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Report of a case: Retroperitoneal mucinous cystadenocarcinoma with rapid progression

Hirohiko Kamiyama[a,*], Ai Shimazu[a], Yurika Makino[a], Ryosuke Ichikawa[a], Takahiro Hobo[a], Shuei Arima[a], Shigeo Nohara[a], Yuji Sugiyama[a], Masafumi Okumura[a], Masahiko Takei[a], Hiroyoshi Miura[a], Koji Namekata[a], Hidenori Tsumura[a], Motoi Okada[b], Masaru Takase[b], Fumio Matsumoto[a]

[a] Department of Surgery, Koshigaya Municipal Hospital, Higashikoshigaya 10-47-1, Koshigayashi, Saitama 343-8577, Japan
[b] Department of Pathology, Koshigaya Municipal Hospital, Higashikoshigaya 10-47-1, Koshigayashi, Saitama 343-8577, Japan

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ABSTRACT

INTRODUCTION: Retroperitoneal mucinous cystic neoplasms are uncommon, and little is known about the etiology of the disease. Malignant forms of these are extremely rare. Here, we report a case of primary retroperitoneal mucinous cystadenocarcinoma (PRMC), which demonstrated unexpectedly aggressive progression despite finding only a limited area of adenocarcinoma.
PRESENTATION OF CASE: A 62-year-old woman with a complaint of abdominal discomfort was admitted to the hospital. Abdominal CT and MRI showed multiple large retroperitoneal cysts dislocating the right kidney nearly to the center of the abdomen. Transabdominal resection of the cysts was performed. Those cysts contained 1100 ml of mucinous fluids in total. Cytological examination of those fluids revealed no malignant cells. The cyst wall was lined with mucinous epithelial cells, and contained some ovarian-type stroma. Also, there was a focal area of adenocarcinoma in the cyst wall, and the lesion was diagnosed as primary retroperitoneal mucinous cystadenocarcinoma. Eight months later, the patient developed lumbar bone metastasis. Chemotherapy with S-1, an oral fluoropyrimidine, and docetaxel had been begun immediately; however, the disease had rapidly spread in the retroperitoneum. Eventually, the patient died of the disease 15 months after surgery.
DISCUSSION: Retroperitoneal mucinous cystic neoplasms are considered to be metaplasia of embryonal coelomic epithelium. Complete excision without rupture is essential. However, variance of biological aggressiveness might exist in PRMCs.
CONCLUSION: Retroperitoneal mucinous cystadenocarcinoma is a rare tumor, and it is urgently necessary to elucidate the etiology of an effective therapy for the disease.

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 multicystic 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, and caudal smaller cysts were high in T1 and 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

* Corresponding author. Tel.: +81 48 965 2221; fax: +81 48 965 3019.
E-mail address: hkamiya@juntendo.ac.jp (H. Kamiyama).

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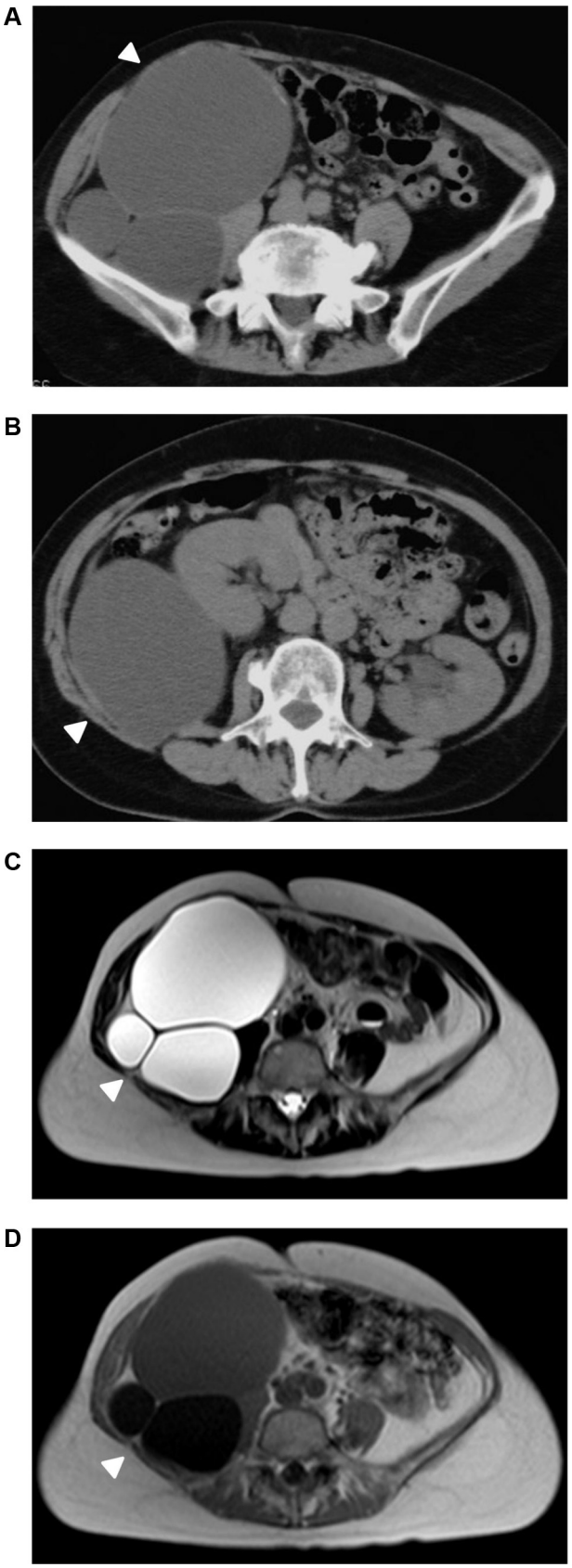

Fig. 1. (A) Abdominal CT showed multiple retroperitoneal cystic lesions (arrow head). (B) The cystic lesions displaced the right kidney to the median of the patient's body (arrow head). (C) Abdominal MRI (T2 weighted image) showed homogenous cysts (arrow head). (D) Abdominal MRI (T1 weighted image) showed different intensities of cystic lesions (arrow head).

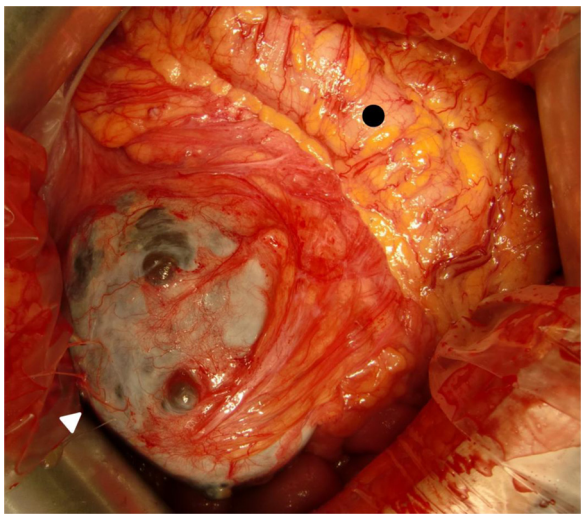

Fig. 2. Operative finding: cystic lesion (arrow head) displaced the ascending colon (black dot).

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.

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/m^2 for the first two weeks in 3 weeks and injection of docetaxel of 60 mg/m^2 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

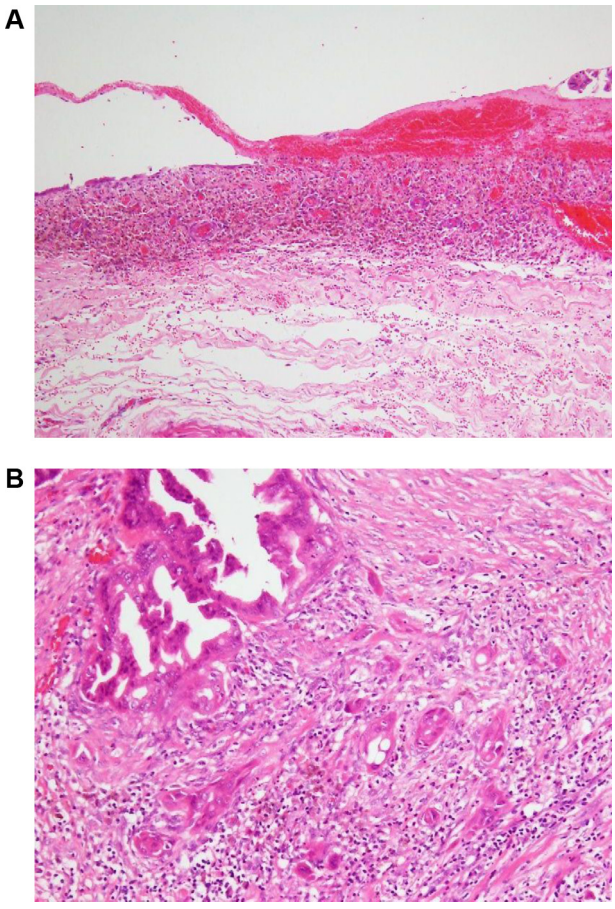

Fig. 3. (A) Hematoxylin and eosin (H & E) stained sections reveal a cyst wall focally lined with mucinous epithelial cells and showing partial erosion and patchy hemorrhage. (B) A small focus of moderately to poorly differentiated adenocarcinoma arising in a back ground of mild chronic inflammation in the caudal smaller cyst seen on this H & E stained section.

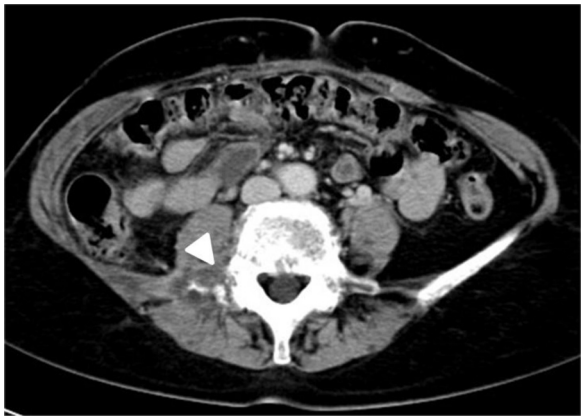

Fig. 4. Abdominal CT, taken 8 months after the surgery, showed osteoclastic lumbar bone metastasis at L4.

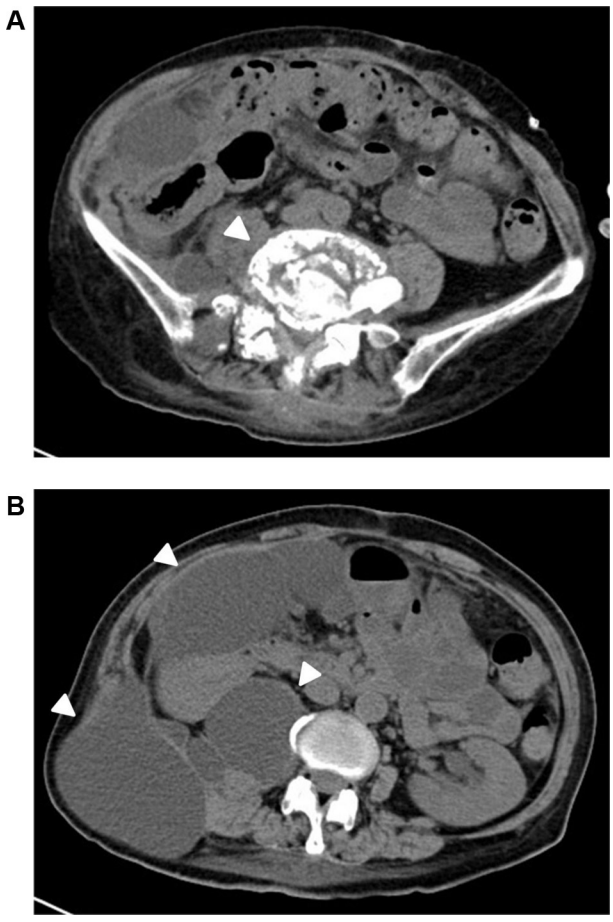

Fig. 5. (A) Abdominal CT, taken 15 months after the surgery, showed massive osteoclastic metastasis at L4. (B) In addition to progression of osteoclastic bone metastasis, massive local recurrence of retroperitoneal mucinous cystadenocarcinoma had developed.

schwannoma) and nonneoplastic lesions (pancreatic pseudocyst, nonpancreatic pseudocyst, lymphocele, urinoma, and hematoma) [9–10]. Retroperitoneal mucinous cyctadenoma and cystadenocarcinoma largely affect females, although some male cases had been previously reported [2,5–6,11–13].

Several hypotheses have been postulated regarding the pathogenesis of retroperitoneal mucinous cystadenoma/adenocaricnoma: (1) heterotopic ovarian tissue [14], (2) monodermal variant of teratomas [15], (3) embryonal urogenital 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 cystadenoma, 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 cystadenoma.

Currently there is no significant chemotherapy for PRMC [22–23]. Therefore, resection should be considered for

retroperitoneal cystic lesions in case the lesion contains carcinoma, 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 prospected that variance in biological 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 intraoperative 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 elucidate the etiology of and effective therapy for the disease.

Conflicts of interest

All authors have no conflict of interest.

Funding

All authors have no funding of research.

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 manuscript.

Authors contribution

Hirohiko Kamiyama was a clinician in charge, and wrote the manuscript. Ai Shimazu, Yurika Makino, Ryosuke Ichikawa, Takahiro Hobo, Shuei Arima, Shigeo Nohara, Yuji Sugiyama, Masafumi Okumura, Masahiko Takei, Hiroyoshi Miura, Koji Namekata, Hidenori Tsumura, and Fumio Matsumoto were clinicians in charge. Motoi Okada and Masaru Takase were in charge of pathological diagnosis.

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