

## LOW-GRADE APPENDICEAL MUCINOUS NEOPLASM PRESENTING AS A LARGE RIGHT ILIAC FOSSA CYSTIC MASS MIMICKING COMPLICATED APPENDICITIS: A CASE REPORT AND REVIEW OF LITERATURE

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### Abstract

Low-grade appendiceal mucinous neoplasms (LAMNs) are rare appendiceal epithelial tumors characterized by mucin production and potential for peritoneal dissemination. Their clinical and radiologic presentation frequently mimics inflammatory conditions such as complicated appendicitis or appendiceal abscess, leading to diagnostic uncertainty. A 50-year-old male presented to our facility with right iliac fossa pain. Ultrasound demonstrated a cystic mass. Contrast-enhanced CT revealed an enlarged appendix (7.7 mm) with its tip encasing a thick-walled, rim-enhancing mass measuring  $10.35 \times 6.01 \times 6.45$  cm (~208.6 mL), associated with periappendiceal fat stranding and reactive lymphadenopathy. Findings were suggestive of longstanding appendicitis with abscess, though appendiceal mucocele was considered. Appendectomy was performed. Histology showed abundant acellular mucin dissecting the appendiceal wall with low-grade epithelial dysplasia, consistent with low-grade appendiceal mucinous neoplasm (LAMN). The patient had an uneventful recovery and was referred for oncologic surveillance. LAMN can closely mimic inflammatory appendiceal pathology and appendiceal abscess radiologically. A high index of suspicion, early surgical intervention and histopathological evaluation are essential to prevent progression and complications such as pseudomyxoma peritonei.

**Keywords:** Appendiceal mucinous neoplasm, LAMN, Appendiceal abscess, Pseudomyxoma peritonei, CT imaging

### Introduction

Appendiceal neoplasms are rare entities, accounting for approximately 1–2% of appendectomy specimens, with mucinous tumors forming a distinct pathological subgroup<sup>1</sup>. Among these, mucinous tumors represent a unique pathological spectrum ranging from benign mucoceles to malignant mucinous adenocarcinomas<sup>2</sup>. Low-grade appendiceal mucinous neoplasm (LAMN) is recognized as a unique entity characterized by low-grade

epithelial proliferation and cytologic atypia and abundant extracellular mucin production and accumulation and potential for peritoneal dissemination<sup>3,4</sup>.

Although histologically indolent, LAMNs possess significant clinical relevance due to their propensity to rupture and seed mucin-producing cells into the peritoneal cavity, resulting in peritoneal dissemination, leading to pseudomyxoma peritonei (PMP)—a rare but potentially fatal condition marked by progressive mucin accumulation within the peritoneal cavity<sup>3</sup>. PMP most commonly arises from rupture or leakage of appendiceal mucinous tumors, a condition associated with significant morbidity and mortality if untreated<sup>4</sup>. PMP is characterized by progressive intraperitoneal accumulation of mucin, leading to abdominal distension, bowel obstruction, and eventual organ compromise emphasizing the importance of early diagnosis of LAMN and its management<sup>4</sup>.

LAMNs most commonly occur in individuals in the fifth to sixth decades of life and often present with nonspecific or misleading clinical features<sup>5</sup>. Patients may present with: Right iliac fossa pain mimicking acute appendicitis, Palpable abdominal mass, Features suggestive of appendiceal abscess as seen in the patient in this case report. They may also be incidental findings during imaging or surgery.

Clinically, LAMNs are notorious for their heterogeneous and deceptive presentation. Patients may present with symptoms indistinguishable from acute appendicitis, chronic abdominal pain, or intra-abdominal abscess<sup>4</sup>. In many cases, diagnosis is incidental or made postoperatively due to nonspecific clinical and imaging findings.

This overlap in presentation frequently leads to preoperative misdiagnosis, with several studies reporting that LAMNs are often initially treated as complicated appendicitis<sup>6</sup>.

Radiological imaging plays a central role in evaluation. Computed tomography (CT) is considered the most sensitive and specific modality for assessing appendiceal pathology, capable of demonstrating cystic dilation, wall thickening, mural calcifications, and periappendiceal inflammation<sup>7</sup>. However, despite advances in imaging, distinguishing LAMN from inflammatory or infectious processes remains challenging.

Histopathological examination remains the gold standard for diagnosis, with defining features including low-grade epithelial dysplasia, mucin accumulation, and absence of infiltrative invasion<sup>3</sup>.

This case highlights the diagnostic dilemma posed by LAMN, particularly when it presents as a large cystic mass mimicking appendiceal abscess, and provides a comprehensive review of current literature.

## **Case Presentation**

A 50-year-old male presented with a history of progressively worsening right-sided abdominal pain localized to the right iliac fossa. The pain was of gradual onset, persistent,

and non-radiating, with no associated nausea, vomiting, altered bowel habits, or urinary symptoms. The patient also complained of feeling a “hard area” in his right lower abdomen. There was no history of prior abdominal surgery or known chronic medical illness.

On physical examination, the patient was hemodynamically stable. Abdominal examination revealed localized tenderness in the right iliac fossa without guarding, rigidity. Deep palpation showed the feeling of a mass in the region of the right iliac fossa. There were no signs of generalized peritonitis. Laboratory investigations demonstrated mild leukocytosis, while other routine hematological and biochemical parameters were within normal limits.

Initial ultrasonographic evaluation of the abdomen demonstrated a cystic mass in the right iliac fossa, raising suspicion for a localized intra-abdominal collection. To further characterize the lesion, a contrast-enhanced computed tomography (CT) scan of the abdomen was performed. CT imaging revealed an enlarged and inflamed appendix measuring approximately 7.7 mm in diameter. The appendiceal tip was observed to be closely related to, and partially encasing, a well-defined, thick-walled, rim-enhancing isodense collection located in the right iliac fossa. This collection measured approximately  $10.35 \times 6.01 \times 6.45$  cm (anteroposterior  $\times$  width  $\times$  height), with an estimated volume of 208.6 mL. Associated findings included mild periappendiceal fat stranding and inflammatory changes, as well as multiple periappendiceal and mesenteric lymph nodes of varying sizes, two of which measured approximately  $1.04 \times 0.87$  cm and  $1.01 \times 0.62$  cm, respectively. Importantly, there was clear delineation between the lesion and adjacent small bowel loops, and no evidence of an overt invasive appendiceal mass was identified. Based on these findings, a radiologic impression of longstanding appendicitis complicated by an extensive appendiceal abscess was made. However, the possibility of an appendiceal mucocele or mucinous neoplasm was also considered as a differential diagnosis.

Given the imaging findings and persistent symptoms, the patient underwent surgical intervention. Intraoperatively, the appendix was found to be enlarged and involved in a localized mass without evidence of rupture or dissemination of mucin within the peritoneal cavity. No gross peritoneal implants or ascites were identified. An appendectomy was performed without complication, taking care to avoid spillage of contents.

Histopathological examination of the resected specimen revealed a neoplastic lesion characterized by abundant acellular mucin dissecting through and splitting the muscularis propria of the appendix. The appendiceal mucosa demonstrated epithelial cells with low-grade to minimal dysplastic changes. Additionally, variable lymphocytic infiltrates were observed within the muscularis layer. There was no evidence of destructive stromal invasion. These findings were diagnostic of a low-grade appendiceal mucinous neoplasm (LAMN).

The postoperative course was uneventful. The patient recovered well and was subsequently referred to the oncology unit for further evaluation and surveillance, given the known risk of progression to pseudomyxoma peritonei in cases of appendiceal mucinous neoplasms.

## **Justification of the Study**

Low-grade appendiceal mucinous neoplasm (LAMN) is an uncommon tumor of the appendix that often presents with vague and nonspecific symptoms, making diagnosis difficult before surgery. In many cases, it closely resembles more common abdominal conditions such as acute appendicitis or appendiceal abscess, which can easily lead to misdiagnosis and delays in appropriate management.

This case was considered worthy of reporting because the patient presented with a large cystic mass in the right iliac fossa that radiologically suggested complicated appendicitis with abscess formation. However, histopathological examination later revealed a low-grade appendiceal mucinous neoplasm with abundant acellular mucin and low-grade dysplastic epithelial lining. The unusual presentation created significant diagnostic uncertainty and demonstrated how LAMN can mimic inflammatory abdominal disease both clinically and radiologically.

The study is also important because early recognition of LAMN has major implications for patient outcome. If not properly identified and managed, the lesion may progress to complications such as pseudomyxoma peritonei, a condition associated with significant morbidity due to intraperitoneal mucin spread. Highlighting this case may therefore improve awareness among surgeons, radiologists, and pathologists regarding the need to consider appendiceal mucinous neoplasms in patients with atypical right iliac fossa cystic lesions.

Furthermore, there are relatively few documented reports of LAMN from Nigeria and other parts of sub-Saharan Africa, particularly cases presenting predominantly as a large cystic abdominal mass. Reporting this case contributes to the limited regional literature and emphasizes the value of correlating radiologic findings with histopathologic diagnosis in achieving accurate patient management.

Finally, this report reinforces the importance of routine histopathological evaluation in presumed cases of complicated appendicitis, especially where imaging findings are atypical or suggestive of appendiceal mucocele or mucinous pathology.

## **Discussion and Review Of Literature**

Low-grade appendiceal mucinous neoplasm (LAMN) represents a rare but clinically significant subset of appendiceal epithelial tumors, accounting for approximately 0.2–0.3% of appendectomy specimens and less than 1% of gastrointestinal neoplasms<sup>8,9</sup>. Despite their low-grade cytologic features, these tumors exhibit a unique biological behavior characterized by progressive mucin production and the potential for peritoneal dissemination, distinguishing them from other appendiceal pathologies<sup>9,10</sup>.

One of the most challenging aspects of LAMN lies in its clinical and radiological mimicry of inflammatory conditions, particularly acute or complicated appendicitis. Several

contemporary studies have emphasized that the presentation of LAMN is often nonspecific, ranging from vague abdominal pain to findings indistinguishable from appendiceal abscess or phlegmon<sup>10,11</sup>. In fact, up to half of cases may initially be managed as acute appendicitis due to overlapping features such as right iliac fossa pain, leukocytosis, and localized inflammatory changes<sup>12</sup>. This diagnostic ambiguity was clearly demonstrated in the present case, where imaging findings strongly suggested a complicated appendiceal abscess.

Radiologically, computed tomography (CT) remains the cornerstone for evaluating appendiceal pathology. However, distinguishing LAMN from inflammatory conditions remains difficult, particularly in the presence of secondary inflammatory changes. Typical CT features of mucinous neoplasms include well-defined cystic dilatation of the appendix, low-attenuation lesions, and preservation of surrounding structures, whereas inflammatory abscesses tend to demonstrate more irregular borders and diffuse inflammatory changes<sup>13</sup>. In this case, the presence of a large, well-defined, rim-enhancing collection with preserved fat planes and lack of invasive features represented subtle clues toward a mucinous process, although these were overshadowed by associated inflammatory findings. Recent radiologic reviews emphasize that large encapsulated cystic masses with minimal invasive characteristics should raise suspicion for mucinous neoplasms, even in the presence of inflammation<sup>14,15</sup>.

Other pathologies can mimic LAMN as seen in some studies where LAMN was mistaken for ovarian mass. Ultrasound scan showed a right adnexal mass which could not be separated from the adjacent right ovary. An MRI was done pointed to an ovarian cystic mass, However, laparotomy and histology confirmed the diagnosis of LAMN<sup>16</sup>.

From a pathophysiological standpoint, LAMNs are characterized by expansile growth driven by mucin accumulation rather than infiltrative invasion. Progressive intraluminal mucin production leads to appendiceal dilation, wall thinning, and eventual dissection of mucin through the appendiceal layers<sup>10,17</sup>. The histopathological finding in this case—abundant acellular mucin dissecting the muscularis propria—is a hallmark feature and reflects this pressure-mediated expansion. Unlike invasive adenocarcinoma, LAMNs lack destructive stromal invasion and instead demonstrate “pushing” margins, a distinction that is critical for diagnosis and prognostication<sup>9,17</sup>.

At the molecular level, recent studies have identified recurrent mutations in KRAS and GNAS genes, supporting a neoplastic origin and offering insight into tumor behavior and progression<sup>18</sup>. These molecular alterations are thought to contribute to mucin overproduction and may explain the propensity for peritoneal dissemination seen in advanced cases.

The most clinically significant complication of LAMN is the development of pseudomyxoma peritonei (PMP), a condition characterized by progressive accumulation of mucin within the peritoneal cavity. PMP typically arises following rupture of the appendiceal wall, allowing dissemination of mucin-producing epithelial cells.<sup>3,10,19</sup> Once established, PMP leads to progressive abdominal distension, bowel obstruction, and ultimately significant morbidity if

untreated<sup>20-22</sup>. Importantly, the absence of rupture in the present case represents a favorable prognostic factor, as early-stage LAMN confined to the appendix is associated with excellent outcomes following complete surgical excision<sup>23</sup>.

The presence of periappendiceal and mesenteric lymphadenopathy in this case warrants consideration. While lymph node enlargement often raises suspicion for malignancy, in LAMNs it is typically reactive and related to local inflammation rather than metastatic spread<sup>11</sup>. This distinction is important in differentiating LAMN from more aggressive appendiceal adenocarcinomas, which are more likely to demonstrate nodal involvement and invasive features<sup>11</sup>.

Management of LAMN remains guided by disease extent. For lesions confined to the appendix without rupture or peritoneal involvement, appendectomy alone is generally sufficient, as performed in this case<sup>24</sup>. In contrast, more extensive disease may require right hemicolectomy or cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (HIPEC), particularly in cases complicated by PMP<sup>24</sup>. Current literature strongly emphasizes the importance of careful surgical handling to avoid intraoperative rupture, as this significantly influences long-term prognosis<sup>24</sup>.

Prognostically, patients with localized LAMN have excellent survival outcomes, with recurrence rates remaining low when complete resection is achieved<sup>23</sup>. However, long-term surveillance is recommended due to the potential for delayed peritoneal dissemination, even in initially localized disease<sup>25</sup>.

This case contributes to the growing body of literature highlighting the diagnostic complexity of LAMN, particularly in atypical presentations involving large cystic masses and inflammatory changes or appearance of an appendix abscess. The unusually large volume of the lesion (~208 mL) in this patient further underscores the capacity of these tumors to reach significant size while remaining clinically deceptive. It reinforces the need for clinicians to maintain a high index of suspicion for underlying neoplastic pathology in patients presenting with atypical or disproportionately large appendiceal masses.

## **Conclusion**

Low-grade appendiceal mucinous neoplasm is a rare but clinically significant entity that may present with features closely resembling complicated appendicitis or appendiceal abscess, as demonstrated in this case. The considerable overlap in clinical and radiological findings often makes preoperative diagnosis challenging, particularly when large cystic collections and inflammatory changes are present. This case underscores the importance of maintaining a high index of suspicion for underlying neoplastic pathology in atypical or disproportionately large appendiceal masses.

Radiological evaluation, especially with contrast-enhanced computed tomography, plays a crucial role in identifying suggestive features; however, definitive diagnosis remains

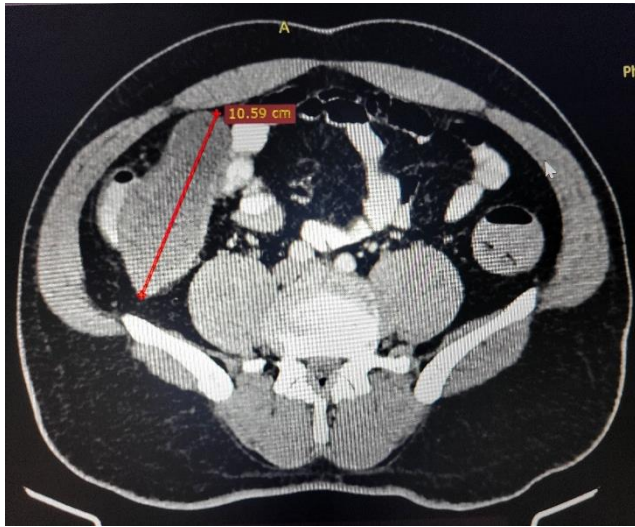
histopathological. Early surgical intervention not only facilitates accurate diagnosis but also prevents progression to pseudomyxoma peritonei, a serious complication associated with mucinous appendiceal tumors.

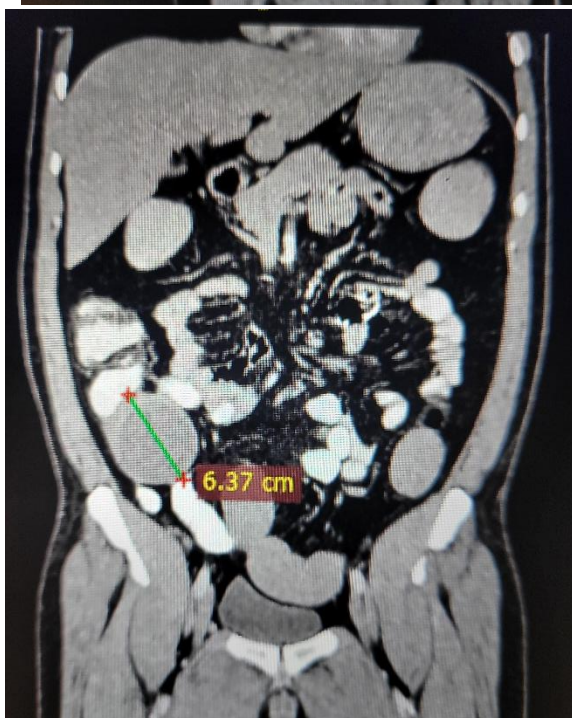
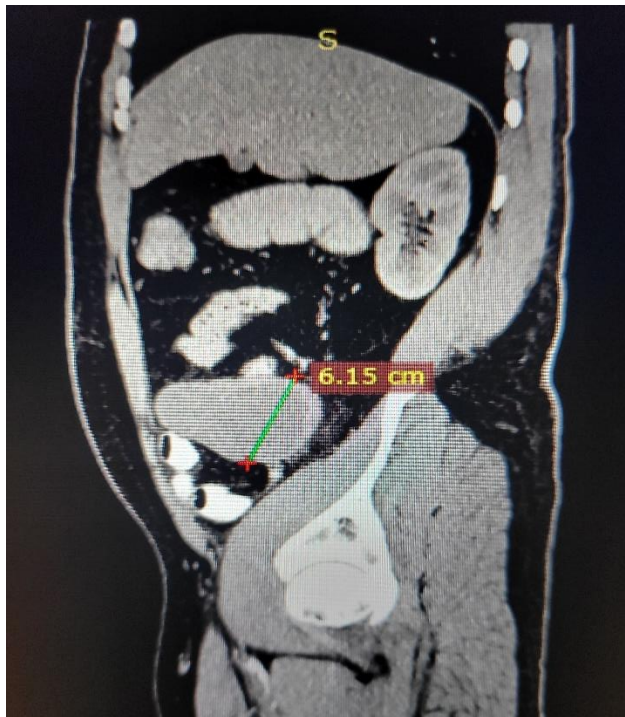
Overall, this report highlights the need for careful radiologic–pathologic correlation and emphasizes the importance of timely management and appropriate postoperative surveillance in optimizing patient outcomes.

### Endnotes

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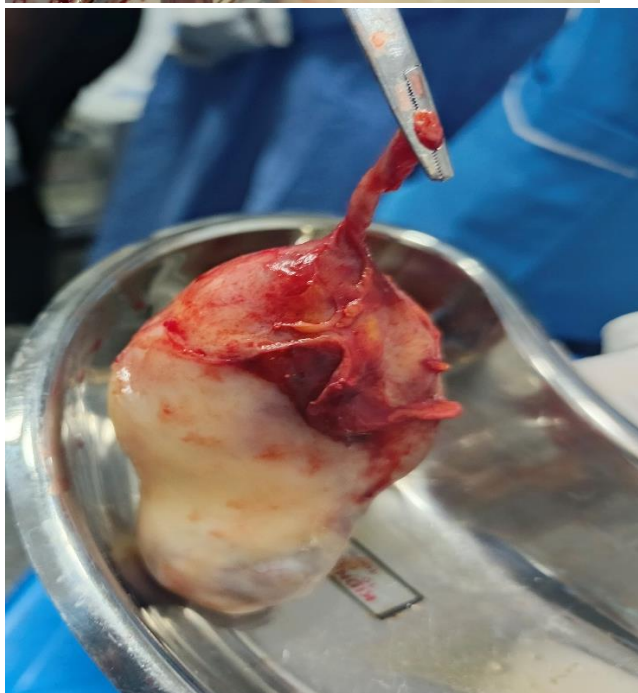
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**Figure 1: Contrast enhanced CT scan of the abdomen and pelvis, a) and b) Axial views c) sagittal view d) coronal view, showing a cystic lesion in the right abdomen. Subsequent exploratory laparotomy and appendectomy revealed a large cystic appendiceal tumor. Histology of the lesion was consistent with LAMN.**

LAMN, low-grade appendiceal mucinous neoplasm





**Figure 2:**

**Intraoperative and gross specimen of a cystic mass placed on a sterile surface. The lesion appears large and cystic with a smooth, glistening surface and focal hemorrhagic areas. Precise gross measurements were not obtained intraoperatively; however, radiologic dimensions were  $10.35 \times 6.01 \times 6.45$  cm, which closely approximate the visual size of the specimen.**