

Diagnostic and Surgical Challenges in Low-Grade Appendiceal Mucinous Neoplasm Complicated by Acute Appendicitis: A Case Report

Running title: LAMN with Acute Appendicitis: A Case Report

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Abstract

Mucinous appendicular tumors (MAT) are rare entities and represent a subset of appendiceal neoplasms. Low-grade appendiceal mucinous neoplasm (LAMN) is characterized by mucin production and can be associated with serious complications such as appendiceal rupture and pseudomyxoma peritonei. Here, we present a case of LAMN complicated by acute appendicitis, emphasizing its diagnostic and surgical challenges. A 52-year-old male presented with acute colicky pain in the lower abdomen, worsening over three days. Imaging studies revealed findings compatible with an appendiceal tumor and mucinous accumulation. The patient underwent laparoscopic appendectomy, and histopathology confirmed LAMN with secondary acute appendicitis. The diagnosis of LAMN can be challenging due to its nonspecific symptoms, often mimicking acute appendicitis. Surgical intervention remains the cornerstone of treatment, with careful consideration of tumor margins and potential for mucinous spread. This case highlights the importance of early detection and appropriate surgical management to prevent complications. Early recognition and surgical management of LAMN are crucial to prevent long-term complications. Laparoscopic appendectomy can be an effective treatment option for localized tumors with minimal invasion.

Keywords: Gastroprotective agents • Low-grade appendiceal mucinous neoplasm • Mucinous tumor • Appendiceal tumor • Appendectomy • Pseudomyxoma peritonei.

Evidence Based Medicine Ranking: Level IV

Introduction

Mucinous appendicular tumors are rare, accounting for less than 1% of appendectomy specimens (1). Mucinous appendiceal tumors represent a rare but significant finding in appendectomy specimens. Low-grade appendiceal mucinous neoplasms (LAMNs) are a distinct subtype characterized by the production of mucin by epithelial cells, which can lead to serious complications such as peritoneal dissemination, commonly known as pseudomyxoma peritonei. Although uncommon, LAMNs account for up to 20% of all appendiceal neoplasms and often present clinically as acute appendicitis, complicating preoperative diagnosis (2).

Recent studies highlight that LAMNs typically present as circumferential proliferation of low-grade mucinous epithelium, often resulting in the obliteration of the mucosal and muscularis mucosa layers, making it difficult to distinguish between appendiceal adenomas and LAMNs. Surgical management of mucinous appendiceal tumors, including appendectomy and, in certain cases, right hemicolectomy, is determined by the neoplastic features of the tumor, such as the degree of cellular atypia and peritoneal involvement. For cases with extension beyond the appendix or positive surgical margins, more extensive surgery is recommended, and in selected cases, hyperthermic intraperitoneal chemotherapy (HIPEC) may be indicated (3).

In this article, we describe the case of a 52-year-old male with LAMN complicated by acute appendicitis, highlighting the challenges in diagnosis and treatment. This case highlights the crucial role of early intervention in reducing the morbidity and mortality associated with this condition. This report follows the SCARE criteria (4).

Case Presentation

A 52-year-old male, self-employed, presented to the emergency department with colicky pain in both lower quadrants of the abdomen. The pain had started three days prior and progressively localized to the right lower quadrant. The patient self-medicated with non-specified analgesics but found no relief. An ultrasound was ordered by a private physician, which showed signs of an appendiceal mass. On further examination at our institution, the patient had tenderness in the right iliac fossa with positive appendicular signs. He had no significant past medical history and denied alcohol or drug use. Imaging Studies: Ultrasound showed a fixed bowel loop with an absence of peristalsis, thickened appendiceal wall, and increased pericecal fat echogenicity. The presence of a heterogeneous mass with mucinous material and calcification suggested a mucocele of the appendix. The findings were compatible with LAMN (Image 1).



Figure 1. The image demonstrates a well-defined, heterogeneous mass with predominantly anechoic areas, measuring approximately 29.7 cc in volume. It is associated with an adjacent fluid collection and periappendiceal edema. A characteristic onion-skin pattern and calcifications are observed, findings consistent with an appendiceal mucocele.

Surgical Approach: In the operating room, following standard surgical safety protocols, the patient was placed in the supine position under general anesthesia. Aseptic and antiseptic measures were applied from the inframammary line to the upper third of the thighs, and sterile drapes were placed in a standardized manner. Access to the abdominal cavity was achieved using the Veress technique through a transumbilical incision, with pneumoperitoneum established up to 14 mmHg. A 12 mm trocar was placed transumbilically, and a camera was introduced through the same trocar for diagnostic laparoscopy. A plastron was found in the right iliac fossa, with a cecal appendix tumor adhered to the lateral abdominal wall (Image 2).

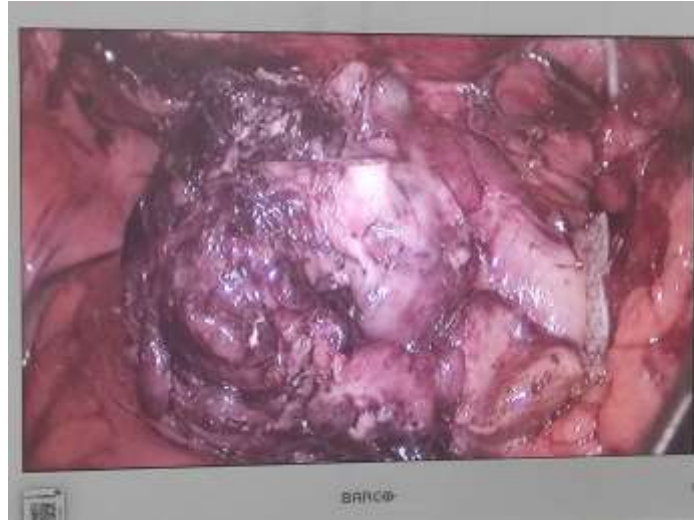


Figure 2. The laparoscopic image reveals an irregular tumoral mass originating from the cecal appendix, adherent to adjacent structures, primarily the lateral abdominal wall. The surface of the mass shows mucinous material, along with evidence of periappendiceal inflammation.

A 5 mm suprapubic trocar and a 12 mm left flank trocar were inserted. Blunt dissection was performed using a harmonic device to release the plastron, revealing an abscessed cecal appendix tumor, which drained mucinous material upon manipulation. The cecal appendix was perforated and adhered to the pericecal mass, with the base of the appendix and cecum showing no apparent abnormalities. Dissection of the mass was carried out, releasing adhesions from the abdominal wall until the base of the appendix was skeletonized. A linear stapler (purple cartridge) was used to transect the appendiceal base. The specimen was extracted through a 12 mm port in the left flank using an endobag (Image 3).



Figure 2. . The image shows a resected appendix with a heterogeneous mass, characterized by irregular mucinous material and areas of hemorrhage. The specimen displays features of mucinous adenocarcinoma, with evident cystic degeneration, fibrotic areas, and a gelatinous texture consistent with mucinous neoplasm.

The right iliac fossa was irrigated with saline solution and aspirated. Laparoscopic ports were removed under direct visualization, ensuring hemostasis. The aponeurosis of the 12 mm ports was closed with 2-0 Vicryl, and skin incisions were approximated with simple 2-0 nylon sutures. Surgical wounds were cleaned, and sterile gauze was applied. The procedure was completed without incidents or accidents, with a complete count of materials and sponges. Minimal bleeding was observed. The patient was transferred to the recovery room with residual anesthetic effects and stable vital signs within normal parameters.

Histopathology Report: The pathological analysis confirmed LAMN with acute appendicitis. The mucinous tumor showed low-grade epithelial proliferation with no signs of muscular invasion. The surgical margins were free of disease, and there was no evidence of disseminated mucinous disease.

Discussion

Low-grade appendiceal mucinous neoplasms (LAMNs) are rare and often discovered incidentally during surgical procedures for acute appendicitis or other abdominal pathologies. In the presented case, the patient displayed classic symptoms of acute appendicitis, which prompted further investigation and subsequent surgical intervention. The discovery of a LAMN highlights the diagnostic challenges, as this tumor often mimics more common appendiceal conditions such as appendicitis or mucocele formation

LAMNs are typically identified through imaging modalities such as ultrasound or CT scans, which may reveal mucin accumulation or cystic dilation of the appendix. In this case, ultrasonography revealed a well-defined, heterogeneous mass with anechoic regions and periappendiceal inflammation, which raised suspicion of a mucocele. Such findings are consistent with previously reported cases of LAMNs, where the diagnostic challenge often lies in distinguishing between benign and malignant lesions preoperatively (5).

Surgical management remains the primary treatment modality for LAMNs, with appendectomy being the standard approach in cases confined to the appendix. In more advanced cases where the tumor extends beyond the appendix or is associated with perforation, right hemicolectomy or cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (HIPEC) may be indicated (6). In the current case, the patient underwent laparoscopic appendectomy, which was sufficient due to the localized nature of the disease and absence of peritoneal dissemination. One of the critical concerns in managing LAMNs is the risk of perforation and subsequent development of pseudomyxoma peritonei (PMP). Several studies have highlighted that perforation, even without immediate clinical signs, can lead to the spread of mucinous material into the peritoneum, increasing the risk of PMP (7). In this case, while the appendix was perforated, no evidence of PMP was found intraoperatively, a fortunate outcome considering the potential for peritoneal seeding in such cases.

Histopathological evaluation confirmed the diagnosis of LAMN, characterized by low-grade epithelial proliferation and mucin production without muscular invasion. This finding aligns with the typical pathological features of LAMNs, which tend to follow a slow progression with a relatively favorable prognosis if managed appropriately (7,8). However, recurrence can occur in cases with positive surgical margins or extensive disease at the time of surgery. In a retrospective analysis of LAMN cases, Guner et al. found that patients with positive margins had a higher risk of recurrence and PMP development (7). In our case, negative margins were achieved, reducing the likelihood of recurrence.

The prognosis for patients with LAMN is generally good, especially in the absence of risk factors such as perforation or positive margins. However, long-term follow-up is critical, as recurrence can occur even years after the initial surgery. In cases of localized LAMN without peritoneal dissemination, appendectomy alone is often curative (9). For patients with perforation or acellular mucin on the serosa, more aggressive surgical approaches such as cytoreductive surgery with HIPEC may be warranted to prevent PMP. Regular imaging and clinical monitoring are essential to detect any signs of recurrence early.

Conclusion

Low-grade appendiceal mucinous neoplasms (LAMNs) represent a rare but clinically significant entity often masquerading as more common conditions such as acute appendicitis. The early detection and proper management of LAMNs are crucial to prevent complications such as perforation and pseudomyxoma peritonei (PMP). In the presented case, laparoscopic appendectomy was sufficient due to the localized nature of the disease, and the absence of peritoneal dissemination highlights the favorable prognosis when diagnosed early and managed appropriately.

Given the risk of recurrence in cases with positive margins or extensive disease, achieving clear surgical margins is essential for reducing recurrence. This case underscores the importance of careful histopathological evaluation and long-term follow-up to monitor for potential complications. As management strategies for LAMNs continue to evolve, this case supports the role of appendectomy as an effective treatment option in localized tumors with minimal invasion, while highlighting the need for more aggressive surgical approaches in cases with risk factors such as perforation or mucin dissemination.

Ultimately, this case contributes to the growing body of literature on the importance of individualized treatment strategies for LAMNs and reinforces the need for ongoing research to optimize long-term outcomes for these patients.

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