Rare disease

Ovarian cystadenocarcinoma and pseudomyxoma peritonei

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Background

Mucocèles resulting from cystadenomas of the ovary are uncommon. Although rare, rupture of the mucocèles can occur without causing any abdominal issue. This report concerns an unusual and interesting case of the right ovary associated with liver, spleen and kidney cysts.

Case presentation

A case of ruptured mucocèle resulting from cystadenoma of the ovary occurred in a 63-year-old woman. The patient underwent x-ray, ultrasound and a CT scan. Localised pseudomyxoma peritonei associated with adenocarcinoma of the right ovary was diagnosed. The patient underwent resection of the ovary, uterus, uterine tube and appendix with excision of the mesenteric lymph nodes and removal of about 5000 ml of yellowish mucoid jelly. She was free of symptoms at 1-year follow-up.

Conclusions

Pseudomyxoma peritonei may occur in patients with ovarian cystadenocarcinoma. In such patients, the ovary should be investigated and transvaginal ultrasound can be carefully performed in cases of ruptured mucocèles and localised pseudomyxoma peritonei. Surgical intervention is the current choice of management.

BACKGROUND

Pseudomyxoma peritonei represents a special case in metastatic peritoneal tumours, which some reports have shown as resulting from cystadenomas of the appendix.^{1–5} However, cystadenocarcinomas of the ovary are uncommon. Although rare, rupture of the mucocèles can occur without causing any abdominal issue or with only painless abdominal distension. There are few reports associating pseudomyxoma peritonei with cystadenocarcinoma of the ovary.^{6,7} Herein, we report an unusual and interesting case of pseudomyxoma peritonei associated with right ovarian cystadenocarcinoma.

CASE REPORT

A 63-year-old woman presented with progressive abdominal distension for 2 months before admission to the hospital. The patient remained in a good general condition with no signs of progression except for the abnormal distension for 1 year before admission. We palpated a non-tender and persistent diffuse mass. The tumour markers α -fetoprotein (AFP), carcinoembryonic antigen (CEA), cancer antigen (CA)19-9 and CA125 were 10.72 ng/ml, 97.28 ng/ml, 127.3 U/ml and 112.72 U/ml, respectively. Abdominal ultrasound showed a large hypoechoic mass in the lower abdomen and an irregular cystic hyperechoic lesion over the right lower quadrant; the appendix appeared normal (figure 1). Gynaecological ultrasound showed the right ovary was multiloculated and enlarged, containing a lot of pultaceous and mucinous material; the left ovary shape was irregular

with an echoless region. A CT scan revealed considerable abdominal mucinous ascites with multiple cysts in the liver, kidney and spleen (figure 2). A jelly-like material was extract by abdominal paracentesis (figure 3). This suggested pseudomyxoma peritonei originating from the right ovary.

An ovariectomy was performed and the pathology revealed a papillary adenocarcinoma of the right ovary (figure 4). On laparotomy, the abdominal cavity was found to be occupied by a jelly-like material with a location suggestive of a ruptured right ovarian mucocèle; resection of the ovary, uterus, uterine tube and appendix was performed with excision of the mesenteric lymph nodes and removal of a yellowish mucoid jelly (about 5000 ml) (figure 5). Pathology revealed a papillary adenocarcinoma of the right ovary (figure 6A) with regional lymph node metastases and left ovary, greater omentum and appendix invasion (figure 6B). Grossly the right ovary measured 8×6×2.5 cm with a fallopian tube measuring 6 cm long. Externally it was ruptured and the inner wall was filled with a jelly-like material. The appendix measured 2×1×0.5 cm. The left ovary measured 6×4×2 cm. Externally it was well encapsulated. The cut surface showed cystic material. Microscopically, a section of the right ovary showed the presence of mucinous papillary adenocarcinoma. A section of the left ovarian capsule wall showed a monolayer applanate epithelium with no presence of the dysplastic gland. A section of the appendix showed infiltration of the dysplastic mucosal glands in the serosa and muscular layer, which suggested

Figure 1 Abdominal ultrasound showing a large hypoechoic mass in the lower abdomen and an irregular cystic hyperechoic lesion over the right lower quadrant.

Figure 2 A CT scan revealing considerable abdominal ascites with multiple cysts in the liver (A), kidney (D) and spleen (B–D).

Figure 3 Jelly-like material extract by abdominal paracentesis.

Figure 4 Ovariectomy was performed and the pathology revealed papillary adenocarcinoma of the right ovary.

origination from the right ovary mucinous adenocarcinoma. Postoperative chemotherapeutic treatment was performed. The patient's hospital course was uneventful. The patient was free of symptoms at 1-year follow-up.

DISCUSSION

Pseudomyxoma peritonei represents a special case in metastatic peritoneal tumours. Its degree of malignant potential is variable; about 50% of patients live 5 years. The

Figure 5 On laparotomy, resection of the ovary, uterus, uterine tube and appendix was performed with excision of the mesenteric lymph nodes and removal of yellowish mucoid jelly.

Figure 6 Pathology revealed adenocarcinoma of the right ovary (A) with regional lymph node metastases and left ovary and greater omentum invasion (B).

sites of origin of the tumour are the ovaries and appendix, and lymphatic or extraperitoneal spread of the tumour is rare. The pathological entities include retention cysts, mucosal hyperplasia, cystadenoma and cystadenocarcinoma. Presenting symptoms include painless abdominal distension and ovarian mass; mucin may accumulate intraperitoneally many years after resection of an ovarian mass.

Diagnosis is made when a jelly-like material is encountered at laparotomy or laparoscopy. A recent retrospective multicentre study of a clinicopathological analysis of pseudomyxoma peritonei showed that, for preoperative tumour markers, the positive rates were 72% (CA125), 47.4% (CA19-9) and 84.6% (CEA).⁸ The patient's tumour markers (CEA, CA19-9 and CA125) were 97.28 ng/ml, 127.3 U/ml and 112.72 U/ml, respectively, which suggests these could be markers in patients with jelly-like material extracted by abdominal paracentesis. Aggressive surgical debulking with copious irrigation of the peritoneal cavity with warm dextrose solution appears to be the treatment of choice.⁹ Adjuvant radiotherapy and chemotherapy has also been used. Unfortunately, chemotherapy appears to be ineffective, and recurrence usually causes bowel obstruction, malnutrition, and can lead to death. Recently, it was reported that surgery combined with intraperitoneal hyperthermic perfusion has been used to treat patients with pseudomyxoma peritonei.¹⁰ An Italian multicentre phase II study revealed that 5-year overall survival, progression-free survival and locoregional progression-free survival rates were 91%, 54% and 69%, respectively. This study confirms the efficacy of the combined treatment on long-term survival and local disease control.¹¹

CONCLUSIONS

Pseudomyxoma peritonei may occur in patients with ovarian cystadenocarcinoma. In such patients, the ovary should

be investigated and transvaginal ultrasound can be performed carefully in cases of ruptured mucocoeles and localised pseudomyxoma peritonei. Surgical intervention is the current choice of management.

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Competing interests None.

Patient consent Obtained.

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