


CASE REPORT

Very rare presentation of perforated appendiceal adenocarcinoma within a prolapsed colostomy

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Abstract

Appendiceal adenocarcinoma is rare with variable presentations. Perforation of the appendix within prolapsed ostomy can have peritoneal spread for which intraperitoneal chemotherapy should be considered. Surveillance is essential for these patients.

KEYWORDS

appendiceal cancer, colostomy, HIPEC, perforated appendix

1 | INTRODUCTION

Appendiceal mucinous adenocarcinoma is a rare disease in clinical practice. We present a case report of a very rare presentation of a perforated appendiceal cancer within an incarcerated prolapsed colostomy, in a patient with a history of abdominoperineal resection for rectal cancer, complicated by delayed peritoneal metastasis.

Primary neoplasms of the appendix are rare with an incidence of approximately 1 case per 100 000 people per year.¹ Appendiceal tumors are broadly classified into neuroendocrine tumors and adenocarcinomas. Adenocarcinomas make 60% of all the cases and are subdivided into colonic, mucinous, and goblet cell-type adenocarcinoma.² They are typically found incidentally after surgery for appendicitis or on CT imaging for another indication. Due to its rarity, optimal management of appendiceal adenocarcinoma remains to be delineated. Due to the paucity of data, their treatment mostly corresponds to colon cancer treatment. Data suggest early diagnosis and resection with negative margins provide patients with the best prognosis.² Given the mean age of presentation at 65 years,³ many affected patients have undergone screening colonoscopy prior to their surgical diagnosis which in theory

could identify some of these patients during the asymptomatic phase. However, current colonoscopes are unable to examine the appendiceal lumen and are therefore effective only at diagnosing pathology that involves the orifice.⁴

2 | CASE HISTORY

A 69-year-old African American man presented to the surgical oncology outpatient clinic with a complaint of a palpable kidney bean size nodule within prolapsed stoma. He denied associated pain, weight loss, or bleeding from the ostomy. Past surgical history included abdominoperineal resection (APR) for stage III rectal cancer in 1996 with adjuvant chemoradiation. Post-oncologic treatment course was complicated by recurrent stomal prolapses requiring multiple colonic resections, parastomal hernia repair, and reciting the colostomy in the right lower quadrant. A ~12 cm prolapsed non-reducible colostomy was noted on examination in the right lower quadrant with a soft, mobile 2.5 × 1 cm nodule palpated within the lateral wall of the prolapsed colon. There were no superficial mucosal lesions noted. The patient was scheduled for a CT scan of the abdomen and pelvis with

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contrast, carcinoembryonic antigen (CEA) testing, and close follow-up at the clinic.

2.1 | Investigations and treatment

The patient presented to the emergency department about a week after with acute worsening of his ostomy prolapse doubling in size from his clinic visit associated with edema and pain without evidence of bowel ischemia. Attempt to reduce stoma at the bedside with granulated sugar was unsuccessful. CT scans of the abdomen/pelvis with IV contrast showed prolapsed ostomy containing fat and vessels approximately 20 cm in length appearing markedly inflamed with subcutaneous soft tissue and mesenteric edema (Figure 1).

The patient was taken to the OR for an urgent revision of ostomy and completion colectomy. The patient's entire abdomen was prepped and draped in the supine position including the incarcerated/prolapsed ostomy (Figure 2). Electrocautery was used to separate the mucocutaneous junction in a circumferential manner. Dissection was continued down to the fascia. The entire colon was found to be intussuscepted into the exteriorized segment of the remnant colon with the terminal ileum forming the intussusceptum. There was edema and congestion of the outer and inner bowel walls. The decision was made to resect all exteriorized bowel and to revise the colostomy to an end ileostomy. There was no suspicion at this point of an appendiceal pathology, and the working diagnosis was worsening incarceration causing strangulation resulting in an obstructed venous flow. The patient had an uncomplicated recovery from surgery and was discharged on post-op day two with instructions to return to the clinic for follow-up.

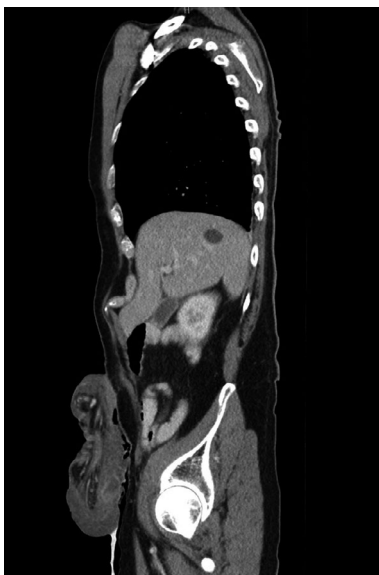


FIGURE 1 Prolapsed right lower quadrant colostomy containing fat and vessels approximately 20 cm in length with no evidence of bowel obstruction. 2.0 cm cyst in the right hepatic lobe is also seen

Gross pathologic examination of the resected specimen showed the cecum and ascending colon were contained within the prolapsed stoma. There was a full-thickness perforation identified 2.7 cm from the appendiceal orifice with surrounding yellow, purulent exudate and hemorrhagic, indurated colonic fat (Figure 3). Microscopic examination showed moderately differentiated invasive mucinous adenocarcinoma of the appendix with metastatic carcinoma identified in 1 of 33 lymph nodes. Pathologic stage classification using AJCC 8th edition guidelines was pT3pN1a. The patient was discussed in multidisciplinary tumor conference, and a recommendation was made for consideration of HIPEC (hyperthermic intraperitoneal chemotherapy), which the patient preferred not to proceed with, and chose systemic chemotherapy (capecitabine/oxaliplatin). He was followed with serial (3 monthly) clinical examination, serum CEA testing and CT chest, abdomen, and pelvis for surveillance.

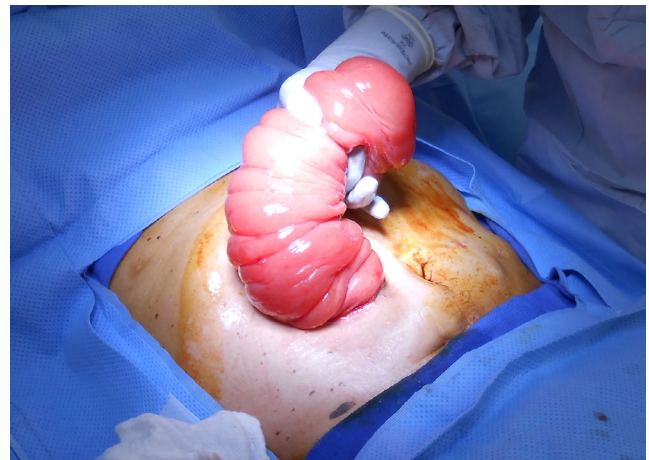


FIGURE 2 Demonstrates acutely incarcerated prolapsed colostomy

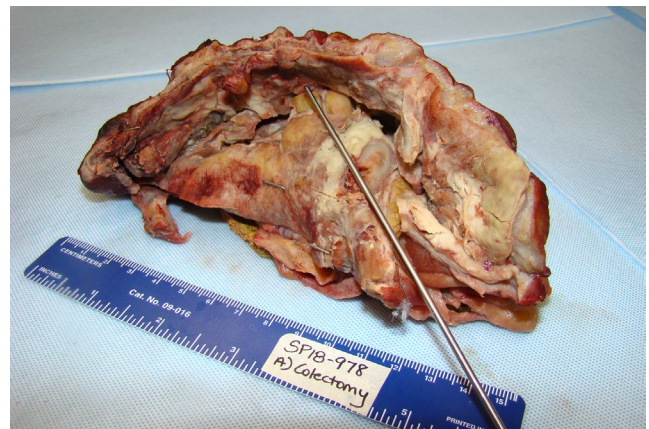


FIGURE 3 Resected right hemicolectomy specimen with suppurative perforated appendicitis with needle through the perforated appendiceal cancer

2.2 | Follow-up

Surveillance CT scan at 12 months after surgery showed interval development of a soft tissue nodule in the right mid-abdomen close to the ostomy concerning for peritoneal carcinomatosis. PET scan confirmed hypermetabolic activity and suspicion for recurrent appendiceal mucinous carcinoma with peritoneal implants. The site of suspicious recurrence underwent CT-guided biopsy, which was consistent with recurrent appendiceal cancer. Diagnostic laparoscopy with possible conversion to laparotomy and HIPEC therapy was offered and discussed with the patient.

3 | DISCUSSION

Mucinous adenocarcinoma of the appendix (MAA) is a rare disease with variable presentations. Patients most often learn of their diagnosis after pathologic analysis of an appendectomy specimen or CT imaging for another indication. Surgical resection is the cornerstone of the treatment. The decision of whether to perform appendectomy vs right hemicolectomy in non-ruptured MAA should be informed by the grade of the tumor.² Low-grade tumors necessitate appendectomy en bloc with special care being taken to preserve the cyst architecture to prevent seeding of the peritoneum. High-grade tumors should be treated with right hemicolectomy with ≥ 12 lymph nodes needed for accurate staging. In addition to its role in staging, removal of a higher number of lymph nodes may provide some curative value.⁵

In cases of perforated MAA, diagnostic laparoscopy should be considered for the evaluation of peritoneal deposits which if positive, complete cytoreduction and HIPEC therapy should be offered in the operative candidate patients.² That includes resection of residual disease plus right hemicolectomy, omentectomy, RLQ peritonectomy, bilateral oophorectomy (in female patients), and hyperthermic intraperitoneal chemotherapy (HIPEC) with mitomycin C (MMC), capecitabine, or oxaliplatin.⁶ When HIPEC is not performed at the time of surgery due to lack of preceding or concurrent diagnosis, an alternative is early postoperative intraperitoneal chemotherapy (EPIC) with floxuridine, MMC, or 5-FU.² Contraindication to curative-intent surgery includes metastatic involvement of extraperitoneal structures such as the liver or retroperitoneal lymph nodes.

Adjuvant chemotherapy can provide overall survival benefit for stage II patients with high-risk features and stage III patients whereas for those with stage IV disease, there is no evidence for significant overall survival benefit.⁷ Close multidisciplinary follow-up is important to monitor postoperative recovery, tolerance of adjuvant chemotherapy regimens, and continued remission of disease as evidenced by

laboratory testing for tumor markers and/or radiologic imaging. Surveillance with history and physical and cross-sectional imaging every 3 months for 2 years and every 6 months for the following 3 years is suggested for these patients.⁸

3.1 | Could the disease have been diagnosed earlier?

The plan for CT scan on his clinic presentation was an attempt at evaluating the nodule in the lateral wall of the prolapsed stoma though it would be hard to pin it to an appendiceal pathology given it is rare to suspect an appendiceal malignancy in a prolapsed stoma. Our patient underwent a screening colonoscopy through his ostomy 6 months before his presentation to the clinic, which was reported as a normal residual colon. The patient's appendiceal pathology was not visualized during the colonoscopy. Some studies have reported the possibility of diagnosing appendiceal cancers during colonoscopy, which would be quite difficult in this case given that the appendix was part of the intussusceptum. Endoscopic changes observed with appendiceal adenocarcinoma include appendiceal lesions, intussusception, and polyps.⁹ However, screening for MAA with colonoscopy is very limited by the inability to visualize beyond the appendiceal orifice, and thus, colonoscopic evaluation provides low yield in diagnosing appendiceal carcinoma.

3.2 | Was a diagnostic exploration and HIPEC indicated after the diagnosis of MAA was identified?

We hypothesized that even though the perforated appendiceal cancer was in the intussuscepted portion the colon in the prolapsed stoma, there was still a risk of peritoneal contamination with possible malignant cells given that anatomically there is no barrier separating this intussuscepted peritoneal lining and the peritoneal lining of the abdomen. Given that this was a surprise pathologic finding, the patient was postoperatively offered re-exploration and HIPEC, which he declined. Surveillance imaging was suggestive of peritoneal metastasis. Options were discussed with the patient including additional systemic chemotherapy vs HIPEC therapy, and he opted for chemotherapy.

4 | CONCLUSION

We report on the first case of ruptured MAA within a prolapsed stoma. Literature review of English medical/ surgical literature did not reveal prior reports. Management options depend on the tumor grade, stage, and perforation status.

Surgical options include appendectomy, right hemicolectomy, and cytoreductive surgery with HIPEC therapy. MAA presentation is often late in the disease process necessitating the use of adjuvant systemic chemotherapy. Multidisciplinary teams approach care and surveillance follow-up are essential for management for patients with similar presentations.

CONFLICT OF INTEREST

None.

AUTHOR CONTRIBUTIONS

GG, YA, LF, and SS: were responsible for data collection, study design, and writing the manuscript. SS: supervised the project.

ETHICAL APPROVAL

Institutional approval was obtained. No identifiable patient information was presented in this report.

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