

Case Report

Ruptured retroperitoneal mucinous cystadenoma with borderline malignancy could occasionally contribute to death: a rare case report and literature review

Yaojun Dun*, Qing Li*, Tao Xu

*Department of Urology, Peking University People's Hospital, Beijing 100044, China. *Equal contributors.*

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Abstract: We describe a 48-year-old woman with ruptured retroperitoneal mucinous cystadenoma with borderline malignancy resulting in death for postoperative intestinal compression. The patient was referred to our institution for abdominal mass with preoperative ultrasonography and computed tomography (CT) revealing retroperitoneal mass. At operation, the 12×9 cm retroperitoneal tumor had ruptured. Tumor resection was performed and the implants were washed out as completely as possible. The pathological findings showed retroperitoneal mucinous cystadenoma with borderline malignancy. Unfortunately, intestinal compression occurred to the patient on 11 days post-operation. She then underwent laparotomy and colon transversum-terminal ileum anastomosis. However, the condition did not improve and she progressed into septic shock and multiple organ dysfunction soon and died finally. Primary retroperitoneal mucinous cystic tumor (PRMC) is a rare tumor with an obvious predominance for females. Primary diagnosis could be made through preoperative imaging modalities including ultrasonography, CT and magnetic resonance imaging (MRI). Total tumor resection was performed in all previously reported cases. In addition, a total abdominal hysterectomy (TAH) with bilateral salpingo-oophorectomy (BSO) and postoperative adjuvant chemotherapy may make sense in some cases. The overall prognosis of PRMC was good. Among all cases with PRMC, our case was the sixth fatal case, and it was also the seventh case in which the tumor capsule had been injured during operation. The death in perioperative period of PRMC which was presented in our case had never been reported in recent literature. We could conclude that the rupture or not of the tumor during operation was an important prognosis factor for PRMC.

Keywords: Retroperitoneal, mucinous cystic tumor, primary retroperitoneal mucinous cystic tumor (PRMC), intestinal compression, literature review

Primary retroperitoneal mucinous cystic tumor (PRMC) is a rare tumor, with only about 90 cases reported in world literature. And little is known concerning its pathogenesis, optimal treatment and prognosis. We describe a case of ruptured retroperitoneal mucinous cystadenoma with borderline malignancy resulting in death for postoperative intestinal compression ultimately, which had never been reported in the recent literature.

Case report

A 48-year-old woman was referred to our institution with a palpable mass in right upper quadrant region for 48 days. She denied any systemic disease or history of drug abuse. Her past

history included one cesarean section and a right salpingectomy for right fallopian pregnancy 19 years ago. Physical examination showed no remarkable findings except for a large mass which was palpable at her right epigastrium. The lesion had first been detected 1 month prior to admission through ultrasonography at local hospital, which showed a hypoechoic mass with distinct and regular margins. Then she underwent abdominal contrast-enhanced computed tomography (CT) to better evaluate the mass, which revealed a 10×8 cm cystic-solid right retroperitoneal mass with wall patchy or strip calcification. The surrounding organs were compressed and dislocated but not infiltrated (**Figure 1**). The laboratory data were within reference ranges. And tumor markers includ-

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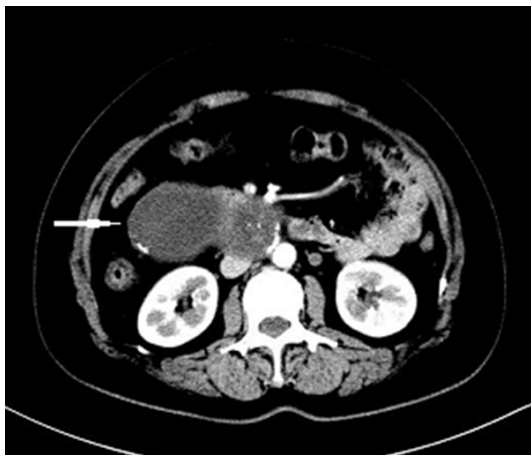


Figure 1. Abdominal computed tomography shows a 10×8 cm cystic-solid lesion (arrow) with wall calcifications in the right retroperitoneal space with mild displacement of surrounding organs.

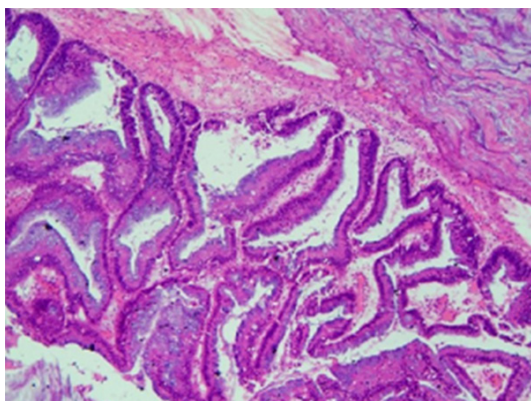


Figure 2. Microscopic examination shows the cyst wall lined with mucinous columnar epithelium (hematoxylin and eosin staining) (×100).

ing CA19-9, CA125 and CEA were within normal limits too.

The patient underwent a surgery to remove the retroperitoneal mass. Because the tumor was close adherent to duodenum and other surrounding structures, the operation was very difficult and time-consuming. Unfortunately, the tumor capsule had been injured during operation, which resulted in tumor rupturing, with mucinous peritoneal implants present nearby. Peritoneal implants were removed and washed out as carefully as possible.

Macroscopically, the tumor was a well-defined mass of 12×9 cm filled with mucin, the wall of



Figure 3. The abdominal computed tomography demonstrates adhesive intestinal compression.

which showed thickening fibrous tissue. And no visible papillary nodules were found. Histologically (**Figure 2**), the cyst was lined by columnar epithelium with mild atypia in some regions. The immunochemistry staining showed CK7 (part +), CK20 (+), CK19 (+), ER (-), PR (-), WT1 (-), CDX-2 (+), TTF-1 (-). A pathologic diagnosis of mucinous cystadenoma with borderline malignancy was made, mixed with mucinous cystadenocarcinoma in part of regions.

The early postoperative course was uneventful, and the patient gradually returned to normal diet with the stitches taken out and the drainage tube pulled-out. However, the patient presented as abdominal distention with nausea and vomiting on 11 days post-operation. A plain x-ray film of the abdomen showed multiple gas-liquid planes.

Although active conservative therapy including gastrointestinal decompression, acid suppression and fluid infusion were accepted, the condition did not improve. Progressive abdominal pain and fever occurred to the patient three days later. And physical examination showed rapid pulse and peritoneal irritation sign with the abdominal CT showing adhesive intestinal compression (**Figure 3**). Thus, the patient was taken to the operating room again to receive laparotomy and colon transversum-terminal ileum anastomosis. Nevertheless, the second surgery did not improve her condition. Septic shock and multiple organ dysfunction was presented on the patient gradually. She died on 30 days post-admission ultimately.

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Table 1A. The treatment and death cause of six patients who died of PRMC

	Age (y)	Sex	Type	Treatment	Follow-up time (mo)	Death cause
Tykkka et al [8], 1975	23	F	malignant	TR	11	Relapse
Gotoh et al [1], 1992	44	F	malignant	TR+Ct	4	Relapse
Mikami et al [9], 2003	38	F	malignant	TR+TAH+BSO+Ct	18	Relapse
Tjalma et al [10], 2008	74	F	malignant	TR+tamoxifen	31	Metastasis
Kamiyama et al [11], 2015	62	F	malignant	TR	15	Relapse
Presented case	48	F	borderline	TR	1	Intestinal compression

Table 1B. The treatment and prognosis of seven patients with PRMC which had ruptured during operation

	Age (y)	Sex	Type	Treatment	Follow-up time (mo)	Prognosis
Tykkka et al [8], 1975	23	F	malignant	TR	11	Death
Tenti et al [12], 1994	46	F	malignant	TR+TAH+BSO+Ct	33	NED
Uematsu et al [7], 2000	86	F	malignant	TR	72	NED
Mikami et al [9], 2003	38	F	malignant	TR+TAH+BSO+Ct	18	Death
Nasir et al [13], 2009	58	F	borderline	TR	12	NED
Williams et al [14], 2010	51	M	malignant	TR+Ct	30	Relapse*
Presented case	48	F	borderline	TR	1	Death [‡]

TR: tumor resection; TAH: total abdominal hysterectomy; BSO: bilateral salpingo-oophorectomy; Ct: chemotherapy; mo: month; NED: no evidence of disease; *The relapse occurred 18 months post-operation. [‡]The patient die of intestinal compression.

Discussion

PRMC is a rare tumor with only 93 cases reported in world literature between 1970 and 2015. There were 74 females and 19 males (female/male ratio = 5.6:1) with an average age of 43 years (range 14 to 90 years). And the size of the tumors ranged from 3.0 to 35.0 cm (mean size 15.3 cm). Abdominal pain, abdominal distention, abdominal mass and abdominal discomfort were the most common symptoms in reported literatures. PRMC could be divided into three categories: primary retroperitoneal mucinous cystadenoma (benign PRMC), primary retroperitoneal mucinous cystadenoma with borderline malignancy (borderline PRMC) and primary retroperitoneal mucinous cystadenocarcinoma (malignant PRMC).

Although the widespread use of imaging modalities including ultrasonography, CT and magnetic resonance imaging (MRI) has increased the detection rate of PRMC, the extreme rarity of similar lesions in the retroperitoneum results in an underdiagnosis of PRMC. And the origin of PRMC is uncertain. Firstly, some authors hold

that PRMC arise in heterotopic ovarian tissue [1]. This theory is largely based on the histologic similarities between ovarian and primary retroperitoneal mucinous tumors. Notwithstanding, in no case has ovarian tissue been found in the tumor. There are several other hypotheses regarding the origin and histogenesis of PRMC: (1) Some authors postulate that PRMC may originate from a primary retroperitoneal teratoma. This may hold true for some cases [2]. (2) Enterogenous duplication cyst as the real site of origin is suggested by others [3]. (3) The hypothesis that the tumor arises from mesothelial invaginations that undergo mucinous metaplasia to become a mucinous tumor is accepted most widely and supported by ultrastructural [4] and immunohistochemical findings [5].

Ultrasonography, CT and MRI are the main methods to diagnose PRMC preoperatively. However, it may be difficult to determine the organ in which the tumor arises and to differentiate a benign from a malignant type through these imaging modalities. In addition, some patients [6] also underwent percutaneous fine-

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needle-aspiration biopsy, but little intracystic content could be aspirated. Thus, the author did not recommend the test because we could not judge the tumor type through aspiration cytology and it may increase the incidence of tumor dissemination.

As for treatment, total excision of the tumor has been performed in all previously reported cases. And a total abdominal hysterectomy (TAH) with bilateral salpingo-oophorectomy (BSO) was recommended by some authors [7] to prevent relapse and metastasis of the tumor, which had drawn lessons from treatment principles of ovarian mucinous cystic tumors. Aside from surgery, postoperative adjuvant chemotherapy may make sense in some reports.

Among 93 cases with PRMC, the follow-up data was available for 66 patients (71.0%). The overall prognosis of PRMC was good during a median of 15 months' (range 2 to 79 months') follow-up. No evidence of disease was presented in 100%, 84.6%, and 71.4% of benign, borderline and malignant PRMC in post-operation course, respectively.

The presented case was the sixth fatal case. The previously reported five fatal cases were all died from relapse or metastasis of the tumor (**Table 1A**). Only our case was the first case died in perioperative period of PRMC. And it was also the seventh case in which the tumor capsule had been injured during operation (**Table 1B**). In our case, the patient died of postoperative intestinal compression. We speculated that the intracystic mucin disseminating into abdomen cavity resulted in adhesive intestinal compression.

We could conclude from **Table 1B** that the rupture or not of the tumor during operation was an important prognosis factor for PRMC. Therefore, the avoidance of tumor rupture during operation could not be more important which may prevent the patient from tumor relapse or distant metastasis and death to some extent. And secondary adhesive intestinal compression could also be avoided if the tumor could be resected as intact as possible.

Disclosure of conflict of interest

None.

Address correspondence to: Tao Xu, Department of Urology, Peking University People's Hospital, Beijing 100044, China. E-mail: xutao@medmail.com.cn

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