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Pseudomyxoma Peritonei - A Case Report

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ABSTRACT

A 53-year-old lady presented with post-menopausal bleeding (spotting) for two days and abdominal pain for three months. CT scan revealed bilateral adnexal mass and ascites; and CEA and CA19.9 was raised. Histopathology of the resected tissues revealed mucinous carcinoma of bilateral ovaries and colon with omental metastasis. Generally, pseudomyxoma peritonei arises from primary ovarian or appendiceal adenomas or adenocarcinomas.

Keywords: Adenocarcinomas; ascites; metastases; mucinous carcinoma; pseudomyxoma peritonei

INTRODUCTION

Pseudomyxoma peritonei is a rare neoplasm with an incidence of 1-2 per million per year.[1] It is a low grade malignancy originating from cancerous or non-cancerous tumors of appendix and/or ovary characterized by diffuse tumor deposits in the peritoneal surfaces and voluminous collection of mucin in the peritoneal cavity forming "jelly belly". It is difficult to diagnose pre-operatively and has a higher recurrence rate.

CASE REPORT

A 53-year-old lady presented to the outpatient department with history of post-menopausal bleeding (spotting) for two days associated with abdominal pain and abdominal distension for three months. She gave a history of dull aching, central pain that was not radiating. On abdominal examination, it was tense, distended, non tender and an irregular, hard, fixed mass was palpable on the left lower abdomen with gross ascites. Under speculum, there was blood-mixed discharge in the vagina. On bimanual examination per vaginum, a similar mass of size around 8x8cm was palpable at the left adnexal region whereas the right adnexa was free. CT examination of her abdomen and pelvis revealed:

Complex cystic lesion with fine enhancing septations in left adnexal region measuring 10.7x9 cm and similar lesion of size 10.6x5.6 cm in right adnexal region.

Bilateral ovaries were not separately visualized.

Gross ascites was seen.

Omental thickening and nodules were seen including deposits along ascending colon/caecum likely of bilateral ovarian malignancy with peritoneal/omental metastases.

Her serum markers CA-125, beta human chorionic gonadotropin and lactate dehydrogenase were normal. However, her carcinoembryonic antigen (CEA) was 82.7ng/ml and CA-19.9 was 159.0 U/ml. The ascitic fluid was aspirated for cytology and reported to be negative for malignancy. Endometrial biopsy was done for post-menopausal bleeding which showed proliferative phase endometrium. Under clinical grounds, she was diagnosed with bilateral ovarian masses and an exploratory laparotomy was performed. On opening the abdomen, around five litres of gelatinous material was found throughout the abdominal cavity (Figure 1).

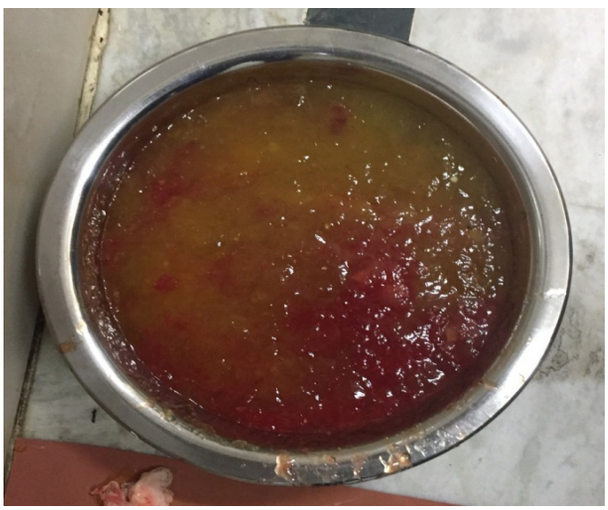

Figure 1. Gelatinous peritoneal fluid removed during laparotomy.

A ruptured mass measuring around 10x10 cm in pelvis, densely adherent to uterus and inseparable from bilateral ovaries was found to be oozing the gelatinous

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material (Figure 2). The mass was in the pouch of Douglas and is a conglomerate mass arising from both ovaries. Another irregular solid mass, measuring 6x5 cm was seen involving the appendix, caecum and proximal part of ascending colon adherent to above described pelvic mass. The omentum was nodular and was adherent to the colonic mass. Multiple nodular deposits were present on the paracolic gutters and bladder base. The uterus with bilateral tubes and ovaries were removed with supracolic omentectomy and limited right hemicolectomy (Figure 3). The abdominal cavity was washed out and was closed in layers. Post-operatively, she made an uneventful recovery.

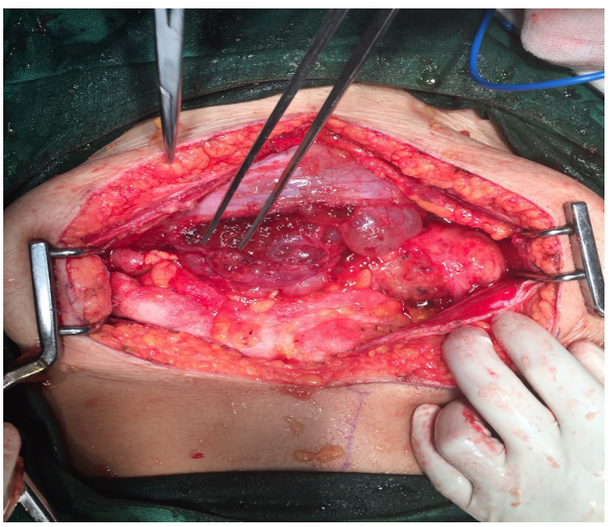

Figure 2. Ruptured ovarian mass in pelvis with mucinous spillage.

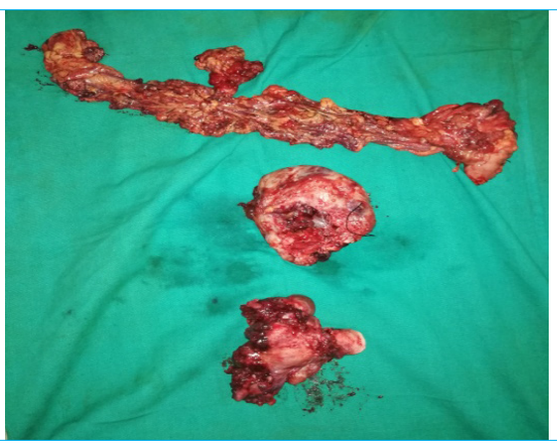

Figure 3. Resected specimen of uterus, ovarian mass and right hemicolon with adherent omentum.

Histopathology report of the resected tissues revealed mucin secreting ruptured mucinous carcinoma of bilateral ovaries with omental and colonic metastasis leading to pseudomyxoma peritonei. The maximum tumor dimension of right ovarian mass was 5 cm and that of left ovarian mass was 11 cm. However, the histopathology of appendix showed it was free of tumor. (FIGO Stage IIIC). Post-operative recovery was uneventful and she received adjuvant chemotherapy.

DISCUSSION

Pseudomyxoma peritonei is a borderline malignancy of ovary and/or vermiform appendix characterized by the dissemination of semisolid gelatinous mucinous implants in the peritoneal cavity. It is a poorly understood disease with controversial origin. Although the primary origin in both men and women is predominantly appendix, a synchronous ovarian and appendiceal tumor is common in females. However, immunohistochemistry and molecular genetic techniques support the hypothesis that the ovarian tumor is metastatic from a perforated appendiceal mucinous tumor in females.[2] Both ovarian and appendiceal adenomas or adenocarcinomas (non-cancerous and cancerous tumors) can cause pseudomyxoma peritonei. Rarely, pseudomyxoma peritionei has been reported among primary tumors of colon, stomach, pancreas and urachus.

The disease is usually diagnosed after the age of 40 and is more common in women than men.[3] Clinically, the patient complains of abdominal distension, pain corresponding to the site of primary tumor and may present with a mass per abdomen. An accurate pre-operative diagnosis is challenging. The tumor markers CEA (Carcino-embryogenic antigen) and CA-19.9 serve as a valuable tool for diagnostic and prognostic evaluation of pseudomyxoma peritonei. The CA-125 is not widely used tumor marker for pseudomyxoma peritonei, instead, used to exclude an ovarian neoplasm.[4] CT is currently the optimal imaging modality for diagnosis, and ultrasound guided tissue biopsy may also be useful.

The conventional treatment of pseudomyxoma peritonei is surgical debulking of tumor, i.e., removal of right hemicolon with appendix, removal of uterus with bilateral tubes and ovaries in females, omentectomy and removal of secondary implants. However, due to the presence of tumor deposits after the first debulking surgery, recurrence is high and overall 5 and 10-year survival rates are approximately 50% and 20% respectively.[5] Hence Sugarbaker et al in 1994 altered the surgical procedure by radical peritonectomy followed by HIPEC (Hyperthermic Intraperitoneal Chemotherapy) with an aim to eliminate macroscopic and microscopic implants as much as possible.[3,4] The surgery included removal of the right hemicolon, spleen, gall bladder, greater and lesser omentum, uterus and ovaries, rectum

in few cases and stripping of peritoneum off the pelvis, diaphragm and liver. Peritonectomy, omentectomy and combination HIPEC with mitomycin C and 5-fluorouracil has been reported to achieve a 10-year survival rate of up to 80%.[5] The reported prognostic factors for pseudomyxoma peritonei are age, histology, residual tumor volume and intraperitoneal chemotherapy.[2]

CONCLUSIONS

Since pseudomyxoma peritonei is a rare disease with non-specific clinical and radiological findings, the pre-operative diagnosis may be missed. In this case, the pre-operative diagnosis of pseudomyxoma peritonei was radiologically difficult, hence, it was only made after surgical exploration and histological analysis of the resected tissues.

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