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A case of low-grade appendiceal mucinous neoplasm with infection treated with ileal resection

Akifumi Okada, Shoichiro Mukai, Toshihiro Nishida, Toshikatsu Fukuda, Hideki Ohdan

ABSTRACT

Introduction: Low-grade appendiceal mucinous neoplasm (LAMN) is a rare tumor found in only 0.27% of appendectomy specimens. It is often misdiagnosed as appendicitis, and patients can remain asymptomatic for years, delaying treatment. This report describes a rare case of LAMN with infection that required surgery 14 years after its initial detection without treatment.

Case Report: A 54-year-old woman presented with right lower abdominal pain and fever for five days. Fourteen years earlier, a computed tomography (CT) scan revealed a cystic lesion in the appendix; however, no treatment was initiated. On admission, her temperature was 38.5 °C, with mild tenderness. Blood tests showed elevated C-reactive protein (CRP) levels, and CT imaging revealed a 9 cm inflamed cystic lesion. Antibiotics were ineffective, and the patient underwent an ileal resection. Laparotomic resection was selected due to the patient's prior history of surgery, suspected adhesions secondary to inflammation, and the considerable tumor size. The tumor was removed intact. Histopathological analysis confirmed a LAMN with associated infection. The postoperative ileus was treated with decompression, and the patient was discharged on day 20.

Conclusion: When a LAMN is removed without perforation, the risk of recurrence is minimal, and the prognosis is excellent. Therefore, early diagnosis and resection are essential. Careful surgical planning is critical to prevent tumor rupture and complications such as pseudomyxoma peritonei (PMP). This case emphasizes the importance of timely intervention.

Keywords: Appendix, Ileal resection, Infectious disease, Low-grade appendiceal mucinous neoplasm

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INTRODUCTION

The 5th edition of the 2019 World Health Organization (WHO) classifies appendiceal epithelial lesions into low-grade appendiceal mucinous neoplasms (LAMN), high-grade appendiceal mucinous neoplasms (HAMN), and adenocarcinomas (mucinous adenocarcinoma, mucinous adenocarcinoma with signet ring cells, signet ring cell carcinoma, and non-mucinous adenocarcinoma colorectal type) [1]. Appendiceal mucinous neoplasms (LAMN and HAMN) are extremely rare, accounting for only 0.27% of appendectomy specimens [2]. Diagnosing LAMN is difficult as it can be misdiagnosed

as appendicitis [3]. Previous studies have reported cases that were asymptomatic for long periods [4]. We treated a case of LAMN with an infection that required surgery after 14 years of no treatment. Given the rarity of this presentation, we report this case alongside a review of the relevant literature.

CASE REPORT

A 54-year-old woman with a five-day history of right abdominal pain and fever was admitted to our hospital. The patient presented with a solitary congenital kidney. She underwent surgery at approximately 20 years of age for a left ovarian cyst and bicornuate uterus, and bypass surgery at 28 years of age for adhesive bowel obstruction due to adhesions in the sigmoid colon. At 46 years, she underwent surgery for postoperative adhesive bowel obstruction, and at 52 years, she underwent a left axillary lipoma resection. Computed tomography (CT) performed 14 years previously indicated a cystic lesion that appeared contiguous with the appendix. Magnesium oxide, epinastine, tizanidine, diazepam, loxoprofen, sodium esomeprazole, and rizatriptan were administered. On arrival at our hospital, the patient's temperature was 38.5 °C; all other vitals were stable. Mild tenderness was noted in the right lower abdomen; however, no signs of peritoneal irritation were observed. An initial hematological investigation revealed a white blood cell count of 7,370/mm³ with 79.0% neutrophils and elevated C-reactive protein (CRP) levels (10.95 mg/dL); there were no other major outliers. Abdominal CT revealed a cystic lesion, measuring 9 cm in diameter, extending from the tip of the appendix, accompanied by surrounding inflammation (Figure 1). Magnetic resonance imaging (MRI) revealed uniformity inside the cyst, which was attributed to fluid retention (Figure 2). Based on these findings, we diagnosed appendiceal mucinous neoplasm with infection and administered intravenous cefmetazole sodium. The patient was scheduled for surgery after the inflammation subsided; however, she did not respond to treatment and underwent ileocecal resection with D2 lymphadenectomy because of the possibility of malignancy on the 8th day of the hospitalization. Given the patient's prior surgical history, the anticipated presence of adhesions to adjacent organs secondary to inflammation, and the substantial size of the tumor, a laparotomic approach was selected for resection. The tumor was located on the outer dorsal surface of the cecum and firmly attached to the cecum but was removed without damage. Histopathological examination revealed a LAMN with infection (Figure 3). The cyst wall was fibrotic and thickened with clusters of chronic inflammatory cells. The cyst lumen was almost devoid of epithelial cells. The remaining epithelium comprised a single layer of adenomatous columnar epithelium. A few atypical cells were observed. Mucus cytology revealed clusters of adenocytes with irregular nuclei.

The patient's inflammatory response improved postoperatively. She experienced postoperative ileus; however, her symptoms improved with decompression through a nasogastric tube, and she was discharged on postoperative day 20.

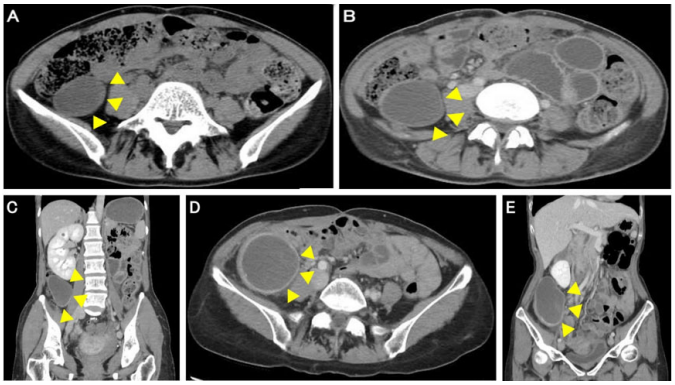

Figure 1: Abdominal CT images. Arrowhead: Tumor. (A) Abdominal CT from 2010 showing a cystic lesion, 4 cm in diameter, in the right lower abdomen. (B, C) Abdominal CT from 2016. The cyst had increased in size to 5 cm and was continuous with the appendix. (D, E) Abdominal CT on admission revealed a cyst approximately 9 cm in diameter with elevated surrounding fatty tissue density.

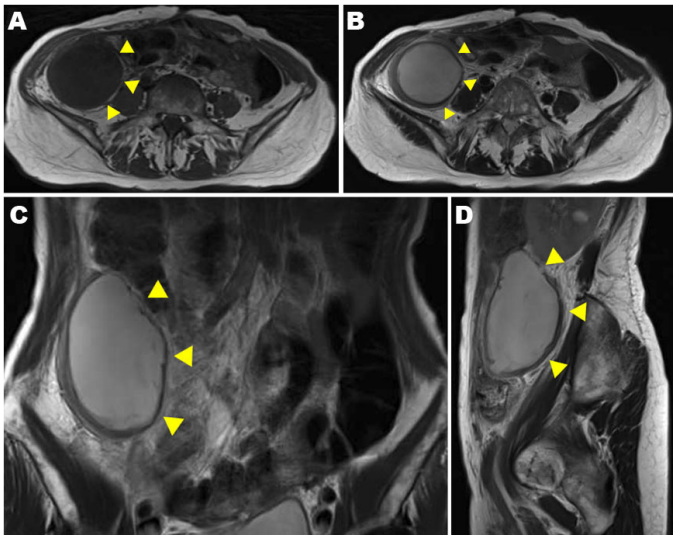

Figure 2: MRI images. Arrowhead: Tumor. MRI images on admission (A: T1-weighted; B–D: T2-weighted). The intracystic area showed low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

DISCUSSION

The presence of low-grade cytologic atypia, together with one of the following, is defined as a LAMN: loss of the muscularis mucosa layer, submucosal fibrosis, pushing invasion, dissection with acellular mucin in the wall, undulating or stratified epithelial growth, appendiceal rupture, and the presence of extra-appendiceal mucin and/or cells. Mucinous neoplasms that display structural features similar to those of LAMN lack infiltrative invasion; however, cytologically high-

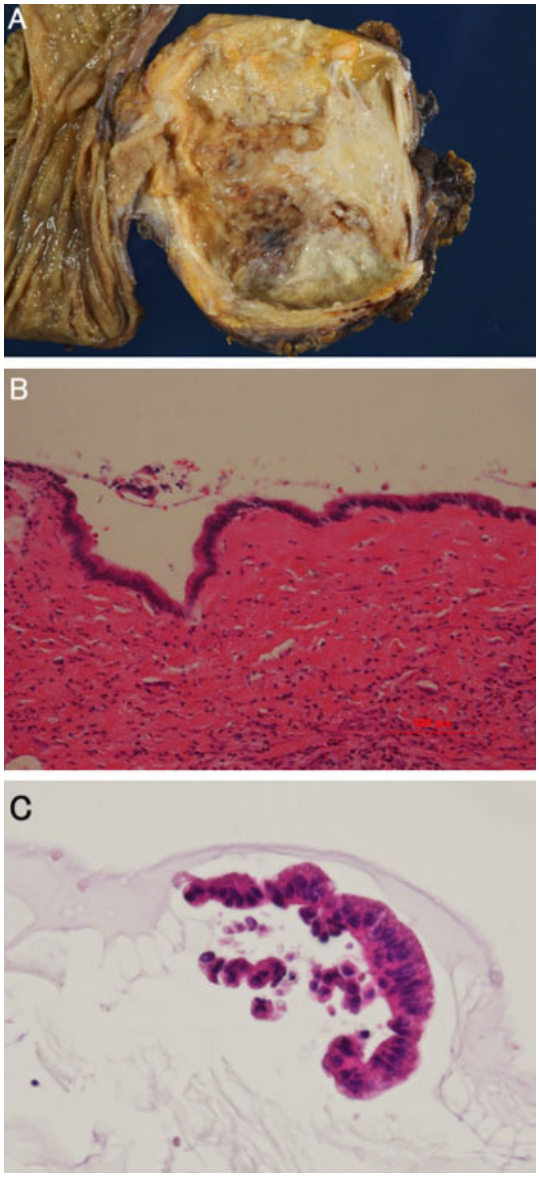

Figure 3: Histopathological images. (A) A unilocular cyst approximately 9 cm in length formed from the base of the appendix to the caudal side, and the lumen was covered with necrotic material and pus. (B) Hematoxylin-eosin stain. Most of the epithelial cells were shed, and the remaining epithelium consisted of a monolayer of adenomatous columnar epithelium with no atypia, suggestive of malignancy. (C) Hematoxylin-eosin stain. Mucus cytology revealed clusters of glandular cells. The nuclei of the glandular cells were small and irregular in shape.

grade atypia has also been identified as HAMN. Mucinous adenocarcinoma is used to describe mucinous tumors with infiltrative invasion [5]. Köhler et al. reported that the average age at diagnosis was 62 years, with 63% of patients being female, and the 5-year survival rate was approximately 80% [6]. An important complication of LAMN is pseudomyxoma peritonei (PMP), which is thought to be disseminated by the rupture of the tumor, causing mucus to disperse into the abdominal cavity [7]. In patients without perforation or PMP, appendectomy is performed; if mucus is disseminated into the abdominal cavity, hyperthermic intraperitoneal chemotherapy perfusion (HIPEC) is recommended in addition to tumor resection [8].

There is no consensus regarding the choice of technique (right hemicolectomy or appendectomy) or approach (laparoscopic or open surgery). With regard to the surgical technique, laparoscopic resection is preferable if the appendix can be removed without perforation. In recent years, there have been increasing reports of laparoscopic resection [9]. Lymph node dissection is not necessary due to the low probability of lymph node metastasis in LAMN [10]. However, a biopsy carries the risk of intra-abdominal dissemination, and a preoperative diagnosis is often not made. Dissection is required when the tumor is malignant, such as in mucinous adenocarcinoma of the appendix. Therefore, we selected a procedure similar to that for malignancy in consideration of the possibility of malignancy.

In this case, a cystic lesion was noted on CT 14 years prior and had been growing ever since; however, no surgical intervention was performed. Fortunately, there were no PMP and the tumor was removed without perforation. Because of the patient's previous surgical history, expected adhesion to the surrounding organs due to inflammation, and the large size of the tumor, we chose to remove it via a laparotomy. Although the prognosis of LAMN is generally good, rupture, torsion [11], and infection have been reported in some cases, as in the present case. In addition, it is difficult to obtain a definitive diagnosis before surgery, and early surgical intervention is desirable for diagnostic purposes. In such cases, the surgical plan should be carefully designed to avoid tumor damage.

The recurrence rate of LAMN was 2.7% [12]. Although there are no guidelines for postoperative surveillance, the most common surveillance method is imaging, and tumor markers are frequently used in combination [12]. Our hospital performs blood tests every six months and imaging tests every year for five years postoperatively. Furthermore, if R0 resection is performed for nonperforated LAMN, the risk of recurrence or postoperative PMP is extremely low, and surveillance may be omitted [13].

CONCLUSION

If LAMN can be resected without perforation, the risk of recurrence is low, and the prognosis is excellent; therefore, early resection should be considered. It is important to carefully plan surgery to avoid damaging the tumor.

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Author Contributions

Akifumi Okada – Conception of the work, Design of the work, Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Shoichiro Mukai – Conception of the work, Design of the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Toshihiro Nishida – Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.