

Endometriosis inflammation mimicking pseudomyxoma peritonei: A case report☆



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1. Introduction

Endometriosis is a common gynecologic condition affecting 6–10% of females and up to 50% of women with pelvic pain and/or infertility [1]. Signs and symptoms of endometriosis include dysmenorrhea, dyspareunia, dysuria, dyschezia, menorrhagia, and infertility. Common clinical findings include endometriomas, pelvic and peritoneal implants, and pelvic adhesions. Endometriosis is most often diagnosed by the presence of pelvic pain in relation to menstruation, although definitive diagnosis is made by laparoscopic identification of implants and pathologic sampling. Endometriosis may mimic other conditions, including ovarian neoplasms, ectopic pregnancy, pelvic inflammatory disease, interstitial cystitis, adenomyosis, iatrogenic pelvic adhesions, irritable bowel syndrome, colon cancer, or diverticulosis [1]. Treatment is based on severity of symptoms and the patient's reproductive plans. Treatment options include pain management, hormonal suppression, or definitive surgical management.

Pseudomyxoma peritonei (PMP) is a rare condition characterized by mucinous ascites and mucinous epithelium within the peritoneal cavity, most commonly associated with gastrointestinal or primary peritoneal carcinomas of the appendix [2]. PMP is suspected if computerized tomography (CT) shows peripheral fluid collections with centralization of abdominal organs and scalloping of abdominal organs, as well as the presence of increased abdominal girth and malnutrition. Laboratory abnormalities include an elevated cancer antigen 19-9 (CA 19-9) and

cancer antigen 125 (CA 125) [2]. Treatment involves surgical resection followed by hyperthermic intraperitoneal chemotherapy (HIPEC). We present a case of a young women with endometriosis whose clinical presentation was consistent with PMP.

2. Case Report

A 25 year-old nulliparous female presented to her primary care physician with three months of diffuse abdominal pain and a 30-pound weight gain. Her medical history was significant for migraines, depression, and idiopathic hemolytic anemia requiring transfusion in 2011. Family history was significant for a brother with leukemia. She had normal menstrual cycles, no dyspareunia or dysmenorrhea, and no change in bowel habits. She did later endorse occasional abdominal pain and brief, monthly episodes of epistaxis. On exam, her abdomen was protuberant with a positive fluid wave. Pelvic exam was limited by ascites but revealed a mobile, non-tender cervix without nodularity, a normal-sized, anteverted uterus, and no palpable pelvic masses. A urine pregnancy test was negative. CT scan revealed multi-loculated, multi-septated, low attenuation fluid filling the abdomen and pelvis and a 4.5 × 6.7 × 6 cm left adnexal mass (Fig. 1). The patient was referred to a gynecologic oncologist for further management.

Further studies included a complete blood count and chemistries, which were normal. CA-125 was elevated at 223. Imaging was concerning for pseudomyxoma peritonei given findings of the multi-septated fluid collections and bowel centralization.

3. Procedure

The patient was referred to gynecologic oncology for surgical intervention. An exploratory laparotomy was recommended for pelvic mass resection, fluid drainage, and possible hysterectomy, bilateral salingoophorectomy, bowel resection, appendectomy, and intraoperative hyperthermic intraperitoneal chemotherapy. Fertility preservation was desired.

She underwent an exploratory laparotomy, removal of loculated ascites, removal of peritoneal endometriotic implants, and ovarian preservation. Intraoperative findings were significant for large, cystic masses filling the abdominal cavity. However, these appeared to arise from the peritoneum rather than ovary or bowel and had no definitive blood supply. Additionally, adhesions and endometriomas were noted

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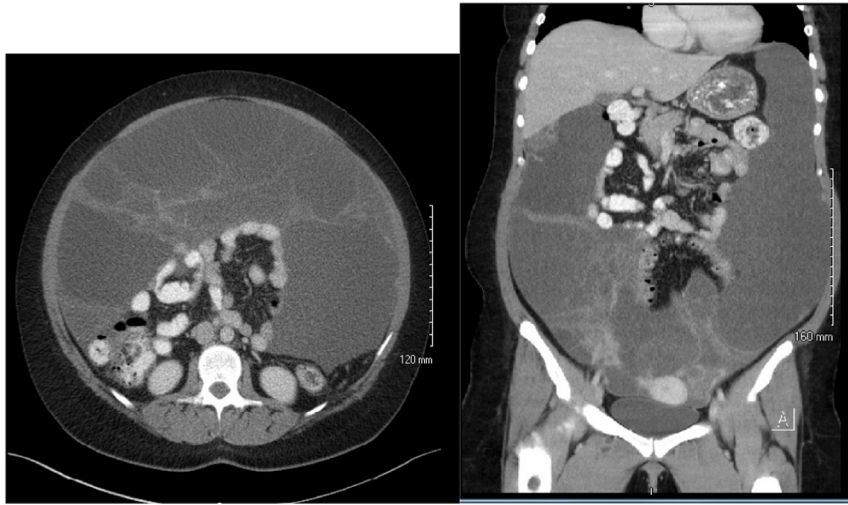


Fig. 1. CT images at presentation demonstrating multi-septated, low attenuation fluid with centralization of the bowel.

in the pelvis, especially the posterior cul-de-sac, and were carefully dissected from the peritoneum. Ovaries and uterus were without masses and were preserved. Appendectomy and omentectomy were performed, but bowel resection and chemotherapy were not indicated given the intraoperative findings.

Final pathology revealed a 30 cm mucicarmine and calretinin negative, WT-1 +/CD10 +/PR + serous cystadenoma with endometriosis. The patient had an uncomplicated postoperative course and was discharged home on postoperative day three with a prescription for an oral contraceptive (Fig. 2).

4. Discussion

The novelty of this case lies in the common features and dramatic differences in management between pseudomyxoma peritonei, an uncommon finding that portends malignancy, and endometriosis, which affects > 175 million women worldwide [3]. Moreover, the finding of a serous cystadenoma is thought to have arisen from preexisting endometriosis.

While endometriosis more commonly presents as isolated, superficial implants <5 cm in aggregate, a review of the literature has identified four case reports of endometriosis presenting as large, cystic endometriomas [4,5,6,7]. Endometriosis presenting alongside frank ascites is evident in the literature as well; however, this is more commonly confused with Meigs syndrome than pseudomyxoma peritonei [8,9]. An increasing body of evidence that endometriosis is associated with a chronic inflammatory state, increased cyclooxygenase-2 activity, elevated activated macrophages, and proinflammatory cytokines suggests

that fluid extravasation and edema is an expected albeit atypical finding in endometriosis [3,10]. This case highlights that endometriosis should be part of any differential diagnosis for premenopausal patients presenting with a complex pelvic fluid collection.

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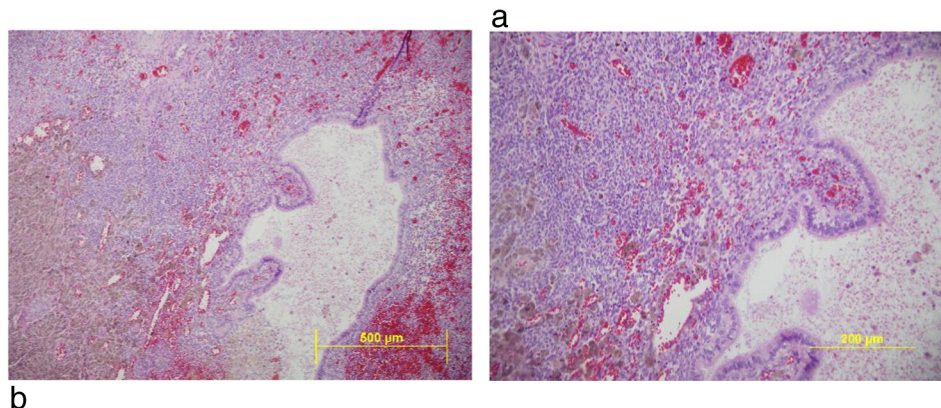


Fig. 2. Histologic images from cystic tissue featuring endometrioid gland with cilia and characteristic hemosiderin deposits (2a), as well as stroma with hyperemic blood vessels (2b).