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Case Report

Pseudomyxoma peritonei from an ovarian mucinous cystadenoma mimicking ovarian cancer

Lamyae Errami*, Hajar Jakhjoukh, Hafsa Taheri, Hanane Saadi, Ahmed Mimouni

Department of Gynecology-Obstetrics and Reproductive Medicine, Mohammed VI University Hospital, Oujda, Morocco

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***Correspondence:**
Dr. Lamyae Errami,
E-mail: lamyae.errami@gmail.com

ABSTRACT

Pseudomyxoma peritonei (PMP) is a rare, borderline malignant condition, most commonly originating from the appendix, with ovarian origin being extremely rare. Rupture of ovarian mucinous tumours can result in dissemination of gelatinous mucus and tumour cells throughout the peritoneal cavity, leading to accumulation of ascites. We report a 56-year-old grand multiparous woman presenting with chronic abdominal pain and progressive distension. Physical examination revealed stable vital signs and overweight status (BMI 27 kg/m²). Laboratory investigations showed mild anaemia, and preoperative serum tumor markers (CA-125, CEA, CA 19-9) were evaluated. Imaging revealed a large multiloculated pelvi-abdominal cystic mass (22×19 cm) with massive ascites and peritoneal calcifications, suggestive of ovarian malignancy. An exploratory laparotomy revealed a 20 cm left ovarian mass with solid and multiloculated gelatinous components, with an appendix slightly enlarged, while the right adnexa and the uterus were unremarkable. Accidental intraoperative rupture released approximately 3 liters of gelatinous ascites. The patient underwent left salpingo-oophorectomy, appendectomy, multiple peritoneal biopsies, and cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (HIPEC). Histopathology confirmed PMP originating from an ovarian mucinous cystadenoma.The patient remains under regular surveillance. Complete cytoreductive surgery with HIPEC offers the best prognosis, though long-term follow-up is essential due to the risk of recurrence. Ovarian-origin PMP is rare and may mimic advanced ovarian malignancy. Accurate diagnosis relies on histopathology, and management requires complete cytoreduction combined with HIPEC, with vigilant long-term follow-up.

Keywords: Pseudomyxoma peritonei, Ovarian mucinous cystadenoma, Ovarian tumor, Cytoreductive surgery, Hyperthermic intraperitoneal chemotherapy, Gelatinous ascites, Peritoneal carcinomatosis

INTRODUCTION

Pseudomyxoma peritonei (PMP) is a rare, borderline malignant condition usually linked to the rupture of mucinous tumors of the appendix, ovaries, or other gastrointestinal tract organs, with the appendix being the most common location of the initial tumor, and the ovaries being an extremely rare origin.[1] The overall incidence of PMP is estimated to be one to two cases per million per year.[2] Rupture of ovarian mucinous tumors results in the dissemination of gelatinous mucus and tumor cells throughout the abdominal and pelvic cavities. This leads to the accumulation of jelly-like ascites, primarily within the peritoneum, as well as in the abdominal and pelvic regions.[3]

CASE REPORT

Our patient was a 56-year-old grand multiparous woman (parity 6) with no relevant past medical history. She presented with chronic abdominal pain and progressive abdominal distention. She reported no nausea, vomiting,

or changes in bowel habits but complained of increased urinary frequency. Physical examination revealed no pallor and stable vital signs. The patient was overweight, with a BMI of 27 kg/m² (weight 78 kg, height 170 cm). Her abdomen was grossly distended with full flanks; it was soft and non-tender. Percussion notes were dull over the entire abdomen.

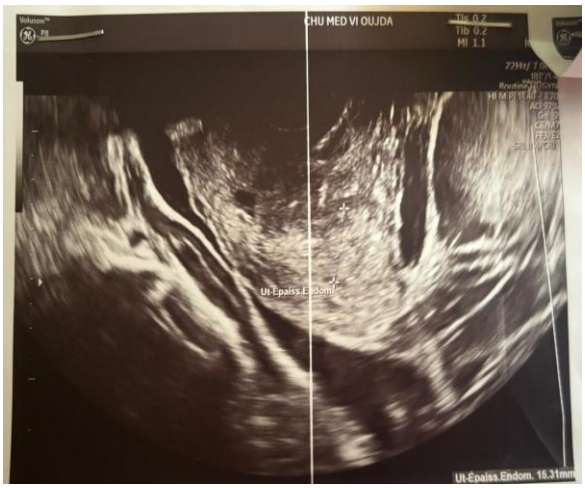

Figure 1 : Transvaginal ultrasound showing a large adjacent multiloculated cystic mass displacing the uterus, associated with ascites and endometrial thickening.

Her vulva, vagina, and cervix were grossly normal. Examination of the cardiovascular and urogenital systems was unremarkable. Laboratory investigations revealed anemia with a hemoglobin level of 9.62 g/dl, while the remaining complete blood count parameters were within normal limits. The coagulation profile was within the normal range. Renal and hepatic function tests were within normal limits. Serum tumor markers were measured, including CA-125, CEA, AFP, and CA 19-9.

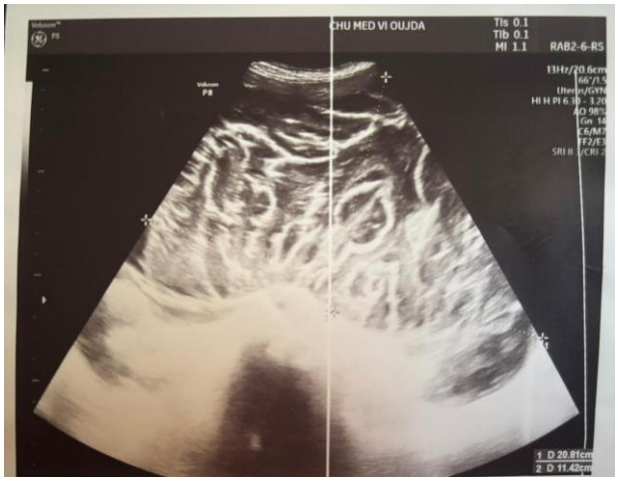

Figure 2 : Transabdominal ultrasound image revealing a large multiloculated cystic mass of suspected ovarian origin with internal septations.

The results showed elevated levels of CA-125 (140 U/ml), CEA (118 ng/ml), and CA 19-9 (65 U/ml), while AFP was within the normal range (<2 ng/ml). Ultrasound examination revealed a large multiloculated pelvi-abdominal cyst measuring 22 cm in the anteroposterior dimension, with internal septations, suggestive of a malignant ovarian tumor associated with massive ascites. The uterus was bulky and was distinguished from the tumor on ultrasound.

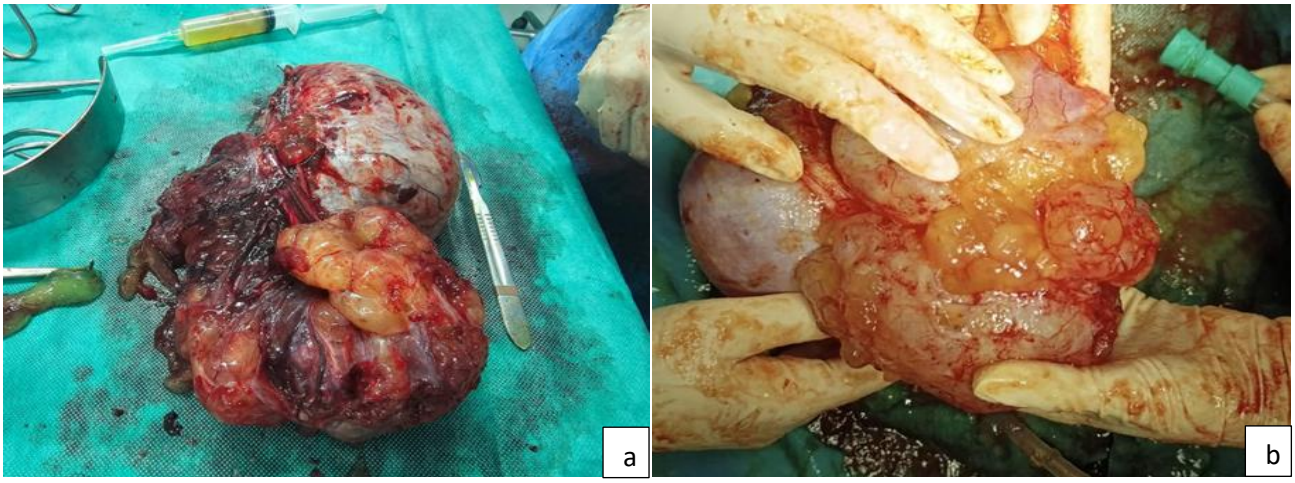

Figure 3 (a and b): Intraoperative findings mimicking advanced ovarian carcinoma, showing abundant gelatinous mucinous ascites and a large left ovarian mucinous mass.

CT scan revealed a large pelvi-abdominal cystic mass measuring approximately 22x19 cm with multiple internal septations, suggestive of ovarian mucinous cystadenocarcinoma. Massive ascites, diffuse the peritoneal calcifications, and peritoneal thickening consistent with peritoneal carcinomatosis were also noted. An exploratory laparotomy was performed, during which accidental rupture of the ovarian cyst occurred, releasing a large volume of gelatinous mucinous ascites with an estimated volume of 3 liters. Upon intraoperative

inspection, a large left ovarian mass measuring approximately 20 cm in its greatest dimension, with both solid and multiloculated gelatinous components, was observed, while the right adnexa appeared unremarkable. The uterus was of normal size, and the appendix was slightly enlarged, located in a lateral-internal caecal position. Gelatinous mucinous ascites and the large left ovarian mucinous mass. Postoperatively histopathological examination revealed no tumor involvement of the appendix, supporting the diagnosis of pseudomyxoma peritonei originating from an ovarian mucinous cystadenoma. The patient underwent a cytoreductive laparotomy (Left adnexectomy and appendectomy, accompanied by multiple peritoneal biopsies) and intraperitoneal chemotherapy.

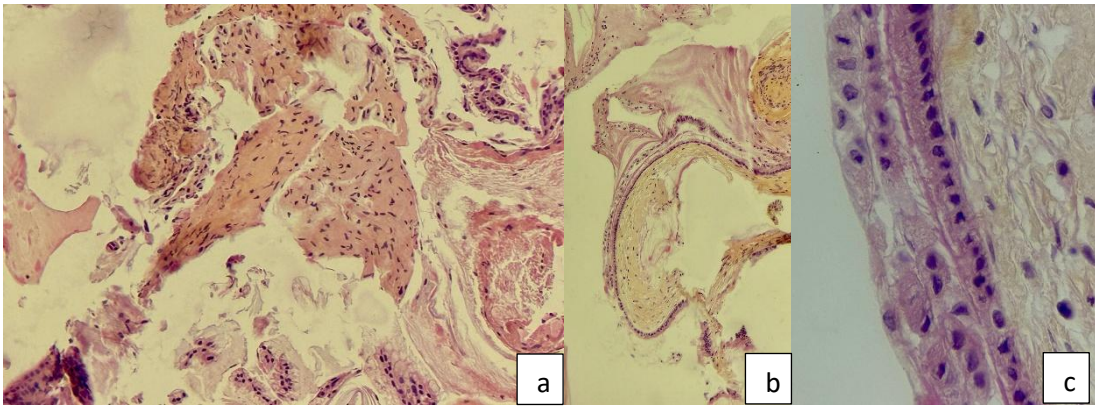

Figure 4 (a-c): Histopathological section of the ovarian mucinous cystadenoma showing mucin-producing epithelial lining without stromal invasion, consistent with a benign lesion (hematoxylin and eosin stain × 200).

DISCUSSION

Clinical presentation

PMP commonly stays asymptomatic until its advanced stages, with many cases being incidentally discovered during surgery.[3] Abdominal distension, abdominal or pelvic pain, the presence of a mass, and weight loss may be observed in cases of ovarian-origin PMP.[1-3]

PMP is classified, according to the histology of peritoneal disease, into 3 categories based on histopathologic features and the volume of tumor cells (low grade, high grade and high grade with signet ring cells).[4]

Imaging findings

Radiologic features of PMP include scalloping of the hepatic and splenic margins due to mucinous peritoneal implants, multiple multilocular cystic masses with rims of curvilinear calcifications, which were often seen in mucin-producing tumors, and compression of abdominal viscera at Computed Tomography, and an echogenic mantle with ascitic septations at ultrasonography.[5,6]

In addition to helping with staging and confirming the nature of the ovarian tumor and ascites, contrast-enhanced CT and MRI imaging can also be used to predict the presence of distant metastases, peritoneal spread, and lymph node enlargement.[7] According to a recent study preoperative ultrasonography was found to have the potential value in predicting pathological grade, complete cytoreduction possibility and the benefits of surgery.[8]

Treatment

The optimal treatment for pseudomyxoma peritonei is cytoreductive surgery (CRS), consisting of complete macroscopic tumor excision combined with hyperthermic intraperitoneal chemotherapy (HIPEC).[9] Residual disease following CRS is assessed using Sugarbaker's Completeness of Cytoreduction (CC) score: absence of residual tumor or residual nodules measuring less than 2.5 mm are classified as CC-0 (complete cytoreduction) or CC-1 (optimal cytoreduction), respectively, whereas residual disease exceeding 2.5 mm corresponds to CC-2 or CC-3 and is considered inadequate cytoreduction.[10] Higher CC scores are associated with increased recurrence rates and reduced overall survival. Systematic appendectomy is currently recommended in the management of pseudomyxoma peritonei, regardless of the macroscopic appearance of the appendix, as occult appendiceal neoplasia may be present and the appendix remains the most frequent primary origin of the disease.[4]

Prognostic/ recurrence

The prognosis of PMP largely depends on several factors, including histopathological grade, tumor burden, and the completeness of cytoreduction. Low-grade PMP is associated with a more favorable outcome compared with high-grade disease or tumors containing signet ring cells. Complete cytoreductive surgery (CC-0/CC-1) has been identified as one of the most important predictors of improved overall and disease-free Survival. Despite optimal treatment, recurrence remains a significant concern, and the principal site of recurrence was the peritoneum. Recurrence rates following cytoreductive

surgery combined with hyperthermic intraperitoneal chemotherapy have been reported to range between 20% and 30%, with most recurrences occurring within the first five years after treatment.[11] Incomplete cytoreduction, high-grade histology, and elevated preoperative tumor markers are associated with a higher risk of recurrence.

Follow up

Postoperative follow-up strategies generally rely on regular clinical assessment combined with imaging and tumor marker monitoring. CT scan or MRI remains the cornerstone of radiological surveillance for detecting peritoneal recurrence, while serum tumor markers may provide complementary information during follow-up.[12] Most recurrences are reported within the first five years following treatment, although late recurrences underline the necessity of extended monitoring. In the présent case, the patient is currently under surveillance and will continue follow-up at regular intervals according to institutional protocol.

CONCLUSION

PMP originating from a benign ovarian mucinous cystadenoma is an extremely rare entity that may clinically and radiologically mimic advanced ovarian malignancy with peritoneal carcinomatosis. This case highlights the diagnostic challenges associated with ovarian-origin PMP and underscores the critical role of histopathological examination in establishing the correct diagnosis. Complete cytoreductive surgery with hyperthermic intraperitoneal chemotherapy remains the mainstay of treatment, while long-term follow-up is mandatory due to the risk of recurrence.

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