


## Case Report

# Primary retroperitoneal mucinous cystic tumor with borderline malignancy: a case report and review of the literature

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### Abstract

Primary retroperitoneal mucinous cystic tumor (PRMCT) is rare and preoperative diagnosis is challenging due to their rarity, lack of specific diagnostic imaging, clinical presentation, and tumor markers. We present an extremely rare case of PRMCT of borderline malignancy and summarize the characteristics of previously reported cases. Although the prognosis and optimal therapeutic strategies require further investigation due to its rarity, we should be considered when dealing with retroperitoneal cystic lesions.

**Keywords:** primary; retroperitoneum; case report; mucinous cystic neoplasm

### Introduction

Primary retroperitoneal mucinous cystic tumors (PRMCT), including mucinous cystadenoma, mucinous cystic tumor of borderline malignancy (PBMCTB), and mucinous cystadenocarcinoma, are a very rare entity [1–3]. Since they are not commonly recognized, preoperative diagnosis is challenging. This case report aims to contribute to the understanding of this rare entity and to aid in making accurate preoperative diagnoses.

### Case report

A 50-year-old female patient initially presented 3 years ago with uterine myoma. At that time, retroperitoneal enhanced computed tomography scan revealed multiple uterine myomas and an 8.0 × 5.8 cm lobulated cystic mass with smooth border in the left lower retroperitoneum (Fig. 1a). The cystic mass was suspected to be a retroperitoneal lymphangioma, and since there were no symptoms related to it, the patient decided to undergo observation rather than surgical removal. Since then, size, number, and feature of myomas and retroperitoneal mass showed no change in the follow-up annual CT (Fig. 1b). Recently, although the patient had no specific symptoms related to the cystic tumor, she felt that her abdomen was gradually becoming more bloated. As a result, she opted for surgical removal. The evaluation for serum tumor markers was not conducted. The surgically resected tumor was a large-sized, well-demarcated multilocular cyst with multifocal wall calcification, measuring 142 g and 10.5 × 6.8 ×

4.5 cm. There were no ascites, lymphadenopathy, or adhesion to surrounding organs. Both ovaries were normal. The cyst contained clear mucinous fluid and showed focal papillary excrescences, measuring 2.4 × 2.0 cm. Microscopically, the cyst was mainly lined by a single layer of mucin-secreting columnar epithelium (Fig. 2a). In papillary excrescences, papillae lined by stratified mucinous epithelium with moderate nuclear atypia were observed (Fig. 2b). The tumor cells were positive for cytokeratin 7, and negative for cytokeratin 20 and calretinin. Ovarian-like stroma was found and showed positivity for estrogen receptor. No stromal invasion was found. Histologic diagnosis was PRMCTB. During a follow-up period of 24 months, there was no recurrence or metastasis.

### Discussion

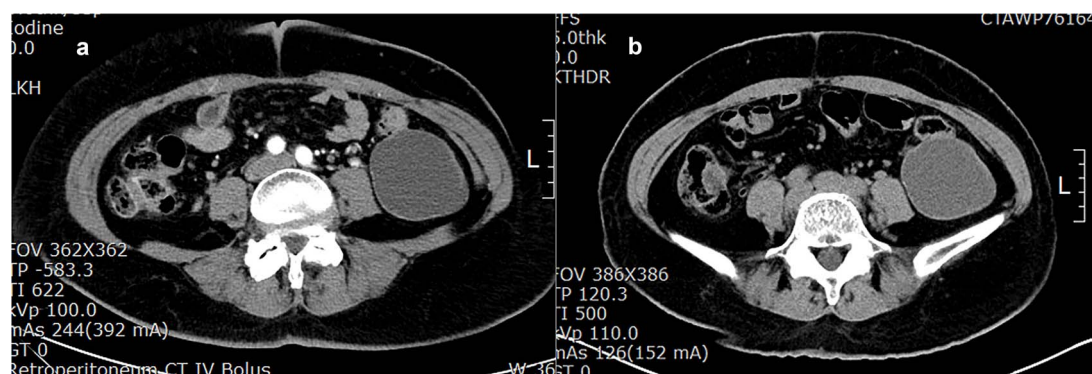
PRMCTs, including mucinous cystadenoma, PBMCTB, and mucinous cystadenocarcinoma are very rare entity and resemble ovarian mucinous cystic tumors [1–3].

Preoperative diagnosis of PRMCT is difficult due to the lack of specific clinical presentation, laboratory findings, or pathognomonic radiologic features [4]. Several hypotheses regarding the origin of PRMCT have been suggested but remain uncertain [2, 5–7]. The one is that PRMCT arises from ectopic ovarian tissue due to the histologic similarity with ovary mucinous cystic tumor. However, PRMCT has been reported in men and most of them did not contain ovarian tissue. Another is that PRMCT originates from teratoma or remnants of embryonal urogenital apparatus. However, the most widely accepted hypothesis is that PRMCT arises

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**Figure 1.** (a) Retroperitoneal enhanced CT shows a lobulated homogeneous cyst in the left lower abdomen. (b) A year later, there was no change in the size, shape, or characteristics of the cyst.

**Table 1.** Clinical features of previously reported cases of primary PBMCTB

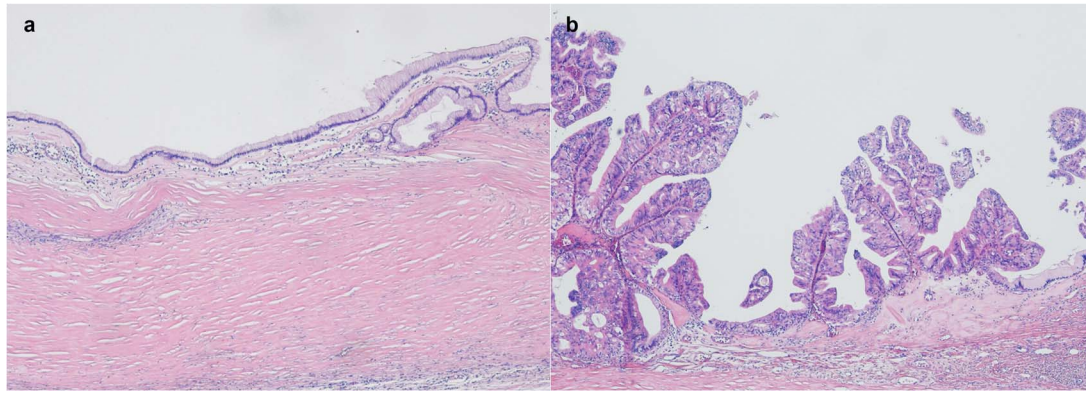
Age/Sex	Symptom	Location	Imaging finding	Size (cm)	Outcome (months)
41/F	Abdominal pain and tumor	ND	ND	12 × 10 × 9	NED (ND)
38/F	Abdominal pain and distension	Left lower abdomen	Multilocular cyst	11	Mediastinal node metastasis (48 months)
47/F	LUQ mass	Left upper quadrant	ND	13	ND
63/M	Abdominal pain	Right kidney	Cyst	11	ND
62/F	Abdominal pain	Right flank	Hypodense mass	12 × 8.5	NED (18 months)
68/M	Right abdominal mass	Right abdomen	Cyst	16 × 14.5 × 10	NED
65/F	Abdominal distension	Left abdominal cavity	Multilocular cyst	21 × 14	NED (12 months)
32/M	Incidental on radiology	Around vena cava	ND	7 × 6 × 4.4	ND
44/M	Abdominal pain	Interaortocaval	Multiple retroperitoneal mass	Multiple (2–5)	NED (12 months)
35/F	Pelvic pain	Rear of the cecum	Cyst	28	NED (24 months)
22/F	Abdominal pain	Superior of uterus	Cyst	20 × 17.5 × 15	NED (24 months)
33/F	Abdominal pain, swelling	Left flank	Cyst	ND	NED (10 months)
33/F	Tiredness	Descending colon	Cyst	15 × 13 × 9	NED (12 months)
48/F	Postprandial fullness	Ascending colon mesentery	Cyst	15 × 13 × 9	NED (8 months)
41/F	Flank pain	Right upper quadrant	Cyst	21 × 16	NED (18 months)
31/F	Abdominal distension	Right retroperitoneal space	Multilocular cyst	21 × 16 × 10	ND
36/F	Abdominal distension	Right lower abdomen	Cyst	12 × 8	NED (6 months)
39/M	Back pain	Left pararenal space	Cyst	6.9 × 4.4	NED
56/F	Painful urination	Right kidney	Multilocular cyst	28 × 22 × 8	Recur (6 months later)
45/F	Abdominal pain	Left lower abdomen	Cyst with nodule	15	NED (12 months)
25/F	Kidney mass	ND	Cyst with papillae	15	NED (148 months)
43/F	Pelvic pain	ND	Cyst with papillae	14	NED (1 months)
48/F	Enlarged mass	ND	Multilocular cyst	7	NED (34 months)
37/M	Abdominal pain, mass	Right abdomen	Multilocular cyst	22 × 11	NED (12 months)
26/F	Abdominal distension, pain	Left lower quadrant	Cyst	13 × 10 × 8	ND
65/F	Mass	ND	Septate cyst	ND	NED (6 months)
23/F	Mass	ND	Cyst	ND	ND
68/F	Palpable abdominal mass	Left kidney	Multilocular cyst	15 × 11 × 9	NED (12 months)
56/F (this)	Incidental on radiology	Retroperitoneum	Unilocular cyst	8 × 5	NED (24 months)

ND, not described; NED, no evidence of disease

from invaginated peritoneal mesothelium, which undergoes subsequent mucinous metaplasia leading to cyst formation [2, 5].

We searched the reported cases of PRMCTB using PubMed and reviewed 28 cases without carcinoma component (Table 1). Including this case, twenty-three cases were women, and 6 were men. The median age was 43.8 years (22 to 68 years). The most common symptom was nonspecific abdominal pain and distension, and it predominantly appeared as a unilocular or multilocular cyst of variable size on imaging. Tumor markers, such as CA125, CA19-9, and CEA were not useful for diagnosis. Six cases revealed ovarian-like stroma, which suggests that female

hormone may affect the development or growth of PRMCT [8]. All the tumors were removed through complete total resection, and their sizes ranged from 2 to 28 cm (mean, 14.96 cm). When reviewing the 29 reported cases so far, follow-up data for 20 patients were available, with an average follow-up period of 22.4 months (range: 1–148 months). During this period, one patient experienced a recurrence, and another patient had mediastinal lymph node metastasis. Due to the rarity and the lack of long-term follow-up data, it is difficult to determine the patient's prognosis of the PRMCTB. The management of PRMCTB is not yet well-established, but the best approach is complete surgical tumor removal. During



**Figure 2.** (a) Mucinous cystadenoma with a single layer of columnar epithelium with abundant intracellular mucin (HE, 4×). (b) PBMCTB shows papillary growth of stratified mucinous epithelium with nuclear atypia (HE, 4×).

surgery to remove the cystic mass, it is important to prevent rupture or spillage of cystic contents to reduce peritoneal dissemination and tumor recurrence [9]. Adjuvant chemotherapy after tumor resection also remains controversial. Some recommended just close follow-up, while others suggested treatment based on its histologically similar ovarian cancer [3, 4, 10, 11].

In conclusion, although PRMCTB is rare entity, we need to be aware of this borderline tumor, as it can transform into mucinous cystadenocarcinoma, and early detection is crucial for appropriate treatment.

### Conflict of interest statement

None declared.

### Funding

None declared.

### Ethical approval

The study was approved by the ethics committee of Yeungnam University College of Medicine. The patient consent waiver was approved by the Institutional Review Board (IRB).

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