Case Report

Appendiceal Carcinoma Presenting as Adnexal Mass With Pseudomyxoma Peritonei—A Case Report and Review of Literature

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Clinical Practice Points

- Pseudomyxoma peritonei (PMP) is an uncommon tumor presenting with mucinous ascites and pressure symptoms. Females usually present with abdominal distension and ovarian mass. Cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy is the treatment of choice.
- Our patient had typical presentation of PMP who presented with ovarian mass. Left ovary showed a big tumor with mucinous ascites, and the appendix was normal looking. Histopathology revealed the primary appendiceal tumor, and mass in the ovary was metastatic.
- This case report highlights the importance of complete surgery including appendicectomy as most of the times primary tumor arises from the appendix.

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Introduction

Pseudomyxoma peritonei (PMP) is an uncommon clinical condition characterized by a slow and progressive accumulation of peritoneal implants and mucinous ascites. Accumulation of mucin in peritoneal cavity results in massive symptomatic distension of abdomen. Twenty to thirty percent of female PMP patients present with an ovarian mass found during the evaluation for lower abdominal pain, a pelvic mass, menstruation problems, or infertility.[1,2] Although earlier there was controversy in nomenclature, it is now generally accepted that PMP arises as a result of neoplastic mucin-secreting cells with low-grade cytologic features disseminating within the peritoneal cavity. Appendiceal and ovarian neoplasms give rise to majority of the cases. We are presenting a case of adnexal (ovarian) mass with PMP resulting from appendiceal carcinoma.[1-3]

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Case Report

A 45-year-old multiparous woman presented with gradually progressive abdominal distension and loss of appetite and weight for 6 months. She had bronchial asthma and uterovaginal prolapse for 10 years. There was no alteration in bowel or bladder function or awareness of mass. She attained menopause 6 years ago, and perimenopausal transition was smooth. She had 5-term vaginal deliveries and used barrier contraception.

On examination, patient's general condition was good with pulse rate of 100/min, blood pressure of 140/80 mm Hg, and respiratory rate 30/min with mild pallor. Chest auscultation revealed bilateral rhonchi. Abdominal distention was present, with umbilicus everted, and small umbilical hernia was seen with moderate amount of free fluid but no palpable mass. On local examination, second-degree cervical descent with large rectocele and minimal cystocele was noticed. Uterus was normal sized, and there was no palpable adnexal mass. Paracentesis of approximately 30 mL revealed thick mucinous fluid. Preoperatively, she received *salbutamol* and *ipratropium* nebulization, intravenous *hydrocortisone*, and *deriphyllin* along with supportive treatment in consultation with pulmonologist for acute exacerbation of bronchial asthma. She was planned for laparotomy and taken up for surgery under high risk in view of bronchial asthma. Preoperative investigations revealed hemoglobin of 8.0 g/dL, renal and liver function tests were within normal limits, x-ray chest was normal, pulmonary function tests showed mild obstruction, arterial blood gas on room air ($pH/pO_2/pCO_2/HCO_3^-$/Base excess/$SatO_2$)

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was 7.41/145/32/20/−4/99%, and CA-125 = 29.6 U/mL. Peritoneal cytology was suggestive of PMP. The peritoneal cytology showed thick mucinous material with a few scattered mucinous epithelial cells having cytoplasmic vacuolation and eccentric nucleus with bland chromatin admixed with a few macrophages. There was no evidence of frank mucinous carcinoma. Ultrasonography revealed normal liver, spleen, gall bladder, and both kidneys; uterus normal size with thin endometrium; and large cystic mass in pelvis with internal septations and heterogenous foci. Mass showed coarse echotexture and was full of complex soft tissue echoes.

She underwent laparotomy under combined epidural and general anesthesia. Five kilograms of mucinous ascetic fluid was drained. Uterus, both tubes, and right ovary were normal. Left ovary was replaced by a tumor of 10 × 6 cm with capsule ruptured and mucinous material coating the whole of peritoneal cavity. Surface of liver, undersurface of diaphragm, and bilateral paracolic gutters were free of tumor, and there were no palpable retroperitoneal lymph nodes. Appendix was retrocecal 3 × 1 cm and buried in adhesions. Total abdominal hysterectomy with bilateral salpingo-oophorectomy and infracolic omentectomy and appendicectomy was done. Small paraumbilical hernia defect of 2 × 2 cm was present, which was repaired. Peritoneal wash was done with 5% dextrose. Pelvic floor repair was deferred because of high risk of anesthesia with prolongation of surgery. Postoperative period was uneventful, and she received intravenous amoxyclavulanic acid and metronidazole. She was discharged on the second postoperative day.

Histopathology revealed the following in the left ovary: mucinous tumor, borderline malignant, and metastatic. Tumor deposits were present on serosal aspect of left fallopian tube. Right ovary, right tube, endomyometrium, and cervix were unremarkable. No tumor deposits were seen on the omentum. Appendix showed mucinous tumor of borderline malignancy. Peritoneum revealed PMP. Overall features were suggestive of PMP, disseminated peritoneal adenomucinosis type (Figures 1-4).

Postoperatively, she received 6 cycles of oxalplatinum and 5-flourouracil and under regular follow-up.

Figure 1 Ascitic Fluid Smear Showing Thick Mucinous Material With Low Cellularity (Papanicolaou Stain, Oil Immersion Microscopy ×100)

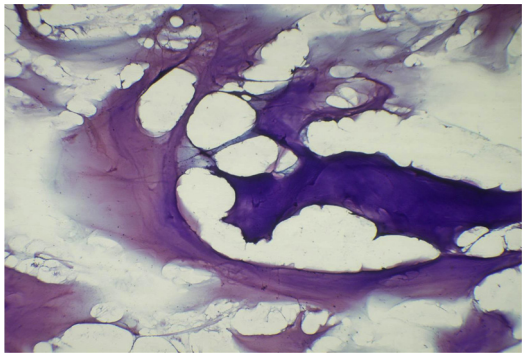
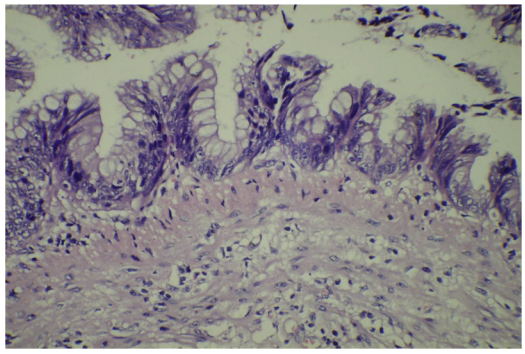


Figure 2 Appendicular Tumor Showing Features of a Borderline Mucinous Neoplasm (Hematoxylin-Eosin Stain, Oil Immersion Microscopy ×200)



Discussion

PMP is an enigmatic syndrome characterized by copious production of mucinous ascites that, over time, fills the peritoneal cavity. It is a rare condition, incidence being 1 per million per year. Classically, it presents at laparotomy as "jelly belly." The term was coined by Werth in 1884 when he described the condition in association with mucinous carcinoma of the ovary. In 1901, Frankel described a case of PMP in association with a cyst of the appendix. Definition, origin, etiology, and treatment of this peculiar condition are controversial. There is a spectrum of disease varying from mucinous cystadenomas of appendix to mucinous cystadenocarcinoma arising anywhere in abdominal cavity. Recently, it has been proposed that the term PMP syndrome be strictly applied to a pathologically and prognostically homogenous group of cases characterized by histologically benign peritoneal tumors that are frequently associated with an appendiceal mucinous adenoma and have a unique natural history. Pathologically, the peritoneal mucinous lesions are termed "disseminated peritoneal adenomucinosis" or simply adenomucinosis. Cases of peritoneal

Figure 3 Serosal Aspect of the Appendix Showing Thick Mucinous Material Confirming Pseudomyxoma Peritonei (Hematoxylin-Eosin Stain, Oil Immersion Microscopy ×200)