

Severe Erosive Esophagitis Secondary to Gastric Outlet Obstruction Related to Pseudomyxoma Peritonei

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ABSTRACT

Pseudomyxoma peritonei (PMP) is a rare clinical condition characterized by a mucin-producing tumor. PMP tumor cells migrate to abdominal and pelvic sites, eventually enveloping intra-abdominal organs and compressing the gastrointestinal tract. Patients with PMP are often asymptomatic in early stages of the disease, but in later stages develop symptoms including abdominal pain, acute abdomen, increased abdominal girth, vomiting, and bowel obstruction. Nonspecific symptoms combined with a relatively modest accuracy of imaging modalities frequently lead to delay in PMP diagnosis and treatment, thereby increasing morbidity. We present a case demonstrating severe erosive esophagitis as a result of PMP-associated gastric antrum compression.

INTRODUCTION

Pseudomyxoma peritonei (PMP) is a rare tumor that occurs in 1–4 persons per million a year.¹ PMP begins as a mucin-producing tumor that predominantly originates from the appendix and ovary, but can also originate from the colon, rectum, gallbladder, bile ducts, stomach, urinary bladder, small intestine, pancreas, lung, breast, and fallopian tube.^{2–4} PMP metastasis mainly occurs after rupture of the primary lesion, which releases tumor cells into the abdominal cavity. Progressive gelatinous deposits eventually envelop the intra-abdominal organs and compress the gastrointestinal tract.⁴ Abdominal computed tomography (CT) scans of patients with PMP often show scalloping of the liver caused by compression by gelatinous ascites (“jelly belly”) from overproduction of mucin in the peritoneal cavity. This compressive physiology can also affect other organs including the gastrointestinal tract. Few cases have been reported in which PMP-associated symptoms originate from gastric antrum compression. We present a case demonstrating severe erosive esophagitis as a result of PMP-associated gastric antral compression.

CASE REPORT

A 70-year-old woman was evaluated for a 3-week history of intractable nausea, vomiting, and constipation. She endorsed no bowel movements for over 1 week despite daily over-the-counter osmotic laxatives, progressive abdominal distension, and emesis of the majority of her oral intake and salivary secretions. Medical history was significant only for hypertension, type 2 diabetes mellitus, hyperlipidemia, and peripheral vascular disease. Social history was significant only for a remote history of tobacco use. On admission, a noncontrast abdominal CT showed thickening of the cecum with stool throughout the colon, a large volume of abdominal ascites, and a nodular liver suggestive of cirrhosis. Laboratory studies showed normocytic anemia with a hemoglobin level of 8.3 g/dL; transaminases (alanine transaminase 16 U/L; aspartate transaminase 23 U/L), platelets ($228 \times 10^3/\mu\text{L}$), bilirubin (0.6 mg/dL), and international normalized ratio levels (1.2) were within normal limits.

The patient’s constipation resolved with administration of enemas and lactulose, but her nausea and vomiting persisted. A subsequent esophagogastroduodenoscopy showed severe circumferential erosive, necrotic-appearing esophagitis (Figure 1). Although the scope could be advanced into the duodenum, there was marked nondistension of the gastric antrum noted during the procedure

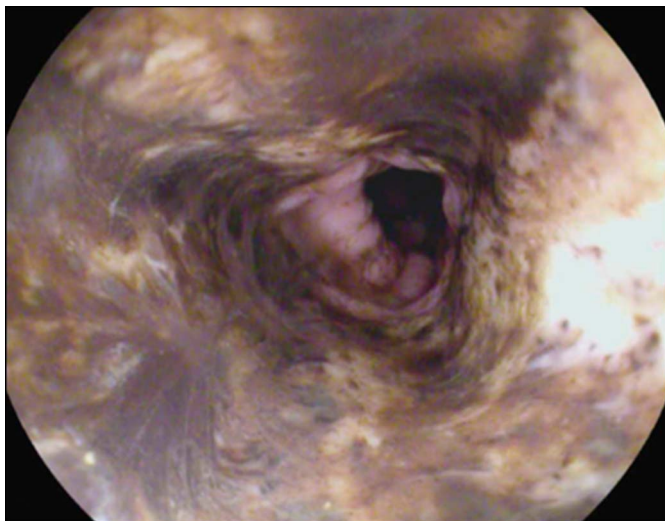


Figure 1. Endoscopic view of the distal esophagus and gastroesophageal junction revealing severe circumferential erosive esophagitis.

concerning for extrinsic compression (Figure 2). Antral biopsies were negative for malignancy. Ascitic fluid obtained through endoscopic ultrasound (EUS) was noted to be thick and difficult to aspirate; no obvious peritoneal nodules were observed on EUS. Analysis of the ascitic fluid was consistent with a nonperitoneal cause of ascites and showed elevated protein levels (3.6 g/dL) and a serum-ascites albumin gradient of 1.1 g/dL; the ascitic fluid was negative for malignant cells. A follow-up abdominal CT with contrast showed appreciable scalloping of the liver contour and mass effect to the stomach, suggestive of PMP (Figure 3). The levels of tumor markers carbohydrate antigen 125 and carcinoembryonic antigen in the blood were elevated at 120 U/mL and 132 ng/mL, respectively.

A diagnostic colonoscopy was attempted because of concern for cecal thickening in the setting of PMP, but was aborted due to

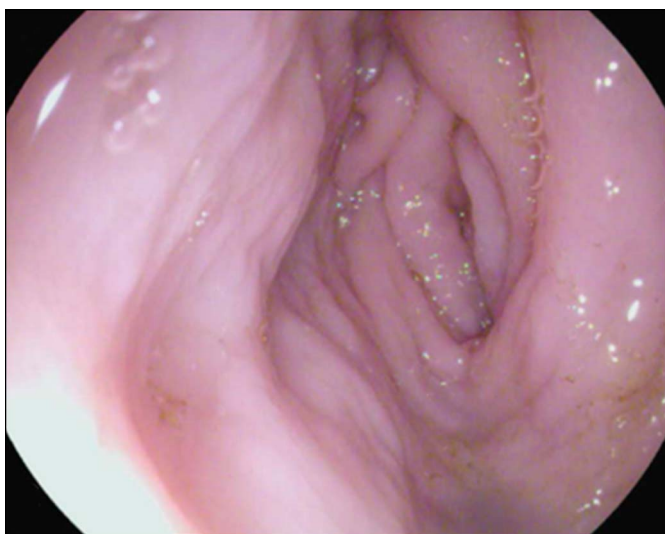


Figure 2. Endoscopic view of the gastric antrum showing non-distensible mucosa in the prepyloric region.

inability to traverse a nondistensible sigmoid colon. Ultimately, a diagnostic laparoscopy was performed for peritoneal fluid sampling. Peritoneal biopsy revealed multiple peritoneal masses with adherence to the adjacent small bowel. Peritoneal biopsy confirmed the diagnosis of poorly differentiated adenocarcinoma with signet ring cell morphology (Figure 4). In retrospect, the erosive esophagitis noted on initial esophagogastroduodenoscopy was attributed to corrosive injury in the setting of protracted vomiting due to delayed gastric emptying. Given the patient's medical comorbidities and her poor prognosis, oncology and palliative care were consulted, and she was discharged to inpatient hospice.

DISCUSSION

PMP remains a diagnostic challenge for physicians and radiologists. During initial stages, patients with PMP are often asymptomatic or exhibit symptoms that are misdiagnosed as irritable bowel syndrome.⁵ Vague symptoms and the limited sensitivities of current imaging modalities often delay the diagnosis of PMP, resulting in worsened clinical outcomes. Several imaging techniques can be used to diagnose PMP, including ultrasound with a parallel fine needle biopsy, magnetic resonance imaging, and positron emission tomography.^{6–8} However, CT with contrast is the most widely used imaging technique to diagnose PMP.⁵ CT aids in diagnosis by showing the distribution and infiltrated range of the primary PMP lesion. The CT scans of patients with PMP often show an abnormal density of ascites as well as omentum thickening, peritoneal infiltration, and mesentery changes; in addition, scalloping of the liver, spleen, and mesentery is frequently demonstrated.⁹ However, previous studies showed that CT identified PMP in only 51% of cases.¹⁰ Similarly, in our case study, the CT scan was not diagnostic for PMP.

Analysis of ascitic fluid can aid in the diagnoses of PMP. Histopathology can reveal varied epithelial cell differentiation as well as abundant extracellular mucinous material or malignant cells.⁷ EUS-guided paracentesis of ascitic fluid has a reasonable sensitivity ranging up to >90% in some studies with low adverse events (1%–4%), but can still miss the diagnosis of malignancy in a fraction of cases.^{11–13} If peritoneal nodules can be identified by EUS, fine needle aspiration of these lesions may improve sensitivity and diagnostic yield.^{11,13} Although case studies have documented the safety and reliability of EUS-guided biopsies specifically in the context of PMP, no larger studies have evaluated the true diagnostic utility of endoscopic biopsy for this condition.^{11,12,14} If EUS paracentesis is non-diagnostic, then laparoscopic sampling may be required as it was in this case study.

The presence of nonspecific tumor markers (eg, carcinoembryonic antigen, carbohydrate antigen 125, and carbohydrate antigen 19.9) can also point to a PMP diagnosis.^{15–18} Tumor marker levels are elevated in most patients with PMP.^{15,19}

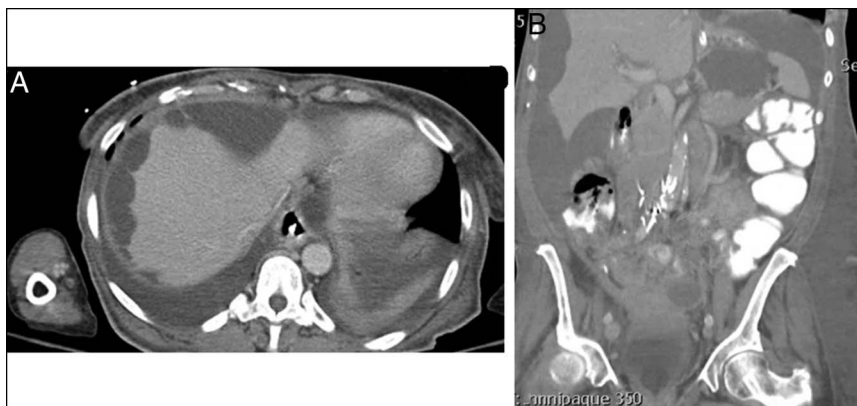


Figure 3. Contrast abdominal computed tomography images, (A) axial view, (B) coronal view, revealing classic “scalloping” of the liver in the setting of gelatinous ascites.

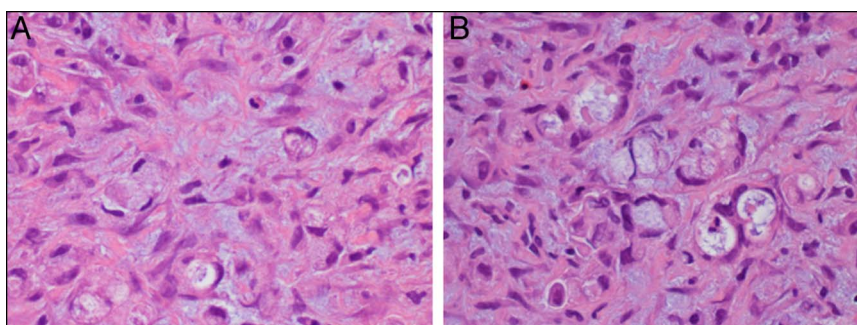


Figure 4. Biopsy of a peritoneal specimen at (A) hematoxylin and eosin stain, 100× magnification and (B) hematoxylin and eosin stain, 400× magnification showing tumor cells with prominent single cytoplasmic mucin vesicle and eccentrically placed hyperchromatic nuclei.

Furthermore, tumor marker levels can be used as a baseline value to assess patient prognosis during postoperative follow-up.⁶ In summary, PMP may present with variable symptoms; therefore, PMP must be considered as a differential diagnosis in a patient with symptoms consistent with gastrointestinal tract compression and new-onset high-protein ascites. An initial negative abdominal CT scan should not dissuade physicians from working up suspected PMP cases; a subsequent peritoneal biopsy must be obtained to confirm the diagnosis.

DISCLOSURES

Author contributions: DS Braun and B. Bushe wrote the manuscript. I. Lytvak provided pathology images. P. Kedia approved the final manuscript. DS Braun is the article guarantor.

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