (5) coelomic metaplasia [18–20], which is most well-described. During embryogenesis, embryonal coelomic epithelium gives rise to the peritoneal mesothelium, the germinal epithelium of the ovary, and the Mullerian duct. In the patient of retroperitoneal mucinous cystadenocarcinoma, inclusion cysts of the coelomic epithelium could be formed and left behind in the retroperitoneum during this differentiation process. Those cysts could receive stimulation by steroids and inflammation for many years and eventually develop into true retroperitoneal cystic neoplasms. Coelomic epithelium is capable of differentiating into serous (tubal), mucinous, endometrioid, and transitional epithelium [21].

PRMC is an extremely rare tumor, and only 63 cases, including our case, have been described in the literature to date [1–7]. Of those, 57 were female, and 12 had mural nodules, which were considered to be signs of malignancy. Although precise prognosis is not available, available data show a wide survival range from 4 months to 10 years.

Preoperative diagnosis of PRMC is challenging. Considerable radiographic findings may include thickening and calcification of the cyst wall or mural nodules on imaging which may suggest malignancy. Aspiration cytology of cyst fluids may also help with diagnosis. CEA and CA19-9 are occasionally positive in serum, cyst fluids, and cyst wall, however, the phenomenon could be observed even in benign mucinous cystadenocarcinoma.

Currently there is no significant chemotherapy for PRMC [22–23]. Therefore, resection should be considered for
retroperitoneal cystic lesions in case the lesion contains carcino-
ma, especially when the lesions are large or malignancy is
suspected in imaging. Additional salpingo-oophorectomy has been
performed by some gynecologists to avoid stimulation of female
hormone with the assumption that those lesions could be hormone
sensitive. However, there is limited evidence around the benefit of
this procedure [24]. In addition, marsupialization and aspiration
also have risks of recurrence and dissemination.

In the presented case, although the lesions were large and
multiple, there was no sign suggestive of malignancy on CT and
MRI. Cytology of the cyst fluid did not reveal malignancy, and
the component of adenocarcinoma was so tiny for the large cysts.
However, the patient passed away after rapid and aggressive course
of metastasis and recurrence. We suspected that variance in bio-
logical aggressiveness existed in PRMCs, and the presented case
could be a ‘high-grade’ one. Immunohistochemistry for CA19-9
was positive in the specimen, and serum CA19-9 was increased, as
the disease progressed. Adenocarcinoma was found in the smaller
cysts which contained some blood in the cyst fluid. In retrospect,
the suspicion of blood in the cyst fluid on MRI could have been
a soft sign of malignancy in this case. Finally, despite no intra-
operative spillage of the cyst fluid, and negative surgical margins
in all of the respected cysts, the clinical course of the disease
after surgery was unexpectedly aggressive. We considered the
remote possibility of remnant cysts, but could not definitively rule
it out.

PRMC is a rare tumor; however, it is urgently necessary to elu-
cidate the etiology of and effective therapy for the disease.

Conflicts of interest
All authors have no conflict of interest.

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Consent
Written informed consent was obtained from the patient for
publication of this case report and accompanying images. A copy
of the written consent is available for review by the Editor-in-Chief
of this journal on request. Identifying details were omitted in the
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Authors contribution
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Hirohiko Kamiyama was a clinician in charge, and wrote
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