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CASE REPORT | SMALL BOWEL

Poorly Cohesive Carcinoma of the Nonampullary Small Bowel: A Rare Cause of Recurrent Small Bowel Obstruction

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ABSTRACT

Poorly cohesive carcinoma (PCC) is an uncommon neoplasm characterized by tumorous cells exhibiting a lack of adhesion. PCC has been reported rarely in the small intestine other than at the ampulla of Vater. We present a 40-year-old man with recurrent abdominal pain and small bowel obstruction. Imaging revealed an abnormal appearing distal small bowel, with only nonspecific mucosal changes discovered on antegrade and retrograde enteroscopy. On subsequent diagnostic laparoscopy, an ileal mass was found and resected with histopathology showing PCC with signet ring formation. This is an aggressive cancer with a worse prognosis than other small bowel adenocarcinomas.

KEYWORDS: poorly cohesive carcinoma; small bowel adenocarcinoma; small bowel obstruction

INTRODUCTION

Primary malignancy of the small bowel is rare, accounting for less than 5% of gastrointestinal (GI) cancers. Small bowel adenocarcinoma (SBA) makes up 30%–40% of these cases and is often diagnosed in the context of intestinal obstruction or GI bleeding.[1,2] Poorly cohesive carcinomas (PCCs) are neoplasms characterized by dyshesive spread of tumor cells. While PCC is well documented in the stomach, gallbladder, and ampulla of Vater, there are limited data regarding nonampullary small bowel PCCs. This case describes a patient with unexplained recurrent small bowel obstruction (SBO) ultimately diagnosed with SBA with features compatible with PCC after diagnostic laparoscopy and small bowel resection of an ileal mass.

CASE REPORT

A 40-year-old man with no significant medical or surgical history presented to University Medical Center of El Paso with recurrent abdominal pain and SBO unrelieved by ibuprofen. Computerized tomography (CT) imaging suggested high-grade SBO due to focal areas of bowel wall thickening in the distal ileum. No masses or lymph nodes were identified. Serum and stool inflammatory markers, stool cultures, and tissue transglutaminase were unremarkable. The patient underwent retrograde single balloon enteroscopy revealing normal mucosa in the distal ileum. Random biopsy was unremarkable. The SBO resolved spontaneously, and the patient was discharged.

However, soon after, he developed recurrent abdominal pain and nausea. CT with intravenous contrast showed submucosal fatty infiltration of the third part of the duodenum, suspicious for Crohn's disease. CT enterography revealed persistent small bowel wall thickening in the distal ileum (Figures 1 and 2). Antegrade push enteroscopy with a pediatric colonoscope showed subtle mucosal

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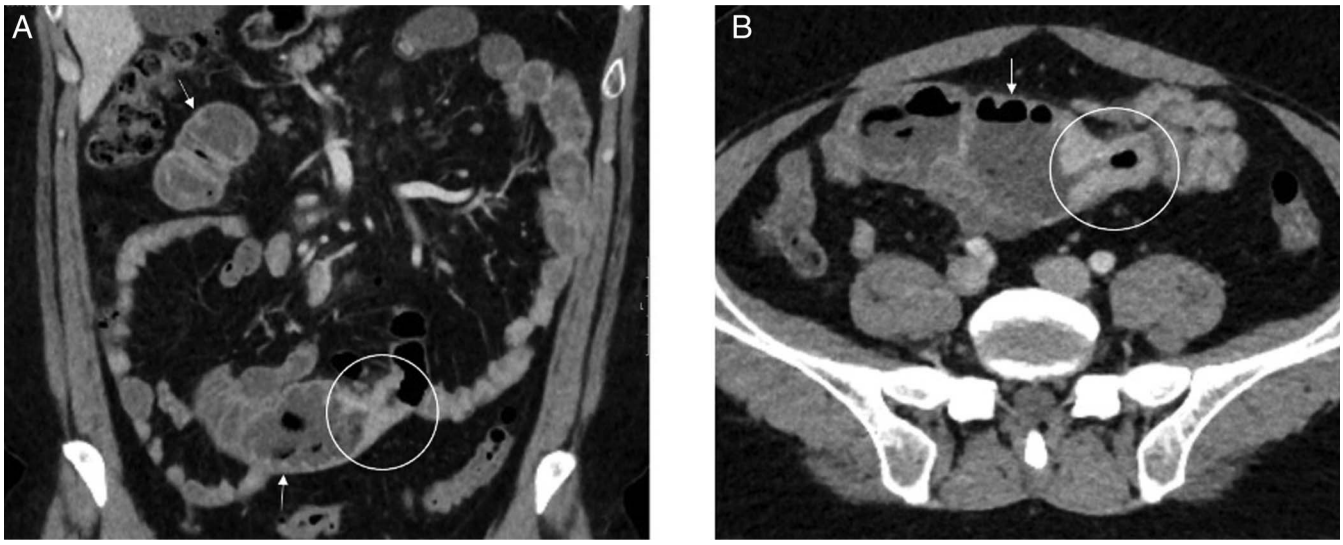

Figure 1. Coronal (A) and axial (B) computed tomography images of the abdomen with intravenous contrast. There are multiple dilated fluid-filled loops of proximal small bowel consistent with a small bowel obstruction (white arrows). Obstruction is secondary to a short segment of circumferential thickening in the ileum associated with luminal narrowing and corresponding to pathologically proven poorly cohesive carcinoma (circle).

erythema in the distal duodenum with a normal ampulla. Duodenal biopsies revealed focal active cryptitis and rare neutrophils in the lamina propria with normal appearing villi. A second retrograde balloon-assisted enteroscopy revealed subtle erythematous changes in parts of the ileum. Maximal insertion was approximately 210 cm from the ileocecal valve, and a tattoo was placed. Pathology of the ileum was normal, while random colon biopsy revealed lymphocytic colitis.

Small bowel capsule endoscopy was not performed due to concern for capsule retention from recurrent SBO. A multidisciplinary discussion followed the discordant findings. Diagnostic laparoscopy revealed an ileal mass with significant desmoplastic effect proximal to the previously placed submucosal tattoo. A hand-assisted laparoscopic resection of a 40-cm segment of small intestine and its mesentery was performed. Pathology showed poorly differentiated adenocarcinoma with features compatible with PCC with signet ring formation (Figures 3 and 5). The tumor was staged as pT4 and pN2, with 9 of 20 lymph nodes positive for PCC. One year later, the patient continues to follow with his oncologist, remains on chemotherapy, and has had 1 hospitalization for small bowel obstruction.

DISCUSSION

We present a case of recurrent SBO caused by a rare type of SBA. Symptoms of SBO classically include abdominal pain, nausea, emesis, abdominal distension, and constipation.[3,4] Differential diagnoses include postoperative ileus, narcotic bowel, pseudo-

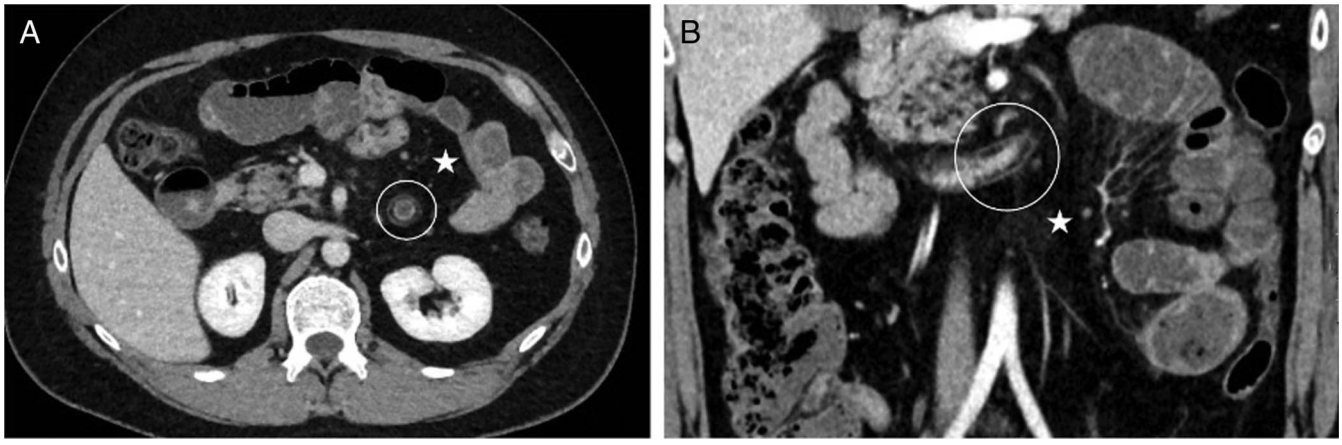

Figure 2. Axial (A) and coronal (B) computed tomography images of the abdomen and pelvis. There is fatty infiltration of the submucosal layer of the small bowel (circle) which appears as low density similar to the surrounding omental fat (star), interposed between the thin-enhancing serosal and mucosal layers. This is referred to as the fat halo sign and is seen as a sequela of chronic bowel inflammation.

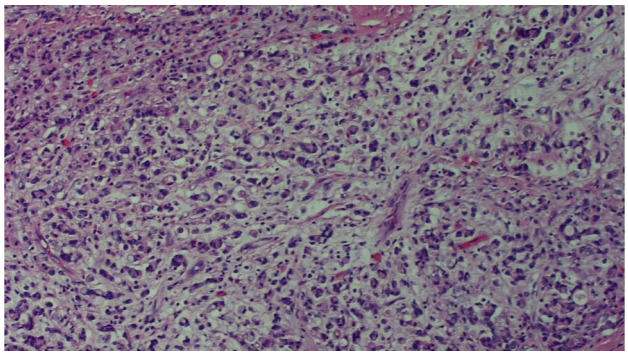

Figure 3. 20× magnification. On lower magnification, this shows a poorly differentiated malignant neoplasm as noted by loss of glandular architecture, pleomorphic nuclei, and an infiltrative pattern.

obstruction, mesenteric ischemia, and large bowel obstruction.[5] Guidelines recommend diagnostic evaluation to distinguish obstruction from ileus and to determine its etiology.[4] While the most common cause of SBO is mechanical, caused by postoperative adhesions causing extrinsic compression, SBO can also result from small bowel disease such as Crohn's disease, nonsteroidal anti-inflammatory drug enteropathy, radiotherapy, or small bowel malignancy.[3]

Initial workup for suspected SBO includes obtaining plain film radiographs.[3,4] If inconclusive, cross-sectional imaging, such as abdominal/pelvic CT with intravenous and oral contrast is recommended.[3,4] In this case, CT enterography identified a high-grade SBO with nonspecific focal wall thickening in the distal small bowel.

It was initially suspected that Crohn's disease was the cause of SBO. While capsule endoscopy is standard for small bowel endoscopic assessment, there was concern for capsule retention.[3] Therefore, the patient underwent antegrade and retrograde single balloon-assisted enteroscopy, which yielded nonspecific pathology results and inconclusive findings to explain symptoms. In Crohn's disease, one would expect to find notable endoscopic and histologic findings of the mucosa,[6] which our patient did not have despite suggestive CT findings. Furthermore, normal villi in the duodenum and negative tissue transglutaminase made celiac disease less likely. The lymphocytic colitis seen on the patient's colon biopsies was unclear, as the patient had no diarrheal symptoms, possibly indicating sequelae of nonsteroidal anti-inflammatory drug-associated injury.[7]

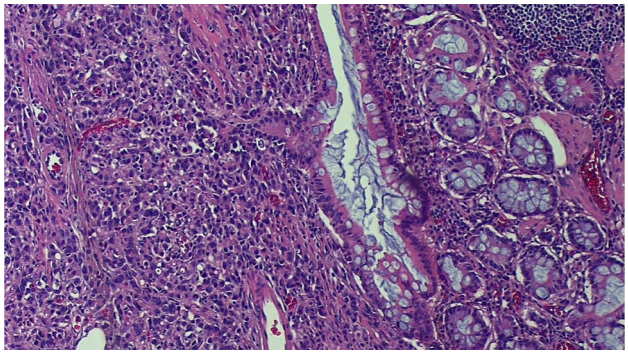

Figure 4. 20× magnification. This malignant neoplasm appears to have a dyshesive cell infiltrative pattern as noted by the spacing in between tumor cells and infiltration into surrounding tissues.

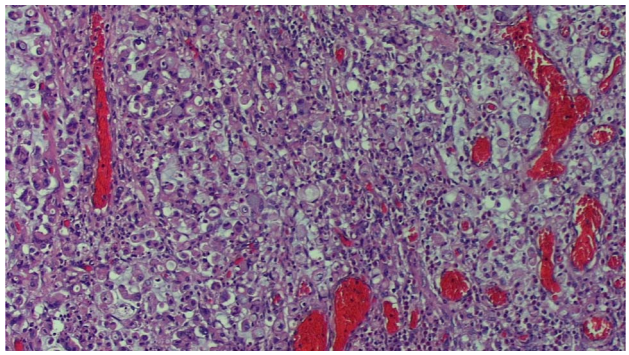

Figure 5. 40× magnification. On higher magnification, this malignant neoplasm shows dyshesive cells in which some have a signet ring cytology. These features support a poorly differentiated adenocarcinoma, described as a poorly cohesive carcinoma.

In 2019, the World Health Organization identified nonampullary PCC as a distinct subtype of SBA with unique clinical and pathologic characteristics and a worse prognosis compared with other SBAs. PCCs were initially described only in the stomach, gallbladder, and ampulla of Vater. The College of American Pathologists recognized nonampullary PCC, with or without signet ring cells, as a distinct histologic type in their 2021 protocol for examining small bowel carcinomas.[8] Studies have indicated that histologic subtypes of SBAs may offer prognostic significance, with noncohesive histology having a worse prognosis than cohesive histotypes.[8] PCCs exhibit a more aggressive nature, often spreading extensively into the peritoneal cavity.[8] They are typically diagnosed at a younger age and strongly associated with Crohn's disease with higher rates of lymphovascular and perineural invasion.[8] PCCs are often detected in the setting of SBO or GI bleeding. Patients with PCC generally have worse cancer-specific survival compared with those with nonspecified SBAs.[8] In addition, PCC is associated with hereditary factors such as Lynch syndrome or familial adenomatous polyposis, as well as immune-mediated diseases such as celiac disease or Crohn's disease.[8]

Treatment of SBA is similar to that of colorectal cancer, involving en bloc resection of the tumor. To date, however, there is no standard adjuvant regimen proven to be effective against SBA following resection.[9–11] For locoregional or resectable disease, wide resection with lymphadenectomy is the recommended treatment. Postoperative complications include adhesions that increase the risk of further bowel obstruction, which may explain our patient's recurrent obstructive symptoms. For metastatic SBA, systemic chemotherapy has been the primary treatment.[11,12]

In conclusion, we present a rare case of a small bowel PCC causing SBO. It is crucial to consider SBA in the differential of

SBO, especially in the absence of IBD or mechanical causes. The evaluation for Crohn's disease is important, given the differences in therapy and its association with PCC.

DISCLOSURES

Author contributions: B. Lee: wrote the manuscript. J. Guzman: consulted on manuscript development. O. Padilla: revised the manuscript and provided pathology images and descriptions. S. Laks: revised manuscript and provided radiology images and descriptions. N. Ng: revised the manuscript and provided details of surgery. SE Elhanafi: revised the manuscript and provided details of endoscopy. R. Badillo: revised the manuscript and consulted on manuscript development. M. Zuckerman: principal investigator, manuscript supervisor, and article guarantor.

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Informed consent was obtained for this case report.

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