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

David S. Braun, MD1, Bryce Bushe, MD2, Irina Lytvak, MD3, and Prashant Kedia, MD2

1Internal Medicine, Methodist Dallas Medical Center, Dallas, TX
2Gastroenterology, Methodist Dallas Medical Center, Dallas, TX
3Pathology, Methodist Dallas Medical Center, Dallas, TX

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 antral 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 × 10^3/uL), 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.

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Correspondence: David S. Braun, MD (DavidBraun@mhd.com).
concerning for extrinsic compression (Figure 2). Antral bi-
opies 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
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. His-
topathology 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 ad-
verse events (1%–4%), but can still miss the diagnosis of ma-
lignancy 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.11,12,14 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, carcinoem-
bryonic 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

DISCUSSION

PMP remains a diagnostic challenge for physicians and ra-
diologists. 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
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diagnostic for PMP.

A diagnostic colonoscopy was attempted because of concern for
cecal thickening in the setting of PMP, but was aborted due to
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 ade-
nocarcinoma with signet ring cell morphology (Figure 4). In
retrospect, the erosive esophagitis noted on initial esoph-
gastroduodenoscopy was attributed to corrosive injury in
the setting of protracted vomiting due to delayed gastric emp-
tying. Given the patient’s medical comorbidities and her poor
prognosis, oncology and palliative care were consulted, and she
was discharged to inpatient hospice.
Furthermore, tumor marker levels can be used as a baseline value to assess patient prognosis during postoperative follow-up. 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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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.
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