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# LOW GRADE APPENDICEAL MUCINOUS NEOPLASM – A RARE CASE REPORT

Pathology

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## ABSTRACT

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare mucinous tumour of appendix with low grade cytologic atypia. Patients presents with variable symptoms depending on the clinical manifestations. They can be often misdiagnosed as acute appendicitis or as retroperitoneal tumour. The most frequent complication of LAMN is Pseudomyxoma peritonei (PMP) which leads to mucin implants in the adjacent peritoneum. They are treated surgically with complete resection of the appendix, when it is non – metastatic. We report an unusual case of a 60-year-old male found to have LAMN status post elective appendectomy, six months after being treated for an appendiceal abscess.

## KEYWORDS

LAMN, PMN, Appendicectomy

## INTRODUCTION:

Appendiceal neoplasms constitues 0.5% of all gastrointestinal tract tumors [1]. Most appendiceal tumors consist of primary epithelial neoplasms which is further classified into Mucinous and Non – mucinous tumor and second most common is neuroendocrine tumors of appendix [2].

Appendiceal mucinous neoplasms (AMNs) are rare tumors, with a benign to malignant potential [3]. AMNs are classified as Low-grade AMNs (LAMNs), High-grade AMNs (HAMNs), Mucinous adenocarcinomas and poorly differentiated mucinous adenocarcinomas with signet ring features[4, 5, 6]. Most LAMNs are incidental findings during an appendicectomy and clinically patients can manifest with weight loss, nausea and vomiting, palpable mass, and abdominal distention [7, 8].

## Case Presentation:

A 60 year old male came with chief complaints of high grade intermittent fever, abdominal pain and loose stools for past two days. On palpation there is tenderness present in the right iliac fossa, right lumbar region and umbilicus. He was initially treated symptomatically, after 3 days inview of appendicular abscess patient was started with IV antibiotics. CECT abdomen shows dilated appendix with maximum diameter of 11.8mm with periappendiceal inflammatory changes and collection of around 39cc (Figure 1). Interval laproscopic appendicectomy was performed after six weeks when the patient was asymptomatic.

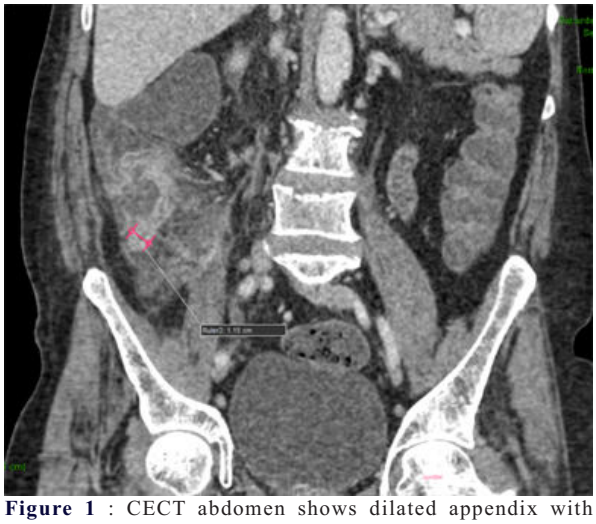

**Figure 1** : CECT abdomen shows dilated appendix with Periappendiceal fluid collection

**Gross:** Received appendicectomy specimen measuring 3.5x2x1cm. External surface of appendix is covered with exudate and focal areas show congestion. Cut surface shows obliterated lumen.

## Microscopy:

Sections studied shows undulating mucinous epithelium with low grade cytologic atypia. Architectural complexity , broad pushing front of the muscularis mucosa is seen (Figure 2). Invasion of the wall is absent. Adjacent appendix shows chronic inflammation suggestive of resolved appendicitis (Figure 3). The resected margin (base) is free of tumour.

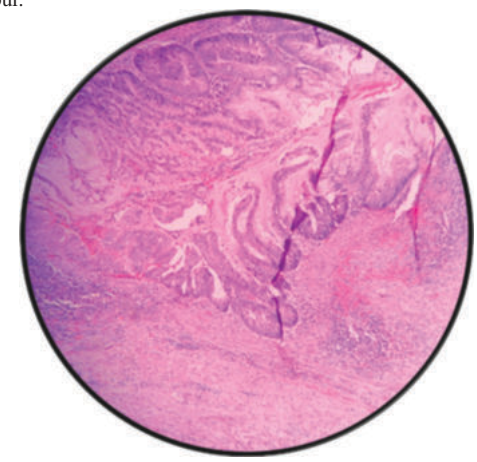

**Figure 2:** Photomicrograph shows architectural complexity with broad pushing borders into muscularis mucosa. 4X, H&E.E

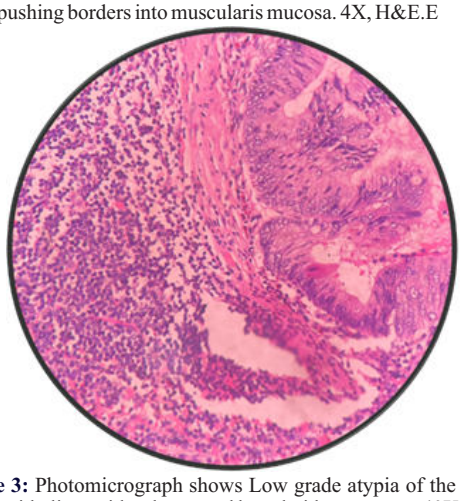

**Figure 3:** Photomicrograph shows Low grade atypia of the mucosal lining epithelium with submucosal lymphoid aggregates. 40X, H&E

## DISCUSSION:

LAMNs are rare tumor localized in the appendix or the surrounding appendiceal mucosa wall. Most primary appendiceal adenocarcinomas are of the mucinous type, arise from an adenomatous

polyp or serrated adenoma and frequently present with the Pseudomyxoma peritonei syndrome. These neoplasms are more common in men, particularly in the sixth decade of life. LAMNs are often incidental finding in asymptomatic patients, but rarely LAMN can present with abdominal pain, intussusception and obstruction. Most common complications of LAMN includes intussusception, ureteral obstruction, volvulus, small bowel obstruction (SBO), rupture, and PMP[1-2].

Often, this malignancy is misdiagnosed as acute appendicitis, retroperitoneal tumors in the right iliac fossa, or an adnexal mass [3].

Grossly, LAMN specimens show swollen appendix due to accumulation of gelatinous mucin inside the lumen with hyalinization and dense fibrosis of the appendiceal wall. LAMNs < 2 centimeters (cm) are rarely malignant and are classified as benign simple or retention mucoceles. Masses > 6 cm present with a higher risk of malignant cells, a higher risk of appendiceal perforation, and development of PMP [3].

Histological features of LAMN includes acellular dissecting mucin with "undulating growth" or "pushing invasion" or "diverticular – like growth " of malignant epithelial cells creeping into the appendiceal wall [4].

Some studies have shown elevation of CEA, Ca 19-9, and Ca-125 in 56 -67% of patients with LAMN [5].

Few studies have shown development of pseudomyxoma peritonei in 20% of the patients with LAMN after complete resection of primary lesion by appendicectomy. PMP is diagnosed by various imaging modalities such as ultrasonography and Computed Topography scan by the presence of gelatinous mucinous nodules in the peritoneal wall. They emphasized few macroscopic and microscopic features in LAMN cases that predicts the further development of Pseudomyxoma peritoni which includes,

1. Macroscopic findings: Smaller luminal diameter (<1 cm) and thicker wall because of separate mucin aggregations.

2. Microscopic findings: Acellular mucin and neoplastic cells presenting on serosal surface and not being confined to mucosa with evidence of microscopic perforation which indicated mucin leakage. In case of presence of diverticulum within the tumor, it can be considered as the weakest point for rupture (6).

The goal of management of LAMN includes the prevention of rupture, seeding, and development of PMP. The practice of right hemicolectomy in the absence of lymph node metastasis has been replaced, with an appendectomy only approach used for the treatment of benign appendiceal mucoceles. In the presence of, infiltrating malignancy into submucosa or with lymph node metastasis, right hemicolectomy with or without omentectomy may be performed [7].

In our case, there was no pathological evidence of malignancy infiltration into the bowel submucosa or lymph node metastasis and no evidence of malignant cells in the mucin pools in the periappendiceal tissue. Thus, further surgical and adjuvant therapies were not required in our patient. Our patient underwent a laparoscopic procedure that allows magnification of the surgical field and rapid patient recovery. The risk of peritoneal seeding increases with the removal of specimens through the port site but can decrease the risk of seeding overall as reported by Fujuni et al [8].

Lymph node metastasis is a rare and is seen in only 4.2% of patients but would require an aggressive treatment [9]. Active surveillance for every six months in two consecutive years is required for LAMN post appendicectomy patients to prevent tumour recurrence and development of PMP.

Follow-up should continue for five to 10 years with physical examination, annual CT, and monitoring of tumour markers. The five year survival rate for localized LAMN is 95%. Accurate pathological assessment and classification of LAMN are important to assess for malignancy risk, seeding, recurrence, and patient prognosis.[10]

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