

HUGE OVARIAN MUCINOUS CYSTADENOMA AND PSEUDOMYXOMA OVARII/PERITONEI: A CASE REPORT.

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Abstract

Background: Mucinous tumours, the second most common type of epithelial ovarian tumours, are the largest tumours that can be found in the human body. Pseudomyxoma peritonei is a rare clinical entity characterised by progressive accumulation of extracellular mucinous material in the abdominal cavity and tumoural implants on the peritoneal surfaces. Though of controversial origin, ruptured appendiceal mucinous adenoma being most commonly associated with it, mucinous tumours of the ovary have been notable cause (pseudomyxoma ovarii). Patients may present with features of abdominal mass effect among others. Abdominopelvic USS and CT are useful tools for evaluation but definitive diagnosis is often by laparotomy or laparoscopy. Debulking surgery with or without intraperitoneal hyperthermic chemotherapy is the treatment.

Case presentation: A 68-year-old P₄⁺¹(4 alive) woman, known diabetic and 17years postmenopausal, presented with a 4-month history of progressive abdominal swelling and discomfort, leg swelling, generalised body weakness and dizziness, early satiety, anorexia, weight loss and bloatedness. She was chronically ill-looking with pedal oedema and abdominal distension. Results of investigations supported ovarian mucinous cystadenoma with low risk of malignancy. She had appropriate surgery with evacuation of 7.5Litres of clear mucin from the abdominal cavity. She had a good postoperative course and remained well 18 months later. Histopathology findings confirmed diagnosis with no evidence of malignancy.

Conclusion: Pseudomyxoma peritonei is a rare condition, in association with mucinous cystadenomas; without appendiceal involvement, is even more rare hence may go unrecognized and be fatal if poorly managed.

Keywords:Pseudomyxoma-peritonei (PMP), Pseudomyxoma-ovarii, mucin, cystadenomas, cytoreduction, chemotherapy.

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INTRODUCTION

Mucinous tumours are the second most common type of epithelial ovarian tumours. They are the largest tumours that can be found in the human body.^{1,2} Pseudomyxoma peritonei develops from progressive accumulation of mucin in the abdominal cavity from mucinous tumours of ovarian (pseudomyxoma ovarii) or appendiceal origin, often leading to bowel obstruction.¹⁻³

CASE PRESENTATION

A 68-year-old P₄⁺¹(4 alive) woman, known diabetic and 17 years postmenopausal, who presented with a 4-month history of progressive abdominal swelling and discomfort, leg swelling, generalized body weakness and dizziness, early satiety, anorexia, weight loss and bloatedness.

She was chronically ill-looking, mildly pale, with bilateral pitting pedal oedema up to the mid shin. Her abdomen was grossly distended with an ill-defined abdomino-pelvic mass extending to the umbilicus; ascites was demonstrable by fluid thrill. RBS-7 mmol/L, HbA1c-13%, Ca125-21.3 U/ml (0-35), CEA-172.3 ng/ml (<8.5); PCV was 32%, and other baseline investigations were normal. Abdominopelvic USS findings were consistent with ovarian mucinous cystadenoma. Abdominopelvic CT had impression of hydatid cyst, multiple ovarian cysts to rule out serous cystadenocarcinoma. Risk of malignancy index (RMI) was 191.7. She had staging laparotomy and intra-operative findings were: massive thick mucinous ascites (7.5 L of clear mucin), huge left mucinous cystic ovarian mass (20 cm x 15 cm) exuding mucin from its breached surface, and no evidence of metastatic nodules on the visceral or parietal peritoneum. Right ovary was normal in size, but hard in consistency.

She had total abdominal hysterectomy, bilateral salpingo-oophorectomy, infra-colic omentectomy, appendectomy and copious peritoneal lavage. Histopathology findings were: Ovary – mucinous cystadenoma; uterus – squamous metaplasia (of the endocervix), endometrial polyp and degenerating leiomyoma uteri. Omentum-inflammatory, no evidence of malignancy. Appendix-peritonitis, no evidence of malignancy. Ascitic fluid cytology-acellular. She is on follow-up visits in clinic, and at the time of this report (more than 18 months post-surgery), her condition has remained satisfactory.

DISCUSSION

Mucinous cystadenoma is a benign tumour that arises from the surface epithelium of the ovary as the second most common type of epithelial ovarian tumour. It constitutes 15 – 25% of all ovarian tumours. The more massive an ovarian tumour is, the more likely it may be mucinous.^{1,2} It occurs in women between 30 and 60 years, with a mean age of 50 years (though the patient was 68-year-old), 10% are bilateral, about 75 – 85% are benign, and up to 15% progress to mucinous

cystadenocarcinoma.³ It may be asymptomatic, but most often presents with: abdominal swelling and discomfort/pain and gastrointestinal symptoms; which the patient presented with. Other clinical features may include: cardiorespiratory embarrassment, urinary symptoms, oedema of the legs, varicose veins, haemorrhoids and uterine prolapse.¹⁻³ Complications include ovarian cyst accidents, malignancy, and rarely pseudomyxoma peritonei/ovarum. It is the implantation of mucinous tumour cells in the peritoneum with production of copious amounts of mucin (pseudomyxoma peritonei).¹⁻⁴ However if the focus of mucin release is directly from the mucinous tumour cells of the ovary, it may be referred to as pseudomyxoma ovarum, as may have occurred in the patient.

Treatment considerations for ovarian mucinous cystadenoma include: patient's age, clinical presentation, tumour size, nature of cyst - simple, unilateral, unilocular, RMI, surgical fitness and woman's wishes.¹⁻⁴ Treatment options could be conservative; or surgical with or without hyperthermic intraperitoneal chemotherapy (HIPEC). Conservative management may be employed when patient is asymptomatic, cyst is simple, unilateral, unilocular with size- <5 cm, low-risk RMI in the presence of normal serum CA125 levels, woman's wishes, or if not surgically fit. Repeat evaluation should then be done in 4–6 months. For younger women, ovarian cystectomy or unilateral salpingo-oophorectomy can be done, but for women >40 years, total abdominal hysterectomy with bilateral salpingo-oophorectomy is recommended.¹⁻⁴ Treatment for pseudomyxoma peritonei/ovarum and if RMI \geq 200 Include cytoreductive surgery with adjuvant therapy which include HIPEC, mucolytic agents (Irrigation/lavage-with dextrose/water, dextran sulphate, plasminogen activator/urokinase), chemotherapy (post-op intraperitoneal or IV-melphalan, cisplatin), radiotherapy and photodynamic therapy. Our patient did not get the above adjuvant treatment because of her low-risk RMI and low index of suspicion of this rare complication (pseudomyxoma peritonei/ovarum)

before the surgery, as well as the fact that the histology results all showed no malignancy (therefore not requiring adjuvant chemotherapy).



Figure 1: Huge mucinous cystadenoma of the ovary.

CONCLUSION

Pseudomyxoma peritonei is a rare condition, in association with mucinous cystadenomas; without appendiceal involvement, is even more rare hence may go unrecognized and be fatal if poorly managed.

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