Management of a primary retroperitoneal mucinous cystadenocarcinoma: case report

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Summary

Purpose: To review the treatment of primary retroperitoneal mucinous cystadenocarcinoma (PRMC). *Case report:* A 30-year-old woman had a large retroperitoneal mucinous adenocarcinoma treated with conservative laparoscopic surgery. Two years later, she was found to have bilateral ovarian cysts at the time of cesarean section. Since cystectomies revealed mucinous adenocarcinoma, she underwent complete surgical staging and adjuvant chemotherapy at this time. *Conclusion:* A rare case of similar cancer in the ovary following treatment for PRMC was described. It is unclear whether the prognosis is improved by oophorectomy. Further cases and long-term follow-up are necessary.

Key words: PRMC; Ovarian cancer; Laparoscopy; Oophorectomy.

Introduction

Primary retroperitoneal mucinous cystadenocarcinoma (PRMC) is a rare tumor, the pathogenesis of which remains unclear and controversial. Most authors support the theory of coelomic metaplasia [1-3]. Supernumerary ovary or heterotopic ovarian tissue has also been considered a possible source of retroperitoneal mucinous cystic tumors [4, 5]. Another possibility is that this may be an outgrowth of mucinous epithelium over other components from a primary teratoma of the retroperitoneum [6]. Some authors have also mentioned the possibility of enterogenic duplication cyst as the origin [7].

Recently, coelomic metaplasia is being considered as a possibility for ovarian epithelial neoplasms. The process is just like the neoplastic development in the primary Müllerian system [7-9]. Therefore, some patients have undergone not only tumor resection, but also hysterectomy and bilateral salpingo-oophorectomy [1, 4, 8], notwithstanding the fact that in all these cases, the resected genital organs showed no evidence of involvement or tumor infiltration. To improve clinical outcomes, oophorectomy is advocated [1].

Laparoscopy has become the standard approach for the treatment of female patients with an abdominal mass. On the other hand, some problems may occur when tumors provisionally diagnosed as benign preoperatively turn out to be malignant.

A case of similar cancer that occurred in the ovary following treatment for PRMC is presented. A laparoscopic procedure was done for PRMC. On the basis of a review of the literature, management of such cases is discussed.

Case Report

A 30-year-old woman, gravida 1, para 1, was referred to the present hospital for right abdominal pain. On physical examination, an 18-cm mass was found in the right lower abdomen. The mass showed a smooth surface, cystic to elastic hard. On magnetic resonance imaging (MRI), a large, 19×14×8 cm, non-enhanced, cystic mass was seen (Figure 1a). The uterus and both ovaries were normal in shape and size. The tumor had no connection to gynecological organs. The levels of serum tumor markers CA 19-9, carcinoembryonic antigen, and CA 125 were within normal limits. A serous tumor of peritoneal or mesenteric origin was suspected. A laparoscopic procedure was done. A large cystic mass arising from the retroperitoneum at the right iliac fossa was found displacing the ascending colon to the left side (Figures 1b, c). After the serous fluid was aspirated by a sand-balloon tube, the tumor was resected from the retroperitoneal space. At the first surgery, an appendectomy was not performed because the tumor appeared serous. Microscopically, the wall of the cystic lesion was lined by mucinous columnar epithelium showing mild to moderate nuclear atypia, papillary projections, and mild stratification. A focal, apparently stromal lesion was found (Figure 2a). Its features were consistent with mucinous adenocarcinoma. Immunohistochemically, neoplastic cells showed positive staining for cytokeratin, EMA, and CEA and negative staining for estrogen and progesterone receptors. In part of the lesion, mucinous metaplasia was adjacent to the mesothelium (Figure 2b). Negative staining for CEA was seen in the benign single layer of mucinous epithelium. On the other hand, positive staining for CEA was seen in the area of malignant neoplastic cells (Figure 2c). These histological and immunohistochemical findings confirmed that the tumor was mucinous adenocarcinoma that had originated from the retroperitoneal mesothelium. To evaluate for a gastrointestinal primary, colonoscopy and positron emission tomography-computed tomography (PET-CT) were performed. All additional examinations were normal. Because no evidence was found to suggest additional treatments, she was followed using ultrasound and tumor markers (CEA, CA19-9, and CA 125) every month before pregnancy. Afterwards, no abnormalities were seen, and she was observed using only ultrasound after pregnancy. Two years later, when a repeat cesarean section was performed, bilateral cys-

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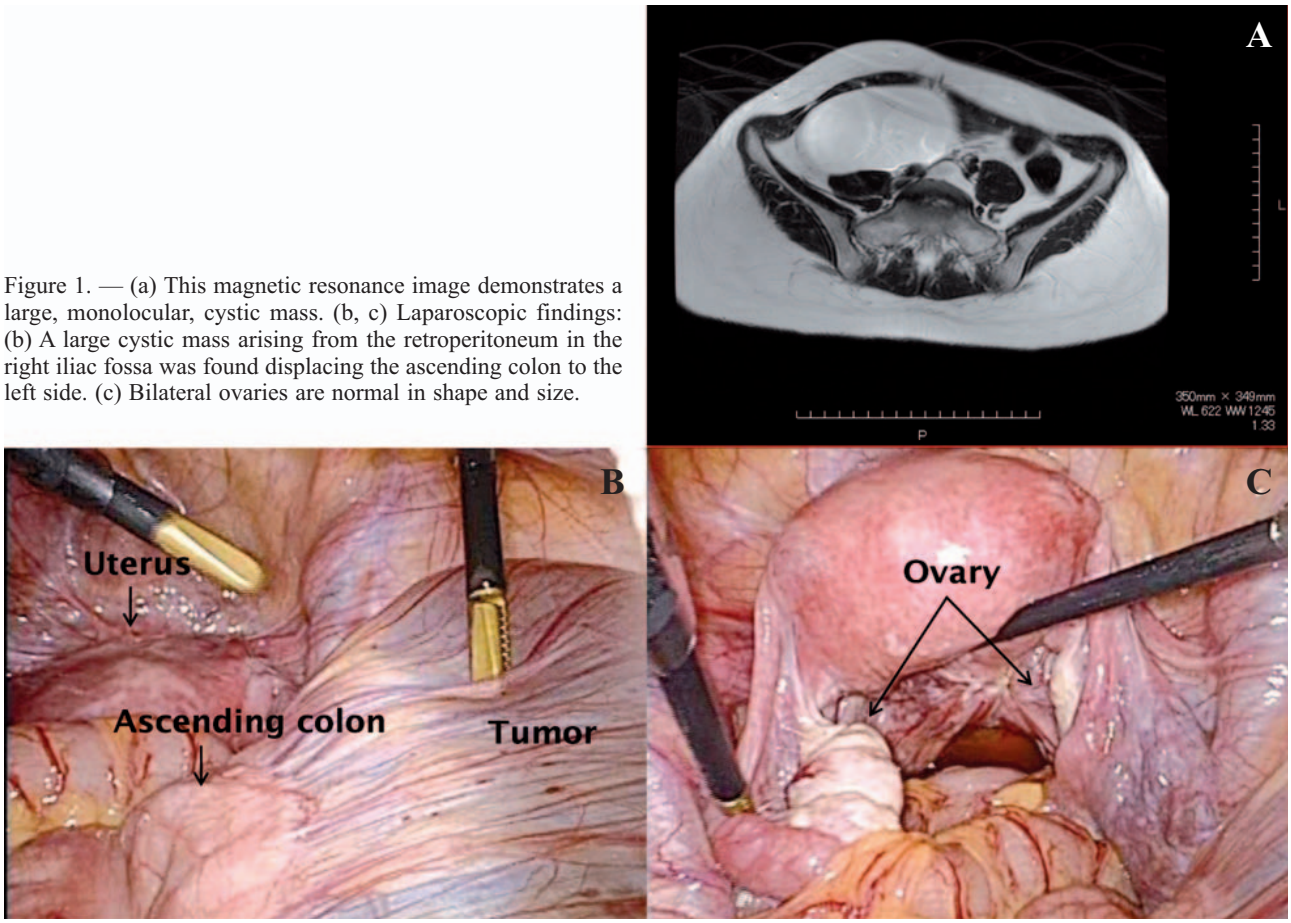

Figure 1. — (a) This magnetic resonance image demonstrates a large, monolocular, cystic mass. (b, c) Laparoscopic findings: (b) A large cystic mass arising from the retroperitoneum in the right iliac fossa was found displacing the ascending colon to the left side. (c) Bilateral ovaries are normal in shape and size.

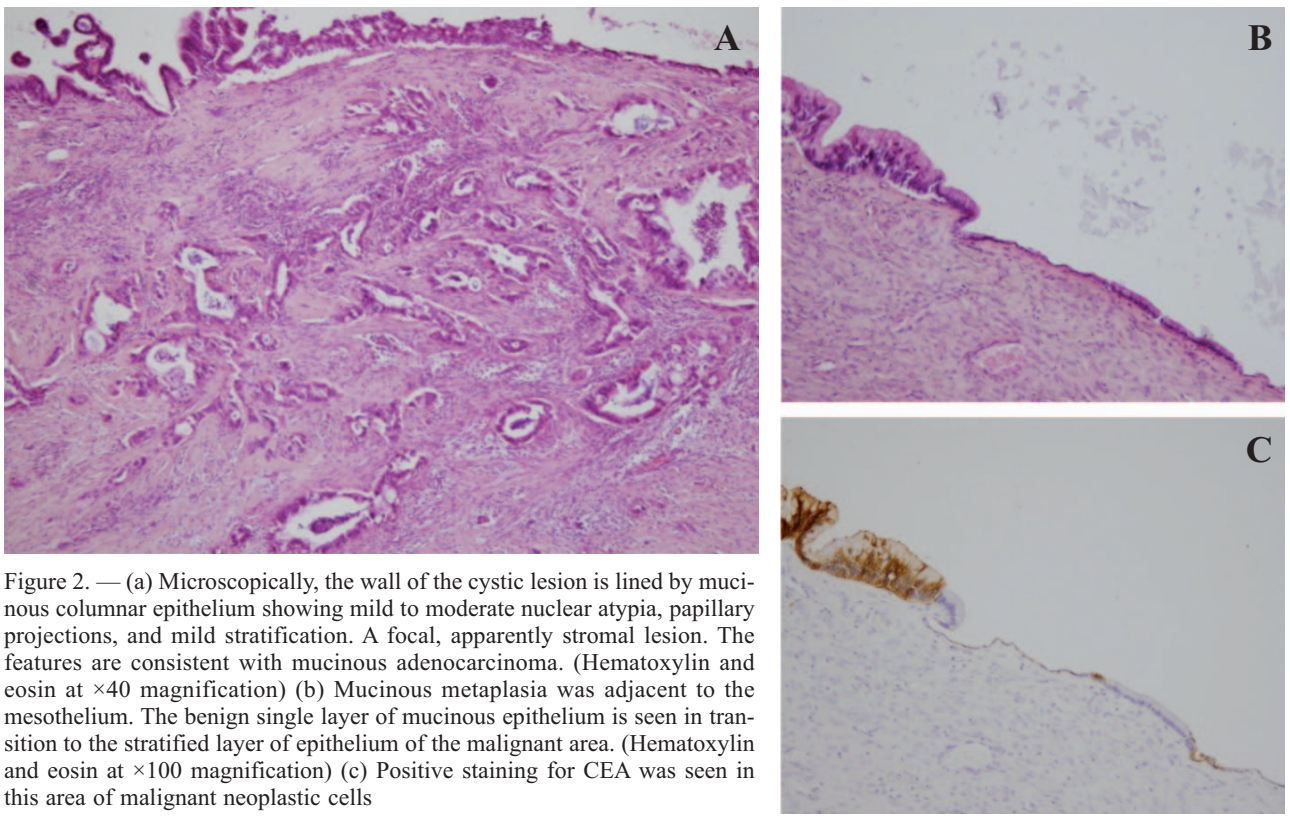

Figure 2. — (a) Microscopically, the wall of the cystic lesion is lined by mucinous columnar epithelium showing mild to moderate nuclear atypia, papillary projections, and mild stratification. A focal, apparently stromal lesion. The features are consistent with mucinous adenocarcinoma. (Hematoxylin and eosin at ×40 magnification) (b) Mucinous metaplasia was adjacent to the mesothelium. The benign single layer of mucinous epithelium is seen in transition to the stratified layer of epithelium of the malignant area. (Hematoxylin and eosin at ×100 magnification) (c) Positive staining for CEA was seen in this area of malignant neoplastic cells

Table 1. — *Primary retroperitoneal mucinous cystadenocarcinoma.*

Author	Year	Sex	Age	Side	Size(cm)	Initial treatment	Adjuvant treatment	Prognosis
Senda	1990	F	42	R	11×8×5	TE	–	–
Kuroda	2006	F	33	L	15	TE	–	DOD
Kawai	2003	F	20	R	7×6	TE	–	NED(18M)
Kaku	2001	F	38	L	16	TE	CAP→Second Surgery→Taxol	DOD(18M)
Tando	2009	F	42	F	12×8×3.5	TE	–	NED(2M)
Matuno	2001	F	39	L	20×18×15	TE	–	NED(24M)
Horiuchi	1991	F	55	L	18×12×10	TE	CA→CPA	NED(19M)
Toyoda	2007	F	72	R	4.5×5.5	TE	–	3M: Rec→DOD
Saikawa	1992	F	50	R	10×17×16	TE	–	NED(4M)
Tamura	1984	F	51	L	1410g	TE	TAH+BSO	–
Thomas M	2002	F	38	L	11×10×10	TE+resection of decending colon+Appe	–	NED(60M)
Roberto D	1996	F	45	L	20×16×15	TE	TAH+BSO+PLND+OMN	NED(16M)
Roth LM	1976	F	48	L	7×7×2	TE		6M: Rec→DOD
Gotoh K	1992	F	44	R	12×10×5	TE	Tegaful-Uracil	4M: Rec→DOD
Song ES	2005	F	72	M	12×12×10	TE	–	2M: Rec→DOD
Suzuki S	2001	F	40	R	12×12×15	TE+resection of ileocecal region+Appe	–	NED(15M)
Tangjitgamol S	2002	F	41	L	12×11	TE+TAH+BSO+Appe+OMT+PALNBx	CDDP+CBDCA	NED(18M)
Uematsu T	2000	F	86	M-L	23×20×12	TE+distal partial gastrectomy with LND	–	NED(72M)
Park U	1991	F	40	R	13×15×24	TE	TAH+BSO+OMBx	NED(3M℃lost)
Lee IW	1998	F	55	L	19×13	TE	TAH+BSO+PLND+PALNBx+OMN	NED(30M)
		F	45	L	17×15×9	TE+TAH+BSO	–	NED(15M)
Carabias E	1995	F	43	R	15	TE	TAH+BSO	NED(24M)
Pearl ML	1996	F	33	L	large	TE	–	NED(10M)
Fujii S	1988	F	69	R	10×15×23	TE+TAH+BSO	–	NED(36M)
Nelson H	1988	F	35	R	20×13	TE+TAH+BSO	–	NED(22M)
Tenti P	1994	F	46	R	20×12	TE+TAH+BSO	Chemo	NED(33M)
		F	45	L	20×16×15	TE+TAH+BSO	–	NED(19M)
Jorgensen LJ	1991	F	38	R	8	TE	–	NED(9M)

R: right
L:left
M:middle

TE: Tumor excision
TAH: total abdominal hysterectomy
BSO: bilateral salpingo-oophorectomy
PLND: pelvic lympho node dissection

PALNBx: para-aortic lympho node biopsy
LND: lympho node dissection
Appe: appendectomy
OMN: omentectomy

tic ovarian tumors were confirmed. The right and left ovaries measured 6 × 4 cm and 4 × 3 cm, respectively. Bilateral cystectomy was done, and the same mucinous adenocarcinoma was diagnosed microscopically. The staging procedure included a total abdominal hysterectomy with bilateral salpingo-oophorectomy, appendectomy, omentectomy, and both pelvic and para-aortic lymphadenectomy. The appendix and the pancreas were grossly normal in size and shape. Remaining cancer was found at both the site of adhesion of the ovary and the pouch of Douglas. Washing cytology was negative. The final diagnosis was Stage IIb with Grade 1 mucinous ovarian cancer. Chemotherapy with six courses of paclitaxel 180 mg/m^2 and carboplatin (AUC = 5) was administered. The patient was doing well without evidence of disease eight months later.

Discussion

Mucinous cystadenocarcinoma is not an uncommon gynecologic malignancy. Mucinous tumors of the ovary are occasionally associated with mucinous tumors at other sites, such as the cervix or appendix. They may sometimes involve two different organs with the same histology, so it is difficult to determine both the primary and metastatic sites. When retroperitoneal mucinous cystadenocarcinoma is assumed to be the result of coelomic metaplasia, the possibility exists that a similar ovarian cancer can occur from a metaplastic change.

Is adjuvant bilateral salpingo-oophorectomy justified at the first laparoscopic operation? Reports regarding the extent of surgical treatment of PRMC were investigated, and these are summarized in Table 1. Many authors advise hysterectomy with bilateral salpingo-oophorectomy [3, 4, 8, 9], either simultaneously or later at re-exploratory laparotomy accompanied by peritoneal washing and lymph node sampling [2-4, 8, 9]. It has been reported that patients who undergo hysterectomy and bilateral salpingo-oophorectomy were free of recurrence [3, 8, 9]. The tumor has been said to be affected by female hormones [4]. However, some of those patients who had metastases in organs other than the ovary developed recurrence and died [4-6]. The question remains as to whether bilateral salpingo-oophorectomy improves the prognosis. Considering conception and ovarian function, sufficient evidence was required to resect ovaries without pathological changes. It is recommended that adjuvant bilateral salpingo-oophorectomy be restricted to women who have completed their child-bearing or are postmenopausal [4].

Choosing between laparoscopy and laparotomy for the treatment of abdominal tumors is difficult. In this case, a laparoscopic procedure was performed because the tumor was preoperatively diagnosed as a benign serous tumor. When serous fluid was aspirated, some of the fluid may

have leaked. A retroperitoneal approach was taken for the retroperitoneal tumor. With the normal left approach, it is difficult to see the right iliac fossa. It was necessary to aspirate the fluid because of the size of the tumor. It has been reported that cyst rupture is more frequent during laparoscopic management, suggesting that laparoscopy may increase the risk of dissemination [10]. However, metastasis of the cancer affected only the ovary. It is hard to ascribe the metastasis in this case to leakage. Moreover, it has been reported that port-site metastasis is linked to unprotected extraction [10]. A recovery bag was used in this case.

In the present case, recurrence was seen two years after the first operation. There is little literature reporting long-term follow-up, but long-term follow-up is required to determine the time of recurrence.

In conclusion, a rare case of a similar cancer that occurred in the ovary following treatment for PRMC was presented. It is unclear whether the prognosis of PRMC is improved by oophorectomy. More cases and long-term follow-up are necessary.

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