

A CASE OF PRIMARY RETROPERITONEAL MUCINOUS CYSTADENOCARCINOMA TREATED WITH FERTILITY-SPARING SURGERY

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Primary retroperitoneal mucinous cystadenocarcinoma is an extremely rare tumor. Preoperative diagnosis is very difficult and the treatment remains controversial. A 37-year-old Korean woman (gravida 0) presented with a huge abdominal mass. Computed tomography scan revealed an 18×11 cm sized unilocular cyst with irregular wall thickening and solid component at right adnexa. Serum CA 19-9 was slightly elevated (37.05 U/mL). At laparotomy, a huge right retroperitoneal cystic tumor originating from right paracolic gutter was found. Frozen section of the cystic tumor revealed a mucinous cystadenocarcinoma. Because the patient wished to remain fertile, fertility sparing surgery was performed. Microscopically, no evidence of metastasis was found and no further treatment was given. Six months after surgery, she has no evidence of recurrence. Fertility-sparing surgery should be considered for women with primary retroperitoneal mucinous cystadenocarcinoma, who wish to remain fertile.

Keywords: Cystadenocarcinoma, mucinous; Fertility; Retroperitoneal neoplasms

Primary retroperitoneal mucinous cystadenocarcinoma is an extremely rare tumor. To date, about 50 cases have been reported [1]. The histogenesis of this unusual neoplasm is not clear. In the majority of cases, preoperative diagnosis is not possible because radiologic study such as ultrasonography or computed tomography (CT) scan is not able to distinguish the exact origin of the lesion. Due to its rarity, the treatment of primary retroperitoneal mucinous cystadenocarcinoma remains controversial. Most of these cases were treated with radical surgery, including total hysterectomy and bilateral salpingo-oophorectomy with enucleation of retroperitoneal tumor [2]. But, fertility-sparing surgery also have been adopted for the treatment of primary retroperitoneal mucinous cystadenocarcinoma and showed good prognosis [1,2]. Here, we report on a case of primary retroperitoneal mucinous cystadenocarcinoma treated with fertility-sparing surgery.

Case Report

A 37-year-old Korean woman (gravida 0) presented with a huge abdominal mass. The patient reported that she had had abdominal discomfort since 1 month ago and visited local clinic. On abdomi-

nal ultrasonography, a huge abdominal mass was detected and she was referred to department of gynecology. Her previous menstrual history was regular and past medical history was noncontributory. On physical examination, her abdomen was markedly distended. CT scan revealed an 18×21 cm sized unilocular cyst with irregular wall thickening and solid component at right adnexa (Fig. 1). Neither enlarged regional lymph node nor ascites was found. Hydronephrosis of right kidney and multiple uterine myomas were found. Laboratory analyses showed normal blood counts and nor-

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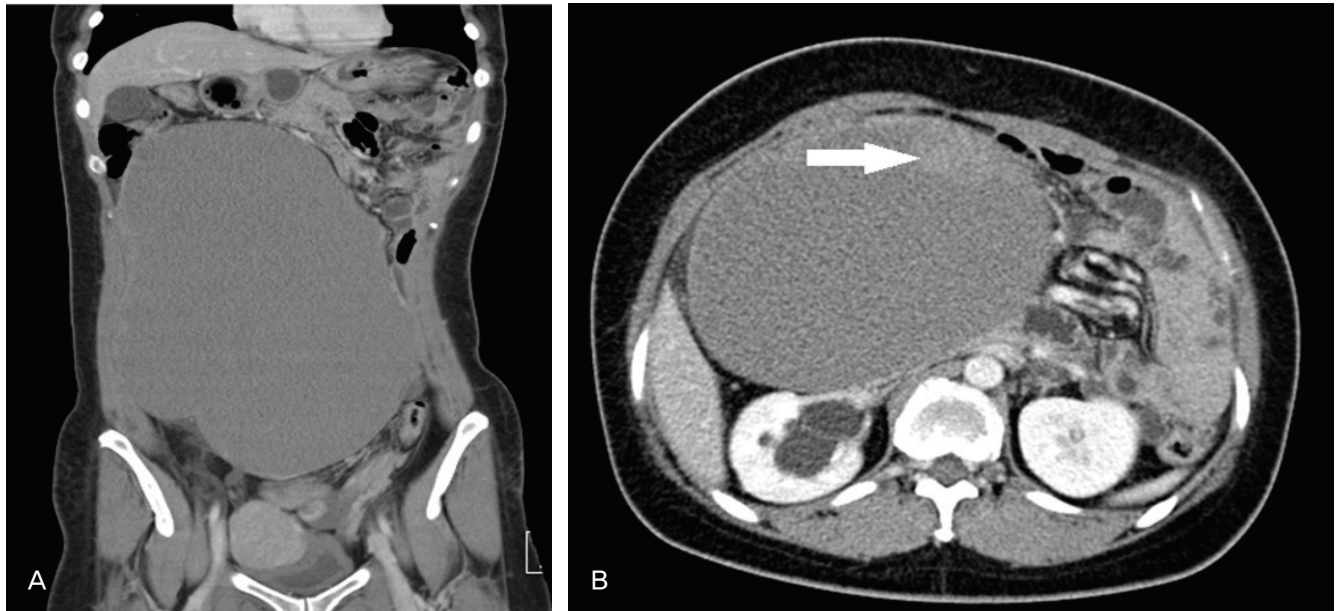


Fig. 1. Computed tomography scan. (A) Unilocular cyst with irregular wall thickening. (B) Solid component (arrow) in the cyst and hydronephrosis of right kidney.

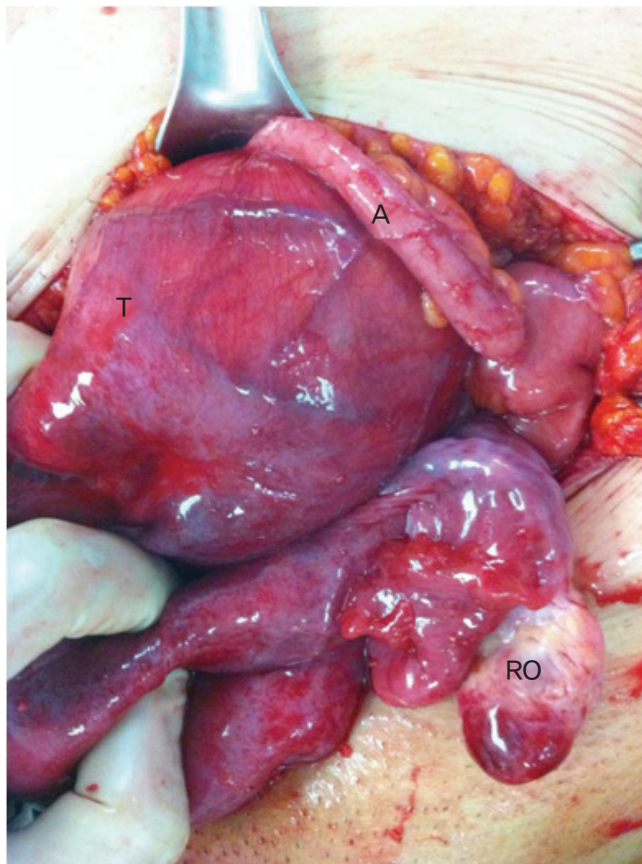


Fig. 2. Intraoperative photo showing the association of the tumor (T), appendix (A), and right ovary (RO). There was no direct connection between them.

mal serum value of CA-125. But, slightly elevated level of serum CA 19-9 was found (37.05 U/mL). Gastroscopy and colonoscopy showed no abnormality.

So, right ovarian neoplasm was suspected and an exploratory laparotomy was performed. A huge right retroperitoneal cystic tumor originating from right paracolic gutter was found. Although the tumor was close to the right ovary and appendix, there was no direct connection between them (Fig. 2). Small nodule was located in the right ovary and multiple uterine myomas were found. Left ovary and fallopian tube appeared normal. The cyst wall, having no apparent connection with any organs and showing no evidence of abdominal spread, was completely excised. Frozen section of the cystic tumor revealed a mucinous cystadenocarcinoma. Right salpingo-oophorectomy, washing cytology, myomectomy, lymphadenectomy, infracolic omentectomy, appendectomy and double J catheter insertion into right ureter were performed. The uterus and left ovary were preserved because the patient wished to remain fertile. Microscopically, no evidence of metastasis was found and no extracapsular invasion or vascular invasion was seen.

Following the surgery the patient recovered without any complication. She has been given no further treatment and 6 months after surgery, she has no evidence of recurrence.

1. Pathologic findings

Grossly, the mass was a well demarcated, unilocular, large cys-



Fig. 3. Gross photography. The tumor is a cystic mass with multiple mural nodules.

tic mass, measuring 19.0×16.0×9.0 cm. It was enveloped by a thick fibrous capsule and contained mucinous fluid. Inner surface showed variable-sized, grayish white, soft mural nodules (Fig. 3). Microscopically, the tumor consisted of glands and papillary structures of columnar mucinous cells (Fig. 4A). The grade of cellular atypia was variable from gland-looking benign mucinous tumor area (Fig. 4B) to glands showing micropapillary architecture, nuclear stratification and atypia, compatible with borderline mucinous tumor (Fig. 4C) and glands of back-to-back arrangement, occasional cribriform pattern and marked nuclear atypia, compatible with well differentiated mucinous adenocarcinoma (Fig. 4D). By immunohistochemistry, the tumor cells were positive for cytokeratin 7, and focally positive for cytokeratin 20 and carcino-embryonic antigen. The Ki-67 index was up to 30%. Considering

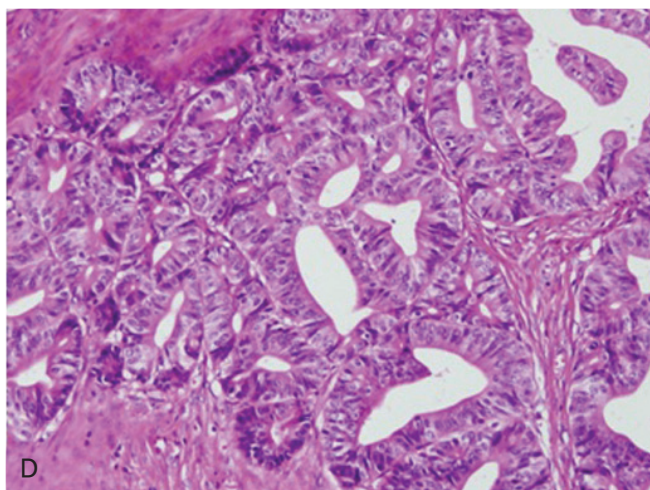
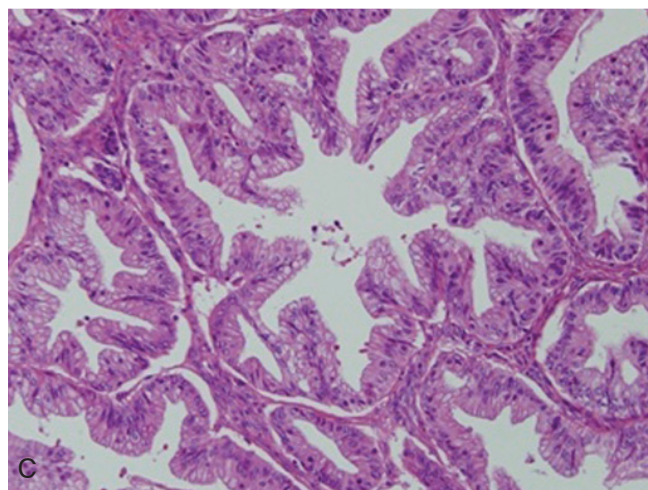
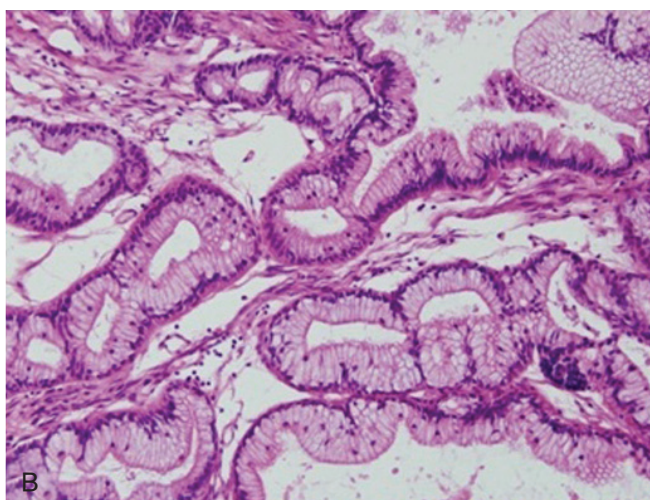
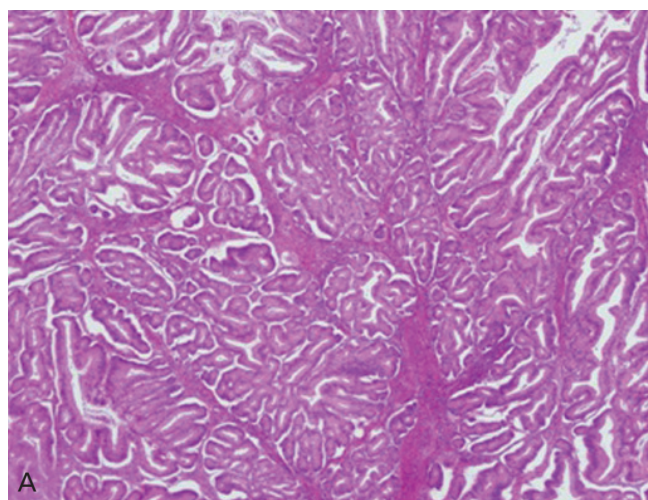


Fig. 4. Microscopic photography. (A) Low power view shows glands and papillae of mucinous cells (H&E, ×40). (B) Benign-looking area (H&E, ×200). (C) Borderline mucinous tumor-like area. Glands show micropapillary architecture and nuclear stratification (H&E, ×200). (D) Adenocarcinoma area. Glands are tightly arranged and show marked nuclear atypia (H&E, ×200).

the clinical feature of the retroperitoneum-confined single mass and the pathologic findings of transition from benign to malignant mucinous tumor, it was diagnosed as a primary retroperitoneal mucinous cystadenocarcinoma.

Discussion

Retroperitoneal tumors of epithelial origin are extremely rare, because no epithelial cells are found in this area. Similar to mucinous tumors of the ovary, these neoplasms are divided into three categories: mucinous cystadenomas, mucinous borderline tumors (tumors of low malignant potential), and mucinous cystadenocarcinomas. Primary retroperitoneal mucinous cystadenocarcinoma was first reported by Roth and Ehrlich [3], in 1977. Although the origin of retroperitoneal mucinous cystadenocarcinomas is not clearly understood, proposed hypotheses include 1) origin from ectopic ovarian tissue [3,4], 2) retroperitoneal primary monodermal teratoma originating from displaced germ cells [5], 3) intestinal duplication, also known as enterogenous genesis [6,7], and 4) coelomic metaplasia [8,9]. To date, the hypothesis that has gained increasing support is coelomic metaplasia, that is, retroperitoneal mucinous cystadenocarcinomas arise from invagination of the peritoneal mesothelium, with subsequent mucinous metaplasia. The ultrastructural findings and immunohistochemical observations support this hypothesis [10].

The age at diagnosis ranges from 17 to 86 years old and the most common complaint at presentation has been abdominal discomfort and a slow-growing pelvic or abdominal mass [7,11]. Ovarian neoplasm rather than retroperitoneal neoplasm was suspected in our patient. Usually, preoperative diagnosis of retroperitoneal neoplasm is difficult because of the non-specific symptoms and the scarce aid of imaging examinations. Although radiologic studies such as ultrasonography and CT scan clearly detect cystic masses in ovary or pelvic organs, diagnosis of retroperitoneal tumor is extremely difficult [7].

CA 19-9 was slightly elevated in our patient, but tumor markers are not very helpful in differentiating the exact origin of the lesion, because CA-125, CA 19-9 may or may not be elevated. Tumor markers help in detecting a recurrent tumor, as in ovarian neoplasm [11].

The treatment of primary retroperitoneal mucinous cystadenocarcinoma remains controversial and no evidence based management guidelines are available. Laparotomy and complete tumor excision should be the principal modality of treatment, but the question

about the extent of the surgery still remains. Most authors treated it as ovarian mucinous cystadenocarcinoma with a standard staging procedure including extirpation of the tumor, total hysterectomy and bilateral salpingo-oophorectomy with or without lymphadenectomy [10]. Most of the follow-up results were excellent. For young ovarian cancer patients who desire a baby, more conservative surgery with preservation of the uterus or ovary may be feasible in a properly selected patient population. According to the last American College of Obstetrics and Gynecology and European Society for Medical Oncology guidelines, fertility-sparing surgery for young women with invasive epithelial ovarian cancer can be adopted for stage IA and non-clear cell histology grade 1 or 2 [12]. Law et al. [2] recommended conservative management should be offered for women with a primary retroperitoneal mucinous cystadenocarcinoma, who desire a baby. They treated a 35-year-old woman by excision of the tumor alone. The patient conceived spontaneously 10 months after initial surgery and had no recurrence 5 years postsurgery. Because our patient also wished to remain fertile and had no evidence of abdominal spread, we performed fertility sparing surgery.

Currently, there is no clear evidence showing the benefit of adjuvant chemotherapy to primary retroperitoneal mucinous cystadenocarcinoma patients. Adjuvant chemotherapy is beneficial when invasion to adjacent structure is evident. Another point in favor of adjuvant treatment is evidence that primary retroperitoneal mucinous cystadenocarcinomas and ovarian mucinous tumor have similar mechanisms in their histogenesis [10]. Patients with this neoplasm generally have a good prognosis after complete removal of the neoplasm [13]. In our patient, the tumor was completely excised and no evidence of metastasis was found. So, we did not perform any adjuvant therapy.

Primary retroperitoneal mucinous cystadenocarcinoma is an extremely rare tumor and the treatment remains controversial. Fertility-sparing surgery should be considered for women with this neoplasm, who wish to remain fertile.

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가임력 보존수술로 치료된 원발성 후복막 점액성 선암 1예

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원발성 후복막 점액성 선암은 극히 드문 종양이다. 수술 전 진단이 매우 어렵고 치료방법은 정립되지 않았다. 임신력이 없는 37세 여성이 거대한 복강내 종양이 발견되어 내원하였다. 컴퓨터단층촬영에서 18×11 cm 크기의 불규칙한 두께의 낭벽과 고형 성분을 가지는 단방성의 낭종이 우측 부속기 부근에서 발견되었다. 혈청 CA 19-9 농도는 37.05 U/mL로 약간 증가하였다. 시험적 개복술을 시행하였고, 우측 결장 옆 복벽에서 기원하는 거대한 후복막 낭성 종양이 발견되었다. 동결절편검사에서 점액성 선암으로 판명되었다. 환자가 가임력 보존을 원하였기 때문에 자궁과 좌측 부속기를 보존하는 가임력 보존수술을 시행하였다. 병리조직학적 검사에서 전이는 발견되지 않았고 보조적 항암화학요법은 시행하지 않았다. 수술 후 6개월까지 추적 관찰하였는데, 재발은 없었다. 가임력 보존을 원하는 원발성 후복막 점액성 선암 환자에서 가임력 보존수술이 고려되어야 한다.

중심단어: 점액성 선암, 가임력, 후복막 신생물