Abdominal Cocoon Syndrome Secondary to Signet Ring Cell Adenocarcinoma: A Rare Diagnostic Challenge

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ABSTRACT

Introduction: Abdominal cocoon syndrome, or sclerosing encapsulating peritonitis, is a rare condition characterized by the encasement of the small bowel in a dense fibrocollagenous membrane, often mimicking symptoms of bowel obstruction. Secondary ACS associated with malignancy is exceptionally uncommon.

Case presentation: A 44-year-old man with a history of ascites presented with intractable nausea, vomiting and severe cachexia. The diagnostic process posed significant challenges, requiring advanced imaging and invasive interventions to uncover the underlying malignancy.

Discussion: This case highlights the diagnostic challenges of secondary abdominal cocoon syndrome, emphasizing the importance of maintaining high clinical suspicion and utilizing advanced diagnostic tools in complex presentations.

Conclusions: Early recognition, advanced imaging, and a multidisciplinary approach are critical to optimizing outcomes in rare and challenging conditions.

INTRODUCTION

Abdominal cocoon yndrome (ACS), or sclerosing encapsulating peritonitis, is a rare condition characterized by partial or complete encapsulation of the small bowel by a dense fibrocollagenous membrane.[1] Most cases are idiopathic, but secondary causes include infections (eg, tuberculosis), malignancies, peritoneal dialysis, and postsurgical adhesions.[2] The incidence of ACS is challenging to estimate because of its rarity, but it has been reported more frequently in men in their 30s through 50s – particularly in regions where tuberculosis is endemic.[1,3] Case reports suggest that secondary ACS associated with malignancies, as observed in this case, is even rarer, making its diagnosis particularly challenging.[4] Existing literature reveals two such malignancy-associated cases: a 49-year-old man with liver cirrhosis and diffuse large B-cell lymphoma who had imaging and autopsy evidence of ACS, and a 16-year-old girl with extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue who had intraoperative findings of ACS.[5,6]

The clinical presentation of ACS often mimics bowel obstruction, with symptoms such as nausea, vomiting, abdominal distension, and cachexia.[1] Diagnostic delays are common because of the nonspecific nature of these symptoms and the lack of clear radiological markers, particularly in cachectic patients in whom reduced intra-abdominal fat can obscure key findings on conventional imaging modalities such as abdominal computed tomography (CT).[4,7] This case underscores the importance of maintaining a high index of suspicion, especially when conventional diagnostic methods fail to yield a clear etiology.

CASE PRESENTATION

A 44-year-old man presented with a 2-month history of intractable nausea and vomiting, accompanied by a 44-pound weight loss and profound cachexia. He reported an inability to tolerate both solids and liquids, necessitating initiation of a nasogastric tube with continuous suction and total parenteral nutrition for nutritional support. His social history included past smoking (10 pack-years), alcohol use, occasional marijuana use, and a lifelong career in roofing. His family history was unremarkable, and his medical history included ascites of unknown etiology, extensively evaluated 2 months earlier at an outside hospital.

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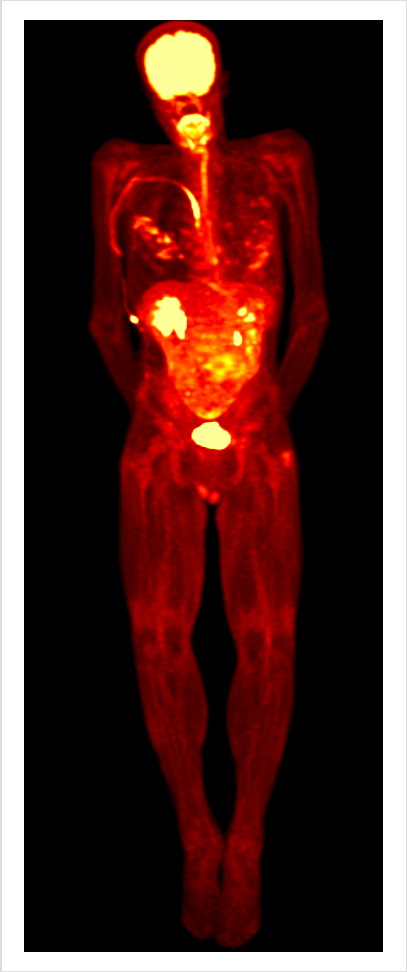
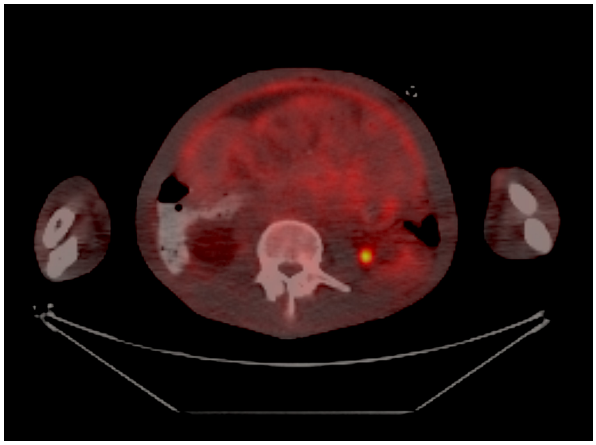
Figure 1. Three-Dimensional MIP PET–CT Image (Coronal View) Demonstrating Abnormal FDG Uptake Around the Peritoneum



Abbreviations: MIP, maximum intensity projection; PET, positron emission tomography; CT computed tomography; FDG, fluorodeoxyglucose.

Figure 2. Axial PET-CT Image Demonstrating Increased FDG Uptake in the Small Bowel With Surrounding Encapsulation



Abbreviations: PET, positron emission tomography; CT computed tomography; FDG, fluorodeoxyglucose.

Investigations at that time included ascitic fluid analysis, which revealed a high protein content (>2.5 g/dL) and a borderline serum-ascitic albumin gradient (SAAG) of 1.2. Cytology was negative for malignancy, and hepatitis and autoimmune panels were unremarkable. A transthoracic echocardiogram showed a normal left ventricular ejection fraction. Microbiological and cytological studies ruled out tuberculosis. Despite these efforts, no definitive cause for the ascites was identified. Abdominal ultrasound revealed mild nodular changes on the liver with otherwise normal morphology, and CT of the abdomen showed no cirrhotic changes. He was discharged with a diagnosis of idiopathic ascites and managed with spironolactone and furosemide.

At the time of the current presentation, the patient exhibited persistent nausea and vomiting, profound temporal wasting, and severe malnutrition. Physical examination revealed a cachectic male with a firm, nontender abdomen and minimal distension. Cardiovascular, respiratory, neurological, and extremity examinations were unremarkable. Laboratory findings showed elevated tumor markers, including CA-125, CA 19-9, and CEA; elevated alkaline phosphatase; and hypoalbuminemia. Endoscopic evaluation was pursued. Esophagogastroduodenoscopy (EGD) revealed Los Angeles (LA) grade B esophagitis, a 6-cm hiatal hernia, congested gastric mucosa, and poor gastric distension attributed to presumed extrinsic compression. Biopsy specimens taken during EGD were negative for malignancy. Endoscopic ultrasound (EUS) demonstrated diffuse gastric wall thickening and abnormal echotexture of the left hepatic lobe but was not conclusive. Of note, multiple EUS attempts were required because of resistance to insufflation. A CT scan of the abdomen and pelvis revealed diffuse peritoneal thickening, bowel wall edema, and nodular liver changes (Figure 1). However, the lack of intra-abdominal fat due to severe cachexia obscured key findings. Given the elevated alkaline phosphatase and nodular changes raising concern for a progressive fibrotic or malignant process, a liver biopsy was performed, which showed no significant fibrosis, cirrhosis, or malignancy.

With no clear etiology identified, the oncology service was consulted, and advanced imaging was pursued. A positron emission tomography (PET)-CT scan revealed fluorodeoxyglucose (FDG)-avid diffuse peritoneal thickening and abdominal cocooning (encasement of the small bowel in a dense fibro-collagenous membrane), suggestive of peritoneal malignancy (Figure 2). However, the primary source of malignancy remained elusive.

To establish a definitive diagnosis, a peritoneal biopsy was performed, which revealed signet ring cell adenocarcinoma of gastrointestinal origin with metastasis to the peritoneum. The final diagnosis of abdominal cocoon syndrome secondary to metastatic signet ring cell adenocarcinoma was established. Unfortunately,

Figure 3. Axial Computed Tomography Image Demonstrating Edema and Thickening of Peritoneum With Compartmentalization of Bowel Loops

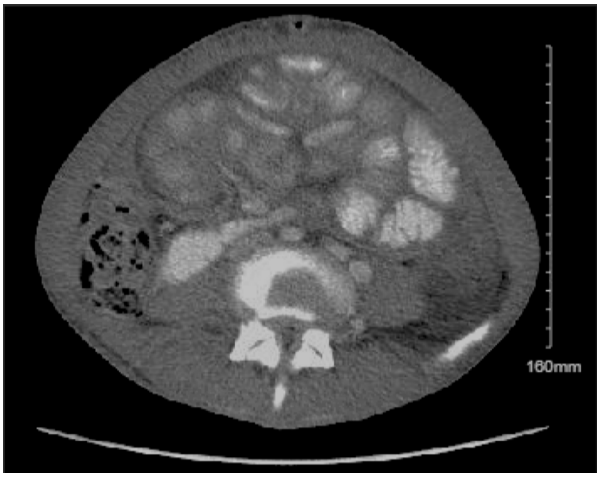
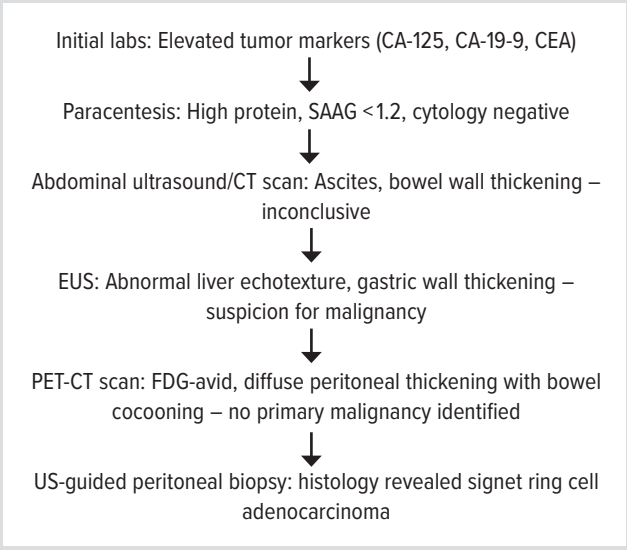


Figure 4. Flowchart Demonstrating Diagnostic Steps Leading to Final Diagnosis



Abbreviations: SAAG, serum-ascitic albumin gradient; EUS, endoscopic ultrasound; CT, computed tomography; PET, positron emission tomography, FGD, fluorodeoxyglucose.

the disease was advanced and deemed inoperable at the time of diagnosis. Due to the diffuse nature of the cancer, oncology considered surgery or radiation therapy unlikely to be helpful and systemic chemotherapy to be the only treatment option. Based on his severely malnourished state, frailty, poor performance status, and disease extent, systemic chemotherapy was cautioned to potentially aggravate the patient's symptoms without a clear idea whether the cancer would respond. He transitioned to palliative care, focusing on symptom management and quality of life. The nasogastric tube was kept in place for decompression, and while vomiting improved during hospital course, he was provided as-needed antiemetics on discharge.

DISCUSSION

Abdominal cocoon syndrome (ACS), also referred to as sclerosing encapsulating peritonitis, is a rare clinical entity typically presenting as intestinal obstruction. While idiopathic cases predominate, secondary causes such as tuberculosis, prior abdominal surgeries, peritoneal dialysis, and malignancy are well-documented.[1,2] This case, highlighting ACS secondary to metastatic signet ring cell adenocarcinoma, underscores unique diagnostic and therapeutic challenges, particularly in the context of severe malnutrition and advanced malignancy.

The rarity of ACS, particularly its secondary form associated with malignancies, poses diagnostic difficulties. Most documented cases are idiopathic or linked to tuberculosis in endemic regions, with reports predominantly involving men in their 30s through 50s.[1,3], In this case, the patient presented with nausea, vomiting, weight loss, and ascites of unknown etiology – symptoms that, while suggestive of gastrointestinal and hepatobiliary disorders, contributed to diagnostic delays because of their nonspecific nature. Although nausea and vomiting are consistent with obstructive symptoms, the presence of weight loss and ascites further complicated the clinical picture, obscuring the underlying cause.[4]

Imaging plays a critical role in diagnosing ACS. CT scans are often instrumental, revealing characteristic findings such as peritoneal thickening, encapsulation of bowel loops, and cocooning.[7] However, in this case, the diagnostic utility of CT imaging was hindered by the patient's severe cachexia, which led to a lack of intra-abdominal fat. Intra-abdominal fat typically acts as a natural contrast against surrounding tissues, making it easier to delineate anatomical landmarks and structures.[8,9] This allows for highlighting abnormalities such as peritoneal thickening or encapsulation with greater ease. The absence of fat in this patient resulted in reduced contrast, making it difficult to differentiate structures and interpret findings accurately, which were largely misinterpreted as nonspecific edema and inflammation. This limitation highlights the importance of recognizing scenarios in which advanced imaging, such as PET-CT, should be expedited to overcome such diagnostic barriers. PET-CT, with its ability to detect FDG-avid peritoneal thickening, was pivotal in this case, raising strong suspicion for peritoneal malignancy causing abdominal cocooning and guiding subsequent diagnostic steps.[10]

Future cases involving malnourished patients with similar clinical presentations may benefit from the early use of advanced imaging to prevent prolonged diagnostic delays. Endoscopic evaluations, including EGD and EUS, are valuable diagnostic tools for gastrointestinal pathologies but were challenging in this case. Multiple EUS attempts encountered resistance to insufflation, initially attributed to edema and inflammation but retrospectively

linked to fibrosis and cocooning characteristic of ACS. This limitation delayed definitive diagnosis by hindering timely tissue sampling, emphasizing the need for alternative strategies or adjunct diagnostic modalities when standard techniques are unrevealing. Signet ring cell adenocarcinoma, the underlying malignancy in this case, is an aggressive cancer often associated with peritoneal dissemination.[11] The absence of an identifiable primary tumor in this patient, coupled with widespread metastatic involvement, compounded diagnostic complexity.

Cases like this highlight the critical role of histopathological confirmation, which ultimately establishes the diagnosis through peritoneal biopsy. This case contributes to the growing literature on secondary ACS, offering valuable lessons for clinical practice. First, while advanced imaging modalities are resource-intensive, their early implementation in malnourished patients with diagnostic uncertainty may expedite diagnosis and improve outcomes. Second, the multidisciplinary collaboration in this case—spanning gastroenterology, radiology, oncology, and pathology—illustrates the importance of a coordinated approach in managing complex cases. Lastly, recognizing the demographic patterns of ACS, with a predilection for males in their 30s to 50s, can guide clinical suspicion and diagnostic prioritization in similar cases.[1,3] By demonstrating the unique diagnostic challenges posed by ACS secondary to an aggressive malignancy, this case is an example of improving diagnostic strategies in rare conditions. The expedited use of advanced imaging and a multidisciplinary approach may lead to earlier diagnosis and potentially life-saving interventions in future cases.

CONCLUSIONS

This case highlights the importance of maintaining a high index of suspicion for abdominal cocoon syndrome (ACS) in patients with nonspecific symptoms and refractory presentations. In complex cases in which cachexia limits conventional diagnostic tools, the early use of advanced imaging modalities such as PET-CT and a multidisciplinary approach can expedite diagnosis and improve outcomes. Recognizing ACS in similar clinical contexts may guide timely and effective management in future cases.

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