Pseudomyxoma peritonei and mucinous pyometral fluid arising from an ovarian borderline mucinous tumor: case report

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Summary

An extremely rare case of a pseudomyxoma peritonei (PMP) and mucinous pyometral fluid, possibly arising from an ovarian borderline mucinous tumor is reported. A 68-year-old Japanese patient received an expolatory laparatomy under a working diagnosis of a PMP, left ovarian cystic tumor and an umbilical hernia. Surgery and platinum-based chemotherapy induced a 15-month disease-free condition.

Key words: Pseudomyxoma peritonei; Ovarian borderline mucinous tumor; Mucinous pyometral fluid; Chemotherapy.

Introduction

Pseudomyxoma peritonei (PMP) is a rare disease with an incidence of only approximately 1/100,000 a year [1]. Ronnett *et al.* have divided PMP into three pathological subtypes: disseminated peritoneal adenomucinosis (DPAM), peritoneal mucinous carcinomatosis (PMCA) and an intermediate group (PMCA-I) [2]. Sometimes, PMCA cases are found to be chemo-resistant and they tend to show a poor prognosis. However, the prognosis of DPAM is thought be much better than that of PMCA [2]. We recently experienced an extremely rare case of a PMP and mucinous pyometral fluid at the same time, possibly arising from an ovarian borderline mucinous tumor. Surgical treatment and platinum-based chemotherapy induced a 15-month disease-free condition.

Case Report

A 68-year-old Japanese female, gravida 3, para 3, noticed lower abdominal distension. In June 2005, she received an exploatory laparotomy under the working diagnosis of a PMP, left ovarian cystic tumor (Figure 1, gray arrow) and an umbilical hernia (Figure 1, black arrow), and pyometra or uterine invasion (Figure 1A, white arrow). At surgery, a yellowish, gelatinous and mucinous fluid (approximately 300 ml, Figure 2A, yellow arrow) was found to be present in the abdominal cavity. The left ovarian cystic tumor had already ruptured, the uterine body was enlarged due to the presence of dark reddish mucinous fluid (Figure 2A, white arrow). She underwent an abdominal hysterectomy, bilateral salpingo-oophorectomy, pelvic lymphadenectomy, and appendectomy with an indwelling of the abdominal port and also repair of the umbilical hernia. A pathological examination showed a left ovarian borderline mucinous cystadenocarcinoma (Figure 2B), metastasis to the appendix and invasion to the uterine serosa. Although the cytological examination from the uterine cavity was positive (Figure 2C), no evidence of tumor cells in serial sections of the endometrium could be detected. No other dissemination or lymph node metastasis could be found in the pelvic or abdominal cavity. From the above findings, the patient was diagnosed as having PMP from a left ovarian borderline mucinous cystadenocarcinoma, with metastasis to the appendix and invasion to the uterine serosa.

From July 2005, the patient received adjuvant chemotherapy with paclitaxel (150 mg/m^2) plus calboplatin (AUC = 5), but her condition later worsened probably due to premedication of cortiosteroids. Thereafter, she received three courses of chemotherapy with systemic cyclophosphamide (350 mg/m^2), terarubicin (30 mg/m^2) and intraperitoneal administration of cisplatin (50 mg/m^2).

After surgery, she received adjuvant chemotherapy and has since been disease-free for 15 months.

Discussion

Regarding chemotherapy for PMP cases, IP administration with platinum is considered to be more effective than systemic administration [3]. The use of IP chemotherapy with platinum proved to be effective for this case, and this case was considered to have either DPAM or PMCA-I type disease.

PMP usually originates from the appendix and sometimes from the ovary [4, 5]. Other primary sites are extremely rare, including the pancreas, stomach, colon, urachus and small bowel. This case showed mucinous fluid, not only in the abdominal cavity but also in the uterine cavity. Although tumor cells were found in the pyometral fluid, no tumor cells were observed in the serial section from the endometrium. We know that the inner diameter is very narrow for tumor cells to pass from the abdominal cavity, however, some pressure by the ileus might enable tumor cells to enter the uterine cavity. As a result, we considered the primary site to be the left ovary in this case.

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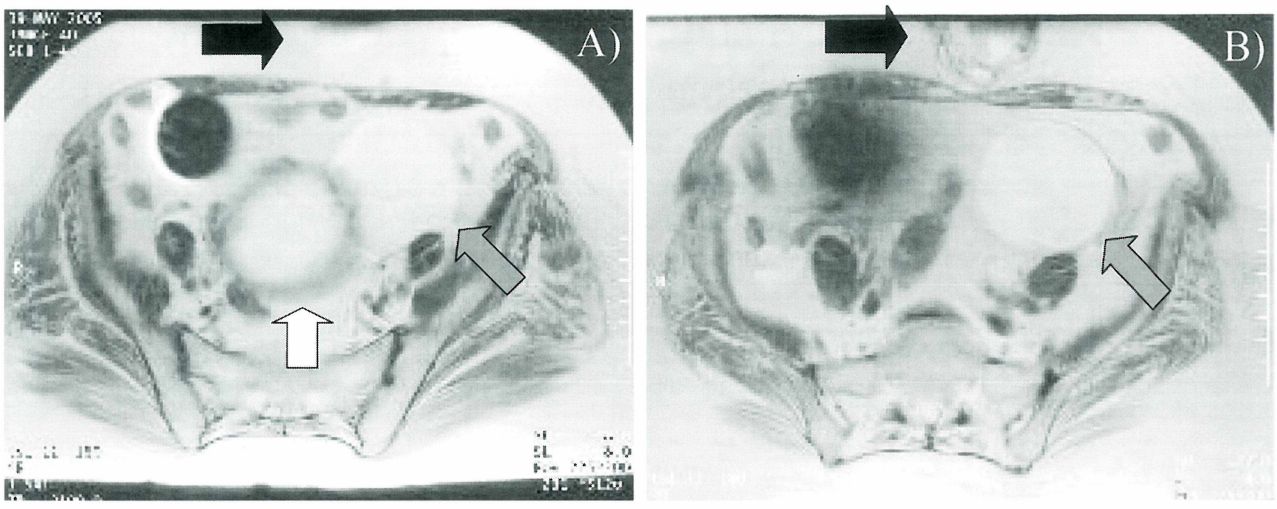

Figure 1. — A) Pelvic MRI T_2 transverse section revealed left ovarian cystic tumor and an umbilical hernia (white arrow). B) Pelvic MRI T_2 sagital section revealed uterine pyometra (grey arrow).

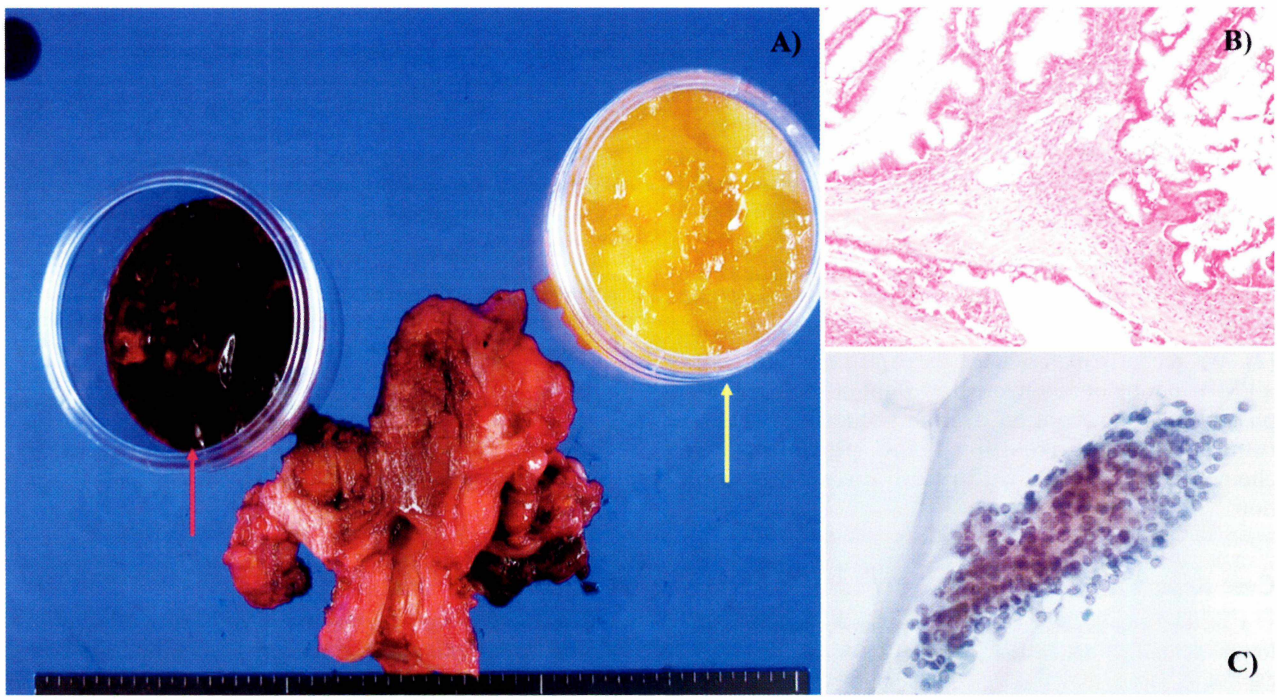

Figure 2. — A) Resected uterus and ovaries. In the uterine cavity, reddish mucinous fluid was present (red arrow). In the abdominal cavity, yellowish mucinous fluid was present. B) Left ovarian mucinous tumor (H&E x 150). C) Cytology from the uterine cavity (Papanicolaou stain x 350).

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