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Intestinal obstruction caused by low-grade appendiceal mucinous neoplasm: A case report and review of the literature

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ABSTRACT

INTRODUCTION: Intestinal obstructions due to appendiceal pathology are rare. Obstructions caused by low-grade appendiceal mucinous neoplasms (LAMNs) are rarer still.

PRESENTATION OF CASE: A 79-year-old woman was referred to our hospital for mechanical small intestinal obstruction. The patient had undergone prior left oophorectomy via a lower abdominal incision. Physical examination revealed abdominal distension and slight tenderness. Laboratory analysis was unremarkable. Contrast enhanced computed tomography (CT) demonstrated dilation of an obstructed small intestinal loop without evidence of strangulation. There appeared to be a low density mass measuring 3.0×1.5 cm with a potential twist in the mesentery near the transition point in the small intestine. The patient was diagnosed with mechanical small intestinal obstruction and was treated conservatively with nasogastric tube decompression for one week. After no clinical improvement, we elected to surgically explore her. Intraoperative findings revealed that the ileum was compressed by the appendix, which had a cystic mass on its tip. There was no evidence of intestinal ischemia. Laparotomy appendectomy alone was performed. Pathology revealed a LAMN measuring 3.0×1.5 cm. She remains disease-free with 18 months of postoperative follow-up.

DISCUSSION: Intestinal obstruction due to external compression by an appendiceal mass is rare, and is often difficult to preoperatively diagnose with CT.

CONCLUSIONS: The etiology of small intestinal obstruction with a transition point in the right lower quadrant of abdomen includes an appendiceal mass on the broad differential for this common problem.

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1. Introduction

Low-grade appendiceal mucinous neoplasms (LAMNs) are characterized by low-grade cytologic atypia and the absence of destructive invasion. These tumors possess the potential for peritoneal spread, giving rise to pseudomyxoma peritonei [1]. They are regarded as low-grade adenocarcinomas according to the 2010 World Health Organization classification [2]. Surgical resection is the standard treatment. The presence of acellular mucin in the right lower quadrant confers a low risk of recurrence, whereas mucin

with mucinous epithelial cells in the right lower quadrant confers a high risk of recurrence. Assessment of surgical margins is an integral part of pathologic tumor evaluation, and the evaluation of LAMNs includes reporting on the status of the margin [3]. However, which specific surgical procedure is most appropriate remains controversial for patients with LAMN. The causes of intestinal obstruction are numerous, and include the presence of a fibrous cord, torsion, internal hernia, or prior abdominal surgery causing adhesions [4]. Intestinal obstructions caused by external compression from the appendix are rare, and those caused by appendiceal masses are rarer still, with only a few cases reported on in the English literature. We report herein a case of intestinal obstruction in the setting of a LAMN. The present work has been reported in accordance to the SCARE criteria [5].

Abbreviations: LAMN, low-grade appendiceal mucinous neoplasm; CT, computed tomography.

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Fig. 1. Contrast enhanced CT showed dilation of an obstructed small intestinal loop. There is a low density mass and potentially twisted mesentery near the transition point (transition point: white arrow, mass: white arrow head).

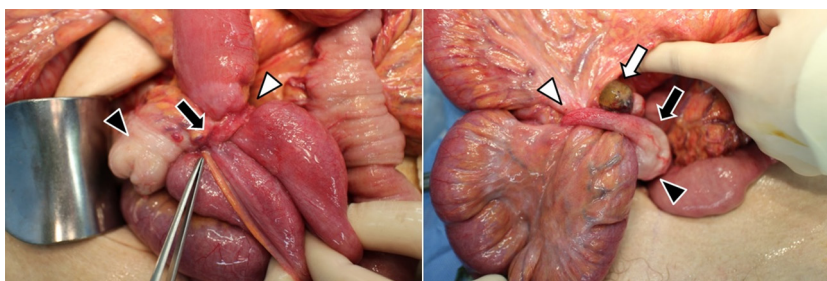


Fig. 2. Operative findings revealed that the ileum was externally compressed by the appendix, which had a cystic mass at its tip (appendix: white arrow head; root of appendix: black arrow; tip of appendix: white arrow; cecum: black arrow head).

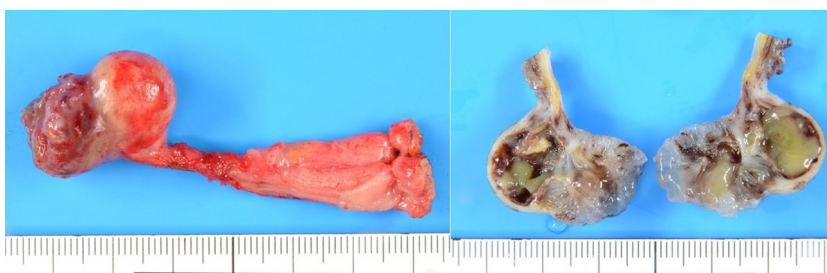


Fig. 3. The resected specimen demonstrated mucus extruding from the mass.

2. Presentation of case

A 79-year-old woman was referred to our hospital for mechanical small intestinal obstruction. The patient had a prior history of left oophorectomy via a lower abdominal incision. Physical examination revealed abdominal distension and slight tenderness. Laboratory analysis revealed the following: hemoglobin, 10.1 g/dL; white blood cell count, $4.10 \times 10^3/\mu\text{L}$; platelets, $21.9 \times 10^4/\mu\text{L}$; serum total protein, 5.7 g/dL; serum albumin, 3.1 g/dL; total bilirubin, 0.5 mg/dL; aspartate aminotransferase, 24 IU/L; alanine aminotransferase, 31 IU/L; lactic acid dehydrogenase, 205 IU/L. Contrast enhanced computed tomography (CT) demonstrated dilation of an obstructed small intestinal loop without strangulation. There was an open loop obstruction and a low density mass measuring 3.0×1.5 cm with a potential twist in the mesentery near the transition point (Fig. 1). The patient was diagnosed with mechanical small intestinal obstruction and was treated conservatively with nasogastric tube decompression for one week. Physical examination and laboratory analysis did not reveal the exacerbation for the term. After no clinical improvement, we elected to surgically explore her. Intraoperative findings revealed that the ileum

was compressed by the appendix which had a cystic mass on its tip (Fig. 2). There was no evidence of intestinal ischemia and root of the appendix was intact. Because of these reasons, Laparotomy appendectomy was performed alone. The resected specimen showed mucus extruding from a cystic appendiceal mass measuring 3.0×1.5 cm and the length of wrap around the ileum was 4.0 cm (Fig. 3). Histopathological examination showed no epithelium covering the cyst and no malignant cells in the mucus. The patient was thus diagnosed with a mechanical small intestinal obstruction due to external compression from a LAMN. The patient's postoperative course was unremarkable and she developed no complications. She was discharged on postoperative day 10. She remained disease-free during 18 months of postoperative follow-up.

3. Discussion

LAMNs are characterized by low-grade cytologic atypia and the absence of destructive invasion. These tumors possess the potential for peritoneal spread giving rise to pseudomyxoma peritonei and can eventually lead to death, even in the absence of overtly malignant cytologic features [1]. Thus, they are regarded as low-grade

Table 1
Reported cases of intestinal obstruction secondary to a mucocele of the appendix.

No.	Author	Year	Age	Sex	Past history of abdominal surgery	Mucocele of the appendix of preoperative diagnosis	Surgery	Size of the mucocele of the appendix(cm)	Pathological diagnosis
1	Mourad [8]	1999	57	M	No	Yes	Ileocecal resection	12 × 7.0 × 7.0	Mucinous cystadenoma
2	Chong [9]	2001	65	F	No	No	Ileocecal resection	8.0 × 5.0	Mucinous cystadenoma
4	Garg [10]	2011	70	M	Unknown	No	Appendectomy	7.5	Mucinous cystadenoma
3	Miyakua [11]	2010	89	F	No	No	Ileocecal resection	9.0 × 3.0	Mucous containing cystic appendix without mucin-producing, neoplastic epithelial cells
5	Zaharie [12]	2012	39	F	No	No	Appendectomy Ileal segmental resection	Unknown	Mucinous cystadenoma
6	Opreanu [13]	2013	51	F	Cholecystectomy Hysterectomy Inguinal hernia repair	No	Ileocecal resection	3.0 × 2.5 × 1.4	Benign mucocele
7	Our case	2018	79	F	left oophorocystectomy	No	Appendectomy	3.0 × 1.5	LAMN

Table 2
The demographic characteristics and clinicopathological factors of patients with intestinal obstruction secondary to a mucocele of the appendix.

factors	No. (n = 7)
Age (mean, range)	64 (39–89)
Gender (male / female)	2 / 5
operation history (yes / no / unknown)	2 / 4 / 1
diagnosis as mucocele of the appendix preoperatively (yes / no)	0 / 7
Intestinal obstruction (strangulation / mechanical)	6 / 1
surgical procedure (appendectomy / ileocecal resection)	3 / 8 / 4
maximum tumor diameter (median) (cm)	7.8 (3.0–12)
pathological diagnosis (mucinous adenoma / mucinous adenocarcinoma / LAMN)	6 / 0 / 1

adenocarcinomas according to the 2010 World Health Organization classification [2].

The causes of intestinal obstruction are numerous, and include the presence of a fibrous cord, torsion, internal hernia, or prior abdominal surgery causing adhesions [4]. Intestinal obstructions caused by external compression from the appendix are rare, and those caused by appendiceal masses are rarer still. Bhandari et al. [6] classified intestinal obstruction caused by the appendix into four groups: 1) adynamic, 2) mechanical (without strangulation), 3) strangulation of the intestine, 4) intestinal obstruction due to mesenteric ischemia. In the present case, the appendix itself was wrapped around the ileum without intestinal ischemic change. It is thus thought that the present case corresponds to a mechanical obstruction without strangulation.

To the best of our knowledge, intestinal obstructions caused by external compression by a twisted appendix or a mucocele are extremely rare, and only a few cases have been reported on; only 16 [7] and 6 cases [8–13] have been reported in the English literature, respectively. Seven cases, including the present case of a mucocele of the appendix, were reviewed in the present study (Table 1). The demographic characteristics and clinicopathological factors of these patients are summarized in Table 2. There were 2 (29%) males and 5 (71%) females with a mean age of 65 (range: 39–89) years. Of these 7 patients, 2 patients (29%) had prior abdominal operations. There were no patients who had diagnosis of mucocele of the appendix preoperatively. Six patients (86%) had strangulation and

1 patient (14%) had a mechanical obstruction. Two patients (29%) underwent appendectomy alone and 5 patients (71%) underwent ileocecal resection. There was median maximum tumor diameter of 7.8 (range: 3–12) cm. It is often difficult to preoperatively differentiate between benign or malignant mucoceles of the appendix and appendicitis. A previous investigator has reported that mucoceles of benign origin are rarely larger than 2 cm [14]. All 7 patients had a maximum tumor diameter larger than 2 cm. Only the present case was found to be a LAMN on pathologic examination. It is often difficult to diagnose the cause of small intestinal obstruction with CT preoperatively. Therefore, a high index of suspicion for an appendiceal cause is necessary in those patients with obstruction and a transition point in the right lower quadrant of the abdomen.

Surgical resection is the standard treatment and is aimed at ameliorating symptoms that may result in numerous complications, and also minimizing the potential for pseudomyxoma peritonei. However, which particular surgical procedure (appendectomy alone, cecectomy, ileocecal resection, or right hemicolectomy in addition to regional lymph node dissection) is most appropriate remains controversial for patients with LAMNs. In the present case, the patient underwent appendectomy alone, as only the tip of the appendix was involved and it was thought correctly that a negative resection margin could be achieved. The patient would have had to undergo a two-stage ileocecal resection in addition to regional lymph node dissection in the case of a pathological diagnosis of mucinous adenocarcinoma or positive resection margin of the LAMN.

Tumor involvement of a surgical resection margin is considered to be an indication for additional treatment or additional surgery, though a previous investigator has reported that patients with LAMN confined to the appendix and without extrusion of mucin or mucinous neoplasia beyond the appendiceal serosa, or without involvement of the appendectomy margin by either neoplastic epithelium or acellular mucin, do not have disease recurrence or peritoneal dissemination [3]. In addition, another previous investigator has reported that cecectomy is the standard treatment strategy for a mucocele of the appendix. This is based upon review of 411 Japanese cases [15]. It is thought that standard procedure for LAMN should be a cecectomy as the rested root of appendix has the potential for malignant transformation and progression to pseudomyxoma peritonei. Others have espoused a two-stage ileocecal resection in addition to regional lymph node dissection if patients

have a pathological diagnosis of mucinous adenocarcinoma or positive resection margin.

4. Conclusions

We report herein a case of intestinal obstruction caused by external compression from a LAMN. It is often difficult to preoperatively diagnose the cause of small intestinal obstruction with CT. Therefore, it is necessary to take into consideration that appendixes themselves can be causes of small intestinal obstructions if there is a transition point in the right lower quadrant of abdomen.

In addition, surgical resection is aimed at removing the symptoms that may result in numerous complications and put the patient at risk for malignant transformation. However, there are no guidelines for the optimal management of LAMN. The surgical procedure for patients with LAMN should be selective cecectomy for negative margins. However, a larger number of patients with LAMN with long-term follow-up data are required to establish optimal, evidence-based treatment.

Conflict of interest

The authors declare that they have no Conflicts of interest.

Sources of funding

The authors declare that this study was not funded externally.

Ethical approval

The study such as this case report was exempted from ethical approval by the Institutional Review Board of Hiroshima City Asa Citizens Hospital.

Consent

When obtaining informed consent for surgical procedures, general consent for publication and presentation was obtained from the patients.

Author contributions

TK drafted the manuscript. TK and TK reviewed and edited the manuscript. MK, KO, MY, AN, YA and MM participated in the care of the patients. MK provided the histopathological examination and diagnosis. TK, JH, HM, and NH participated in critical revision of the manuscript. All authors read and approved the final manuscript.

Registration of research studies

This is case report not research study.

Guarantor

Toshihiko Kohashi.

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