

**A tumor camouflaged as appendicitis. A giant low grade appendiceal mucinous neoplasm in a middle-aged female - Case report and review of literature**

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**Abstract**

Mucocele of the appendix broadly describes a mucus filled appendix that could be secondary to neoplastic or non-neoplastic pathologies. Most common etiology being Low grade appendiceal mucinous neoplasm (LAMN). LAMN are rare tumors accounting for < 0.4- 1 % of all gastrointestinal malignancies and are diagnosed incidentally at appendicectomy. A reliable diagnosis cannot be established using imaging alone and it is recommended that surgical excision without capsular disruption is undertaken. We present a case of a 55 years old woman who had complaints of right lower abdominal pain for 2 days. Imaging showed suspicious acute appendicitis with peri appendiceal fluid collection. She was taken for open appendicectomy which revealed a huge (12 x 6 x 5 cm) mucocele, and histology proved it to be a LAMN with negative margins.

**Keywords:** Low grade appendiceal mucinous neoplasm (lamn), mucocele of appendix, mucinous cystadenoma, open appendicectomy, mucin, appendix.

**Introduction**

Appendiceal mucocele refers to enlarged and abnormally distended appendix filled with mucin due to varied non-neoplastic and neoplastic pathologies (mucosal hyperplasia, simple or retention cysts, mucinous cystadenoma, mucinous cystadenocarcinoma). The most common form of presentation is incidental; however, presentation with appendicitis occurs in a third of cases<sup>1, 2</sup>. Most common etiology being Low grade appendiceal mucinous neoplasm (LAMN). LAMN are rare tumors accounting for < 0.4 - 1 % of all cancers and are diagnosed incidentally at appendicectomy.<sup>3,4</sup> Appendiceal mucoceles have a prevalence of 0.2 - 0.3%

at appendectomy, are seen most commonly in middle aged women.<sup>5</sup>

Recent database studies suggest that the incidence of appendiceal tumors may be increasing from 0.63 to 0.97 per 100,000 persons; however, this may be a consequence of more selective appendectomy or more thorough pathologic evaluation.<sup>6</sup> Adeno carcinoma (mucinous, signet ring or non-mucinous) constitute approximately 20% of AMNs.<sup>7, 8</sup> A reliable diagnosis cannot be established using imaging alone and it is recommended that surgical excision without capsular disruption is undertaken.<sup>9</sup> Patients are asymptomatic in around 25% of cases, others presenting with a right iliac fossa pain similar to appendicitis.<sup>10</sup>

Low grade appendiceal mucinous neoplasm (LAMN) previously known as appendiceal mucinous cystadenoma are mucinous tumors of appendix showing low grade cytological atypia.<sup>11</sup> A low-grade appendiceal mucinous neoplasm (LAMN) is a well-differentiated tumour with a fibrotic, often calcified appendiceal wall that is commonly described as a mucocele by radiologist; however, mucocele is not a pathologic diagnosis. Mucin may be extruded by LAMNs onto neighboring serosal surfaces. If a mucinous lesion is suspected, avoid immediately gripping it since it may rupture and increase the chance of recurrence within the peritoneal cavity if the mucin harbors malignant epithelium.<sup>12</sup>

Serious complications associated with AMN includes intestinal obstruction, intussusception, volvulus, rupture of mucocele and pseudomyxoma peritonei.<sup>3, 4, 13, 14, 15</sup> Elevated CEA, CA 19-9 and CA 125 may be detected in

60 – 65% of AMN.<sup>16, 17</sup> Most of the published literature suggests that simple appendectomy is sufficient for early stage LAMN.<sup>18</sup>

Here, we present a case of a 55 years old woman who had complaints of right lower abdominal pain and found to have a giant LAMN with negative margins, treated by open appendectomy. We follow our case report by review of literature on LAMN.

### **Case report**

55 years old woman, diabetic since 35 years with no past surgical history presented to our surgical department with history of pain in the right lower abdomen for 2 days with imaging results suspicious of acute appendicitis with peri appendiceal fluid collection with possibility of perforation. (Figure No.3) On presentation, patient was vitally stable. Hemoglobin 11.7g/dl, total count of 17,940 cells/mm<sup>3</sup> with neutrophilia of 86%, RBS 359mg/dl.

Patient was consented and was scheduled for an emergency open appendectomy. Intra operatively enlarged appendix measuring approximately 12cms in length was noted which was densely adhered to the surrounding structures. (Figure No.1) Adhesiolysis with appendectomy was performed and specimen sent for histopathological examination.

The patient recovered without incident. Low - grade mucinous appendiceal tumour with uninvolved base of appendix was discovered on histopathology (Figure no 2 & 4). Patient is currently on active periodic follow-up annually.



Figure 1. Intraoperative photograph of appendix specimen 2. Gross specimen (pathology) 3. Ultrasound image showing enlarged appendix with periappendiceal fluid collection. 4. Histopathology image showing myxoglobulosis, lymphoplasmacytic infiltrate and mucin, mucosa lined by tall columnar mucinous epithelium with pseudostratification.

## Discussion

A mucocele of the appendix is an obstructive dilatation by intraluminal accumulation of mucoïd material. It is by one of four processes: retention cysts, mucosal hyperplasia, cystadenomas, and cystadenocarcinomas. Intact mucoceles < 2 cm are almost always benign and larger mucoceles are more likely to be neoplastic. An intact mucocele presents no future risk for the patient; however, the opposite is true if the mucocele has ruptured and epithelial cells have escaped into the peritoneal cavity.<sup>1,2</sup>

During the initial evaluation, it's critical to look for ascites, peritoneal disease, and scalloping of the liver surface on imaging. Because imaging alone is insufficient to make a definitive diagnosis, surgical removal without capsular disruption is indicated. The necessity of careful handling of a mucocele and avoiding rupture cannot be overstated, because in situations of adeno carcinoma, intraperitoneal dissemination of neoplastic cells and subsequent development of pseudomyxoma peritonei are almost guaranteed. If a mucinous tumour of the appendix is suspected, the peritoneum must be thoroughly examined and a

peritoneal cancer index score recorded if mucin is detected. Biopsies to assess epithelial cell, neoplastic cell, and mucin composition can be beneficial.<sup>3,4,5</sup>

Acute appendicitis, retroperitoneal tumors in the right iliac fossa, or an adnexal mass are all common misdiagnoses for this cancer. Ultrasound (US) and computed tomography (CT) are two imaging modalities utilized for diagnosis, with CT being the most widely used radio graphic interpretation for preoperative diagnosis. Cystic dilatation inside the appendiceal lumen with wall calcifications and uneven appendiceal wall thickening are common abdominal CT findings.<sup>13</sup>

The best surgical strategy (laparoscopic versus open), adjuvant therapy, follow-up time, and imaging technique are all still up for debate. The prevention of rupture, seeding, and development of PMP are all goals of LAMN management. In the absence of lymph node metastases, right hemicolectomy has been replaced with an appendectomy-only approach for the treatment of benign appendiceal mucoceles.<sup>18</sup> Right hemicolectomy with or without omentectomy may be performed if cancer has infiltrated the submucosa or if lymph node metastases has been discovered.<sup>18</sup> There was no

evidence of malignancy infiltration into the intestine submucosa or lymph node metastasis in our case, and there were no malignant cells in the mucin pools in the peri appendiceal tissue. Thus, additional surgical and adjuvant therapy are not required.

The therapy of instances of low-grade appendiceal mucinous tumour with positive surgical margins after appendectomy is controversial, and there are no clear standards. Treatment options for early stage low-grade appendiceal mucinous tumour with positive cut margins include simple cecectomy, right hemicolectomy, and monitoring.<sup>19</sup> Arna son et al. compared 6 individuals who underwent cecal resection to 10 patients who were non surgically followed in a retrospective study of 16 patients (LAMN 15 patients, adenoma 1 patient) with included proximal resection margin. Following cecal excision, no residual tumour was seen in these six individuals. No patient in this series developed recurrence or pseudomyxoma peritonei after a mean follow-up time of 4.7 years.<sup>20</sup>

According to Arna son et al., involvement of margins following appendectomy does not indicate recurrence in patients of early stage low grade appendiceal mucinous neoplasm, and these cases can be handled conservatively without further surgery.<sup>20</sup> In our case, regular follow-up and observation were also recommended. In comparison to laparoscopic surgery, open surgical resection is the preferable therapeutic choice. Laparoscopic treatment should be avoided since up to one-third of patients may experience mucocele rupture and mucin leakage in the peritoneum during surgery, which can lead to pseudomyxoma peritonei. In high-risk histopathologic abnormalities such as high-grade poorly differentiated tumors, lymph node metastases, or perforation during surgery, adjuvant chemotherapy with fluorouracil (5-FU)

is advised following surgery. For early-stage low-grade mucinous tumors, adjuvant chemotherapy is not recommended.<sup>19</sup>

During our review of the literature, we discovered that the majority of cases reporting appendiceal mucoceles were in elderly ladies who were treated with open appendectomy. An open surgery for an appendiceal mucocele with retrocecal site was performed on a 64-year-old female. An open appendectomy was performed after a frozen section revealed clear resection margins.<sup>21</sup> Another case involves a 38-year-old man who had frequent episodes of RIF pain and was rushed to the hospital for emergency surgery after being diagnosed with chronic appendicitis. Histology confirmed the diagnosis of appendiceal mucocele after surgery.<sup>22</sup>

An 80-year-old woman was diagnosed with an appendiceal mucocele, which was initially misinterpreted as an ovarian cyst and was discovered intraoperatively during exploratory laparotomy, after which a regular appendectomy was performed. The authors emphasized the difficulty faced by surgeons in such situations, as well as the importance of including appendiceal mucocele among the differential diagnoses of women who arrive with RIF discomfort and mass but no other symptoms of gynecological illness.<sup>23</sup>

Another case of a 48-year-old lady who presented with lower abdominal pain and was diagnosed with a large LAMN (16.5 x 7 x 6 cm) and was treated by open appendectomy.

Another example involves a 38-year-old man who suffered from recurrent RIF pain and was sent to the emergency room with the possibility of chronic appendicitis. Histology confirmed the diagnosis of appendiceal mucocele after surgery.<sup>24</sup>

In our own experience, two more middle aged females underwent open appendicectomy for suspicious acute appendicitis and were diagnosed to have LAMN. Since the margins were negative for high risk features, appendicectomy was deemed sufficient for management and are kept on follow-up with imaging modality every 6 months for first 2 years followed by yearly follow-up for next 5 years.

In the instance of patients who had undergone right hemicolectomy, a 70-year-old woman with appendiceal mucocele underwent open extended right hemicolectomy with Ile transverse anastomosis due to suspicion of cancer and the lack of a frozen section. Histopathology later determined that it was a mucinous cystadenoma with mucocele, and she recovered uneventfully.<sup>25</sup>

Some suggest that the choice of surgery should be based on the size of the mucocele, with right hemicolectomy being the better option for larger mucoceles. However, in order to spare our patient extra morbidity, we opted to perform an appendectomy and then monitor the histopathology to see if a right hemicolectomy was warranted in the event of cancer.

Because our patient had a LAMN with negative margin and no involvement of the base, an open appendicectomy was the procedure of choice.

With scheduled follow - ups and yearly imaging surveillance, our patient is currently doing well.

### **Conclusion**

Appendiceal mucinous neoplasm (AMN) is a rare but relevant diagnostic phenomenon. Early stage AMNs are usually incidentally diagnosed at resection for suspicion of appendicitis. Ultrasonography and CT scan are valuable diagnostic tools, however they are frequently discovered incidentally or during surgery. After conventional appendectomy, low - grade mucinous

neoplasms have an excellent prognosis. The lengths and techniques of post-treatment surveillance are still not standardized. Our patient presented with a giant appendiceal mucocele which was removed by open appendicectomy and later proved to be a LAMN. This should alert clinicians and surgeons to the probability of this diagnosis, particularly in a middle-aged female presenting with chronic abdominal discomfort, and so require radio graphic evaluation to determine the extent of tumour growth and decide on the appropriate surgical care.

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