Appendiceal Mucocele Concurrent With Intestinal Malrotation

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Background	Appendiceal mucoceles and intestinal malrotations are extremely rare conditions that are often found incidentally during the fifth and sixth decade of life. We report a case of concurrent appendiceal mucocele and intestinal malrotation with a review of the previously published literature.
Summary	A 58-year-old female was referred to our clinic because of an abdominal mass in left lower quadrant, detected incidentally during colonoscopy. Abdominal computerized tomography (CT) scan showed an abnormally distended appendix filled with low-density material protruding into the base of the cecum. The patient also had complete intestinal malrotation with the colon and appendix located on the left side of the abdomen. The appendix was removed laparoscopically, with an uneventful recovery. The final diagnosis was a low-grade appendiceal mucinous neoplasm with diffuse involvement of the entire appendix.
Conclusion	Careful operative planning is necessary in cases of concurrent intestinal malrotation and appendiceal mucocele due to the altered anatomy. The mucocele should be removed intact to avoid seeding of the peritoneal cavity with neoplastic mucin-secreting epithelial cells. Postoperative surveillance consists of annual abdominal CT scans and monitoring of tumor markers carcinoembryiogenic antigen and CA19-9.
Keywords	Appendiceal mucocele, low-grade appendiceal mucinous neoplasm, intestinal malrotation

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The authors have no conflicts of interest to disclose.

ABBREVIATIONS:
CT: Computerized tomography
LAMN: Low-grade appendiceal mucinous neoplasm
CEA: Carcinoembryiogenic antigen
USG: Ultrasonography
CRP: C-reactive protein

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Case Description

Appendiceal mucoceles are often found incidentally during the fifth and sixth decade of life and most often in women.[2,3] They can be classified as neoplastic and non-neoplastic based on their pathologic characteristics.[4] Non-neoplastic subtypes are *mucosal hyperplasia* and *simple retention cysts*, whereas neoplastic subtypes are mucinous cystadenoma and *mucinous cystadenocarcinoma*.[5] The term "low-grade appendiceal mucinous neoplasm" (LAMN) replaced "mucinous cystadenoma" in the recent literature to describe the slow growth and spread of the tumors to surrounding structures without destruction.[6]

Intestinal malrotation is often asymptomatic. Its incidence is estimated to be approximately 1:500 in the United States.[7] In general, there is little clinical significance, unless malrotation is complicated with other diseases.[8,9]

Appendiceal mucoceles and intestinal malrotations are independently unusual conditions, but are exceedingly rare together.[1,2] There are only three previous cases of concurrent appendiceal mucocele and intestinal malrotation reported in the literature (Table 1).[8-10]

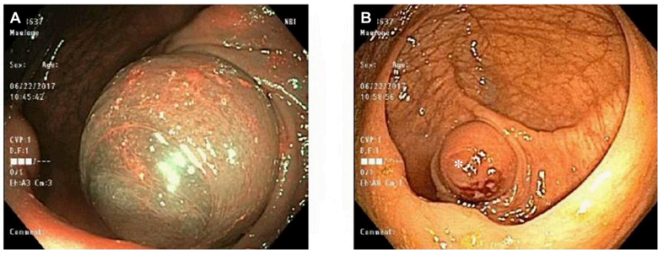

Figure 1. Colonoscopy imaging of the lesion. (A) The close up view of the appendiceal lesion in narrow band imaging (NBI) mode. (B) Appendiceal lesion protruding into cecum in white field imaging mode. White asterix marks the lesion.

with no evidence of malignancy (Figures 1 A & B). Other laboratory tests showed no abnormal findings. The patient had no history of change in bowel habits or weight loss, and denied abdominal pain. On physical exam, her abdomen was positive only for a non-tender mass felt in the left lower quadrant.

We obtained an abdominal computerized tomography (CT) scan, which was remarkable for malrotation of the bowel. The entire colon was located on the left side of the abdomen, resulting in a left upper quadrant cecum. The appendix was distended with low density material and was protruding into the base of the cecum. The most dilated portion of the appendix measured 2.0 cm, and no periappendiceal inflammatory changes were noted (Figures 2 A & B).

Authors	Age/Sex	Location	Size (cm)	Symptoms	Follow up (months)	Recurrence
Sato, 2001[9]	76, F	Left lower quadrant	4x4	Abdominal pain	23	No
Kawashima, 2001[10]	51, M	Lower abdomen	6	Abdominal pain	Not available	Not available
Yap, 2016[8]	65, M	Left upper quadrant	17x8x6	Abdominal pain	12	No

Table 1. Reported cases of appendiceal mucocele concomitant with intestinal malrotation.

The management of a patient with concurrent appendiceal mucocele and intestinal malrotation can be challenging due to the lack of clinical experience and established treatment algorithms. Here, we report a case of concurrent appendiceal mucocele and intestinal malrotation, with management algorithms based on the previously published literature and our own experience.

A 58-year-old female with a non-tender abdominal mass in the left lower quadrant was referred to our clinic following colonoscopy. The patient had a history of colon cancer in two of her cousins and had a prior screening colonoscopy in 2011 that was notable for the extractions of a 10 mm tubular adenoma. She was recommended for a 2–3 year follow-up. In follow-up colonoscopy, a 45 mm polypoid lesion was noted in the cecum. The lesion was described as fixed and firm with no bleeding and the biopsy of the peripheral lesion disclosed superficial columnar epithelium

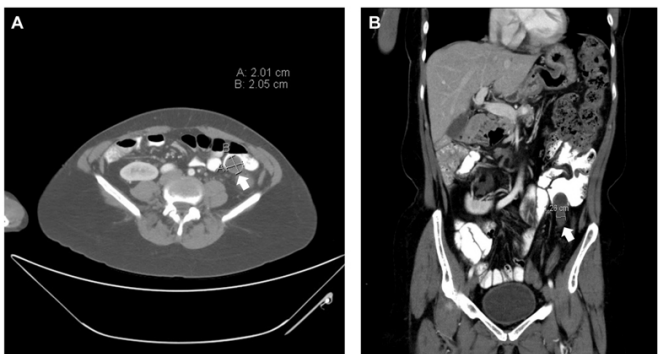

Figure 2. Abdominal CT depicting a left-sided appendiceal lesion protruding into the cecum. (A) Transverse view of the appendiceal lesion with a maximum diameter of 2.0 cm. (B) Coronal view of appendiceal lesion with invasion into cecum and visible intestinal malrotation.

These findings were consistent with appendiceal mucocele confirming the initial colonoscopic diagnosis.

The patient was taken to the operating room for laparoscopic appendectomy. The port placement was a mirror image for that of a typical laparoscopic appendectomy, with a 10mm umbilical port, and 5mm ports in the lower midline and right epigastric regions. Intraoperatively, we observed that the appendix was in the left upper quadrant. It was distended at its mid-portion, but there was no adjacent tissue involvement. There were no Ladd bands. We dissected and removed the appendix and mass carefully to avoid cellular seeding and mucus spillage into the peritoneum. The specimen was extracted with a retrieval bag, and the ports were closed. The patient was kept overnight, tolerated a regular diet, and was discharged home on the morning of postoperative day 1.

The pathologic exam revealed an LAMN measuring 7.1 cm, in essence running the length of the appendix. Acellular mucin was observed invading into the muscularis layer, but without invasion of the serosa and clean surgical margins (Figure 3A). The epithelium was described as adenomatous and pseudostratified with hyperchromatic nuclei and apical mucin (Figure 3B).

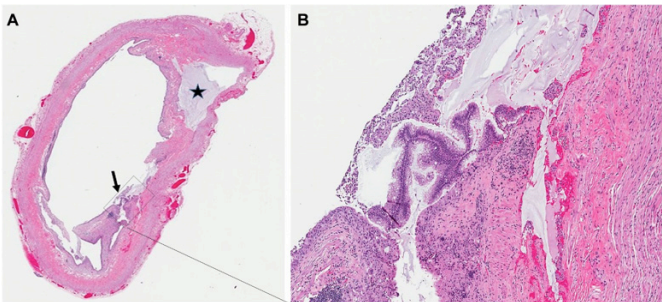

Figure 3. Microscopic cross-section images of the surgical specimen stained with hematoxylin eosin (HE) stain. (A) Low power HE image showing cross section of appendix with dilated lumen containing mucin and focal adenomatous epithelium (arrow) confined by the basement membrane. Acellular mucin is also seen dissecting into the muscularis layer (star) but not penetrating the serosa. (B) Medium power HE image showing a close-up of adenomatous epithelium. The nuclei are hyperchromatic and psuedostratified, and the cells have apical mucin.

The patient's further postoperative recovery was uncomplicated. She was seen in clinic two weeks later, with healed incisions, tolerating a regular diet, and no pain.

Discussion

The appropriate treatment of patients with asymptomatic malrotation is controversial. Traditional teaching favors surgical intervention in all patients with a radiographic diagnosis of malrotation.[11] However, the incidence of intestinal complications in asymptomatic adults with intestinal malrotation is low.[12,13] The most dreaded complication of intestinal malrotation is midgut volvulus, but the risk of volvulus declines significantly after infancy. Most adults require surgical treatment for chronic gastrointestinal symptoms rather than volvulus.[14,15] There is also growing evidence suggesting that many adults with intestinal malrotation remain asymptomatic throughout life[12].

The diagnosis of appendiceal mucocele can be challenging if it is accompanied with intestinal malrotation. The patients can be asymptomatic, as in our case, or may present with abdominal pain in left lower quadrant[9] or left upper quadrant[8] depending on the degree of intestinal malrotation. The diagnostic work up should start with a careful physical exam. Plain radiographs, CT scan, ultrasonography (USG), colonoscopy and barium enema can provide a relatively accurate presumptive diagnosis and exclude other etiologies.[16] Plain radiographs may show characteristic calcifications in a punctate or curvilinear pattern at the appendix site.[16] Additionally, distended bowel loops and air fluid levels can be observed in case of ongoing intestinal obstruction.[10] However, plain radiographs provide little information on the intestinal anatomy.

Abdominal CT is the procedure of choice, as it can both demonstrate the intestinal anatomy and provide accurate staging information about the tumor. Appendiceal mucoceles are typically seen as a well-encapsulated round or tubular cystic mass with low attenuation adjacent to the cecum.[16] A mucinous cystadenocarcinoma should be suspected when there is soft-tissue thickening and wall irregularity without increasing appendiceal wall thickness.[17] Colonoscopic findings are usually similar to the findings depicted in our case; a glossy and rounded mass protruding from the appendiceal orifice into the cecum can be seen.[18] The mass may be firm or soft in consistency, and may exhibit a central indentation, known as the "cushion sign."[19] A "volcano sign" is described as the appendiceal orifice seen in the center of the mucocele protruding into the cecal wall.[20] In addition to imaging, the CEA, CA19-9 and C-reactive protein (CRP) levels might be increased in patients with neoplastic appendiceal mucocele.[8-10]

The presence of abdominal pain in the initial presentation suggests a more complicated clinical course such as the rupture of the tumor or small bowel obstruction.[8-10] A complicated clinical course may also correlate with the increased malignancy. For asymptomatic and uncomplicated cases, a laparoscopic approach can be utilized; however, an open approach may be preferred for the complicated cases.

The extent of resection of appendiceal mucoceles largely depends on the degree of adjacent tissue involvement seen on preoperative imaging.[21] For non-neoplastic lesions, simple excision with clear margins is usually curative, whereas appendiceal cystadenocarcinoma may require right hemicolectomy and lymph node sampling due to the malignant nature of the disease.[9] If the appendix is not ruptured, the tumor should be handled gently intraoperatively in order to avoid rupture and the spillage of mucocele content. The spillage from mucocele may lead to pseudomyxoma peritonei[10] that decreases the survival of the patients significantly (reported 5-year survival 23 percent).[6,8,22] This is especially important in cases with LAMN, since these patients have a 100 percent five-year survival rate if the tumor can be removed without spillage.[22]
Postoperative surveillance for an appendiceal mucocele will depend on intraoperative and pathologic findings. For benign neoplastic lesions, such as LAMN, patients should be followed up with yearly abdominal CT scans for 5–10 years.[21,23] Carcinoembryiogenic antigen (CEA) and CA19-9 levels can also be used to detect possible recurrence of neoplastic lesions.[10,24]

The case presented here is interesting due to the fact that the patient had no symptoms or surrounding organ invasion despite the fairly large size of the tumor. The preoperative imaging also underestimated the size of the lesion; however, we feel that the patient underwent successful laparoscopic management.

In conclusion, careful operative planning is necessary in cases of appendiceal mucocele with intestinal malrotation based on the clinical picture and abdominal imaging. Unruptured, asymptomatic mucoceles should be removed intact. If the mucocele is already ruptured or if the patient is symptomatic, then the tumor is most likely a neoplastic one, and a wider oncologic excision is required.

Lessons Learned

Appendiceal mucoceles accompanied with intestinal malrotation can present with a variety of symptoms depending on the level of the malrotation and the histological subtype of the tumor. An abdominal CT is helpful for both documenting the intestinal malrotation and the extent of the tumor invasion. Tumors that have not ruptured can be safely removed laparoscopically. If the histologic subtype is non-neoplastic or an LAMN, excision with clear margins is curative. Concomitant intestinal malrotation in adults can be managed nonoperatively in the absence of symptoms. If conservative management is pursued, patients should be educated on the signs and symptoms of intestinal malrotation complications that require surgical evaluation.

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