



## A Tumor Masquerading as Appendicitis in a Middle Aged Female Turnout be a Giant Low Grade Appendiceal Mucinous Neoplasm - A Rare Case Report

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### Abstract

Mucocele of the appendix refers to a mucus-filled appendix that may be caused by neoplastic or non-neoplastic illnesses. The most common etiology is low-grade appendiceal mucinous tumor. LAMN are rare tumors that account for <0.4-1% of all gastrointestinal malignancies. They are identified incidentally during appendectomy. Imaging alone cannot provide a good diagnosis, hence surgical excision without capsular disruption is indicated. We present the case of a 50-year-old woman who complained of right lower abdominal pain for two days. Imaging revealed a suspected acute appendicitis with periappendiceal fluid accumulation. She underwent an open appendectomy, which revealed a massive (12 x 6 x 5 cm) mucocele, and histology confirmed it to be a LAMN with negative margins.

**Keywords:** Mucocele; Database; Imaging

### Introduction

The term "appendiceal mucocele" refers to an enlarged and abnormally inflated appendix filled with mucus caused by a variety of non-neoplastic and neoplastic conditions. The most prevalent type of presentation is incidental; nonetheless, appendicitis appears in one-third of cases [1,2]. The most common etiology is low-grade appendiceal mucinous tumor. LAMN tumors are rare, accounting for < 0.4 - 1% of all cancers and are found inadvertent after appendectomy [3,4]. Appendiceal mucoceles have an incidence of 0.2 to 0.3% upon appendectomy and are most common in middle-aged women [5].

Recent database studies indicate that the incidence of appendiceal cancers is increasing from 0.63 to 0.97 per 100,000 people; however, this could be due to more selective appendectomy or more extensive pathologic examination [6]. Adenocarcinomas (mucinous, signet ring, or non-mucinous) account for approximately 20% of AMNs [7,8]. Imaging alone cannot provide a good diagnosis, hence surgical excision without capsular disruption is indicated [9].

Patients are asymptomatic in approximately 25% of cases, with others presenting with right iliac fossa pain comparable to appendicitis [10].

Low grade appendiceal mucinous neoplasms (LAMN), also known as appendiceal mucinous cystadenomas, are mucinous tumors of the appendix with low grade cytological atypia [11]. A low-grade appendiceal mucinous neoplasm (LAMN) is a well-differentiated tumor with a fibrotic, frequently calcified appendiceal wall that radiologists commonly refer to as a mucocele; nevertheless, mucocele is not a pathologic diagnosis. LAMNs may discharge mucin onto surrounding serosal surfaces. If a mucinous lesion is suspected, do not quickly grab it since it may burst and increase the risk of recurrence within the peritoneal cavity if the mucin contains malignant epithelium [12].

AMN is associated with serious consequences such as intestinal obstruction, intussusception, volvulus, mucocele rupture, and pseudomyxoma peritonei [3,4,13-15]. Elevated CEA, CA 19-9, and

CA 125 may be observed in 60-65% of AMN [16,17]. The majority of available literature suggests that a simple appendectomy is sufficient for early-stage LAMN [18].

Here, we discuss the case of a 50 year-old lady who complained of right lower abdomen pain and was later diagnosed with a huge LAMN with negative margins, which was treated with an open appendectomy. Continuing our case report, we conducted a review of the available literature on LAMN.

### Case Report

A 50 yrs old lady who had been diabetic for 25 years and had no previous surgical history reported to our surgical department with a history of pain in the right lower abdomen for 4 days. Imaging results were suspicious of acute appendicitis with periappendiceal fluid collection and the risk of perforation (Figure 3). At the time of presentation, the patient was in good health. Hemoglobin 10.7g/dl, total count 18,440 cells/mm<sup>3</sup> with 79% neutrophilia, and RBS 429 mg/dL.

The patient was counseled and scheduled for an emergency open an appendectomy. An intraoperatively enlarged appendix of approximately 14cms in length was discovered, which was snugly adherent to the surrounding tissues (Figure 1). Open adhesiolysis with appendectomy was performed, and the specimen was sent for histological analysis. The patient recovered with no immediate complications following the operation. Histopathology showed a low-grade mucinous appendiceal tumor with an uninvolved base of appendix (Figures 2 and 4). The patient is currently undergoing active annual follow-up.

### Discussion

An obstructive dilatation caused by an intraluminal buildup of mucoid material is known as an appendix mucocele. Retention cysts, mucosal hyperplasia, cystadenomas, and cystadenocarcinomas are the four processes that cause it. Less than 2 cm intact mucoceles are nearly usually benign, while bigger mucoceles have a higher probability of becoming malignant.

In contrast, if the mucocele has burst and epithelial cells have escaped into the peritoneal cavity, the patient wouldn't be at danger in the future if the mucocele is intact [1,2].



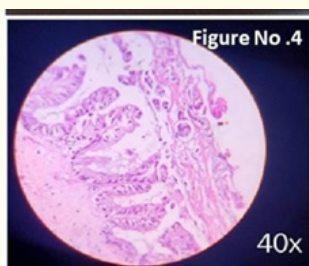
**Figure 1:** Intraoperation photograph of appendix specimen.



**Figure 2:** Gross specimen (pathology).



**Figure 3:** Ultrasound image showing enlarged appendix with periappendiceal fluid collection.



**Figure 4:** Histopathology image showing myxoglobulosis, lymphoplasmacytic infiltrate and mucin, mucosa lined by tall columnar mucinous epithelium with pseudo stratification.

During the initial evaluation, look for ascites, peritoneal disease, and scalloping of the liver surface on imaging. Because imaging alone is insufficient for a conclusive diagnosis, surgical removal without capsular disruption is recommended. The importance of carefully managing a mucocele and avoiding rupture cannot be emphasized, because in cases of adenocarcinoma, intraperitoneal dissemination of neoplastic cells and subsequent development of pseudomyxoma peritonei are nearly inevitable.

If a mucinous tumour of the appendix is suspected, the peritoneum should be thoroughly inspected and a peritoneal cancer index score calculated if mucin is found. Biopsies to determine epithelial cell, neoplastic cell, and mucin composition can be valuable [3-5].

Common misdiagnoses for this cancer include acute appendicitis, retroperitoneal tumors in the right iliac fossa, and an adnexal mass. Two imaging modalities are used for diagnosis: ultrasound (US) and computed tomography (CT), with CT being the most commonly used radiographic interpretation for preoperative diagnosis. Common abdominal CT abnormalities include cystic dilatation inside the appendiceal lumen, wall calcification, and uneven appendiceal wall thickening [13].

The optimal surgical procedure (laparoscopic versus open), adjuvant therapy, follow-up period, and imaging technique are all still debated. LAMN management's goals include rupture prevention, seeding, and PMP development. In the absence of lymph node metastases, right hemicolectomy has been substituted with an appendectomy-only strategy for the treatment of benign appendiceal mucocele [18]. If cancer has spread to the submucosa or lymph node metastases have been identified, a right hemicolectomy with or without omentectomy may be performed [18]. In our case, there

was no evidence of cancer infiltration into the gut submucosa or lymph node metastases, and no malignant cells were seen in the mucin pools of the periappendiceal tissue. Thus, additional surgical and adjuvant therapy are not required.

The treatment of low-grade appendiceal mucinous tumours with positive surgical margins following appendectomy is debatable, and there are no clear cut guidelines. Simple cecectomy, right hemicolectomy, and monitoring are all alternatives for treating an early stage low-grade appendiceal mucinous tumor with positive cut margin [19]. In a retrospective study of 16 patients (LAMN 15 patients, adenoma 1 patient), Arnason, *et al.* compared 6 persons who underwent cecal resection to 10 patients who were nonsurgically followed. The proximal resection margin was included. Following cecal excision, no residual cancer was found in these six people. After 4.7 years of follow-up, no patients in this series developed recurrence or pseudomyxoma peritonei [20].

The study by Arnason, *et al.* involvement of the margins after appendectomy is not indicative of recurrence in individuals with early stage low grade appendiceal mucinous tumor, and these cases can be treated conservatively without further surgery [20]. In our case, regular follow-up and observation were also suggested. In comparison to laparoscopic surgery, open surgical resection is the better treatment option. Laparoscopic treatment should be avoided since up to one-third of patients might experience mucocele rupture and mucin leakage in the peritoneum during surgery, which can result in pseudomyxoma peritonei. Adjuvant chemotherapy with fluorouracil (5-FU) is advised for high-risk histopathologic abnormalities such as high-grade poorly differentiated tumors, lymph node metastases, or surgical perforation. Adjuvant chemotherapy is not advised for low-grade mucinous tumors in their early stages [19].

During our literature review, we noticed that the majority of instances involving appendiceal mucoceles were in older women who underwent open appendectomy. A 64-year-old female underwent open surgery to treat an appendiceal mucocele with a retrocecal location.

An open appendectomy was carried out after a frozen section revealed clear resection margins [21]. Another case involves a 35-year-old guy who experienced recurrent RIF pain and was taken to the hospital for emergency surgery after being diagnosed with chronic appendicitis. Following surgery, histology confirmed the presence of an appendiceal mucocele [22].

An 80-year-old woman was diagnosed with an appendiceal mucocele, which was initially misdiagnosed as an ovarian cyst and identified intraoperatively during exploratory laparotomy, followed by a standard appendectomy. The authors noted the difficulty that surgeons face in such instances, as well as the need of adding appendiceal mucocele in the differential diagnosis of women who present with RIF discomfort and a mass but no other gynaecological symptoms [23].

Another example is a 38-year-old guy who experienced recurrent RIF pain and was admitted to the emergency hospital with the probability of chronic appendicitis. Following surgery, histology confirmed the presence of an appendiceal mucocele [11].

In our experience, two more middle-aged females underwent open appendectomy for suspected acute appendicitis and were identified with LAMN. Because the margins for high-risk characteristics were negative, appendectomy was regarded adequate for therapy, and patients were followed up with imaging every 6 months for the first two years, followed by yearly follow-up for the next five years [11,13].

In the case of patients who had undergone right hemicolectomy, a 70-year-old woman with appendiceal mucocele had an open extended right hemicolectomy with ileotransverse anastomosis due to cancer suspicion and a lack of a frozen section. Histopathology later revealed that it was a mucinous cystadenoma with mucocele, and she recovered normally [24].

Some argue that the type of surgery should be determined by the size of the mucocele, with right hemicolectomy being the best option for bigger mucoceles. However, in order to spare our patient from additional morbidity, we chose to conduct an appendectomy and then watch the histopathology to see whether a right hemicolectomy was necessary in the event of malignancy.

Our patient had a LAMN with a negative margin and no base involvement, hence an open appendectomy was the recommended surgery. Our patient is doing well right now, because of scheduled follow-ups and yearly imaging surveillance.

## Conclusion

Appendiceal mucinous neoplasms (AMN) are a rare but important diagnostic phenomenon. Early stage AMNs are typically discovered after a resection for appendicitis. Ultrasonography and CT scans are useful diagnostic techniques, however they are typically

discovered incidentally or following surgery. Low-grade mucinous neoplasms have a very good prognosis following routine appendectomy. The duration and methods of post-treatment surveillance are still not standardized. Our patient came with a massive appendiceal mucocele, which was removed via open appendectomy and later diagnosed as a LAMN. This should alert clinicians and surgeons to the possibility of this diagnosis, especially in a middle-aged female presenting with chronic abdominal discomfort, necessitating radiographic imaging to establish the amount of tumor growth and the proper surgical treatment.

## Author Contributions

Collection and/or assembly of data : Amar Hegde, Anand Bhandary Panambur, Manuscript writing and approval: Anand Bhandary Panambur.

## Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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