

# Rare Case of Low-Grade Appendiceal Mucinous Neoplasm with Acellular Serosal Mucin: A Case Report and Review of Literature

Ashutosh Mishra<sup>1\*</sup>; Harekrishna Majhi<sup>2</sup>

<sup>1,2</sup>Department of General Surgery, SCB Medical College and Hospital, Cuttack, Odisha, India

Corresponding Author: Ashutosh Mishra<sup>1\*</sup>

Publication Date: 2025/11/13

**Abstract:** Low-grade appendiceal mucinous neoplasms (LAMNs) are rare epithelial tumors of the appendix that can lead to pseudomyxoma peritonei (PMP) if mucin spills into the peritoneal cavity. We report a 65-year-old man who presented with right iliac fossa pain and an appendicular mass. Exploratory laparotomy revealed a dilated appendix with mucinous content and ~200 mL of acellular mucin. Histopathology confirmed a well-differentiated LAMN (Grade 1) with acellular mucin reaching the serosa (pT4a), no invasion, and negative margins. Postoperative recovery was uneventful, and follow-up PET-CT revealed no recurrence. Appendectomy with lavage and surveillance was deemed sufficient. Surgeons should consider LAMN in appendicular masses associated with mucinous collections.

**Keywords:** LAMN, Acellular Mucin, Pseudomyxoma Peritonei, Appendectomy, Appendiceal Neoplasm, HIPEC.

**How to Cite:** Ashutosh Mishra; Harekrishna Majhi (2025) Rare Case of Low-Grade Appendiceal Mucinous Neoplasm with Acellular Serosal Mucin: A Case Report and Review of Literature. *International Journal of Innovative Science and Research Technology*, 10(11), 293-297. <https://doi.org/10.38124/ijisrt/25nov313>

## I. INTRODUCTION

Low-grade appendiceal mucinous neoplasms (LAMNs) are rare, accounting for <0.3% of all appendectomy specimens. They are characterized by villous or flat mucinous epithelium with low-grade cytology and abundant mucin production. Clinically, they mimic appendicitis. The 8th edition American Joint Committee on Cancer (AJCC) staging defines acellular mucin involving the serosa as pT4a. Management depends on histopathologic findings, ranging from simple appendectomy to cytoreductive surgery with HIPEC.

## II. CASE PRESENTATION

A 65-year-old male with no significant comorbidities presented to the General Surgery Department of SCB Medical College and Hospital, Cuttack, with complaints of right iliac fossa pain for five days. The pain was dull, continuous, and localized, without radiation or associated vomiting. The patient denied fever, jaundice, or changes in bowel or urinary habits. There was no prior history of abdominal surgery, weight loss, or gastrointestinal malignancy.

### ➤ Clinical Examination

On general examination, the patient was afebrile, with stable vital signs and no pallor, icterus, or lymphadenopathy. Abdominal examination revealed tenderness and a vague, ill-defined mass palpable in the right iliac fossa, approximately 5 × 4 cm in size, firm in consistency, and mildly mobile. There were no signs of generalized peritonitis or ascites.

### ➤ Laboratory Investigations

Routine hematological and biochemical parameters were within normal limits, except for a mildly elevated serum carcinoembryonic antigen (CEA) level. The findings are summarized in Table 1.

### ➤ Radiological Evaluation

An abdominal ultrasound revealed a markedly dilated, cystic, fluid-filled appendix measuring about 3 cm in diameter, with surrounding soft tissue stranding suggestive of mucocele or appendiceal abscess. No free intraperitoneal fluid or hepatic lesions were detected.

Based on these findings, a provisional diagnosis of appendicular mass (mucocele/complicated appendicitis) was made, and the patient was planned for exploratory laparotomy.

### ➤ Intraoperative Findings

During surgery, approximately 200 mL of clear, gelatinous mucinous fluid was found in the pelvis and right paracolic gutter. The appendix was grossly dilated, thick-walled, and tense, measuring  $7 \times 3.5 \times 3.0$  cm, arising from the ileocecal junction. There was no perforation, serosal breach, or visible tumor invasion into adjacent bowel or omentum. The cecum and terminal ileum appeared normal, and there were no palpable mesenteric or retroperitoneal lymph nodes.

A simple appendectomy was performed with careful handling to avoid rupture or spillage. The appendiceal stump was closed in two layers, and the peritoneal cavity was thoroughly lavaged with warm saline. A pelvic drain was placed before closure. The patient tolerated the procedure well.

### ➤ Gross Pathology

The resected specimen labeled as “*appendicular lump*” consisted of a distended appendix with tense outer surface and areas of transparent mucin beneath the serosa. On sectioning, the lumen was filled with thick mucinous material. No nodular mucosal growths or wall perforations were noted.

### ➤ Microscopic Examination

Histopathology revealed well-differentiated mucinous epithelium forming villous and flat patterns with minimal cytologic atypia, consistent with low-grade appendiceal mucinous neoplasm (LAMN). The muscularis propria was

intact, but acellular mucin was seen dissecting through to the subserosa and reaching the serosal surface. No viable tumour cells were identified in the serosal mucin. There was no lymphovascular or perineural invasion, and all resection margins were free of tumour.

Based on these findings, the lesion was staged as pT4a (acellular mucin on serosa, no invasion) and pM0, according to the American Joint Committee on Cancer (AJCC) 8th edition classification.

### ➤ Postoperative Course and Follow-Up

The postoperative period was uneventful. Oral feeding was resumed on postoperative day 2, and the drain was removed on day 5. The patient was discharged on postoperative day 11 in stable condition.

At 3-month follow-up, the patient was reviewed in the Surgical Oncology department. A whole-body Positron Emission Tomography – Computed Tomography (PET-CT) scan showed no abnormal FDG uptake or metabolic activity, confirming complete disease resolution. Contrast-enhanced CT (CECT) abdomen and pelvis showed only minimal postoperative changes without any evidence of peritoneal deposits or recurrence. Tumour markers (CEA 9.99 ng/mL, CA 19-9 20 U/mL) remained stable.

The patient continues to be on 3-monthly follow-up with serial CEA and CA 19-9 measurements and cross-sectional imaging as indicated.

Table 1 Laboratory Investigations

| Parameter                                        | Result                              | Reference Range                           |
|--------------------------------------------------|-------------------------------------|-------------------------------------------|
| <b>Haemoglobin</b>                               | 13.2 g/dL                           | 12 – 16 g/dL                              |
| <b>Total Leukocyte Count</b>                     | 9,800 / mm <sup>3</sup>             | 4,000 – 11,000 / mm <sup>3</sup>          |
| <b>Platelets</b>                                 | $2.5 \times 10^5$ / mm <sup>3</sup> | $1.5 - 4.5 \times 10^5$ / mm <sup>3</sup> |
| <b>Serum Amylase</b>                             | 180 U/L                             | 30 – 110 U/L                              |
| <b>Serum Lipase</b>                              | 160 U/L                             | 23 – 300 U/L                              |
| <b>Serum CEA (Carcinoembryonic Antigen)</b>      | 9.99 ng/mL                          | < 3 ng/mL                                 |
| <b>Serum CA 19-9 (Carbohydrate Antigen 19-9)</b> | 20 U/mL                             | < 37 U/mL                                 |

The Intra-operative images are shown in Figure 1.



Fig 1 Intraoperative Image Showing Mucinous Mass at Ileocecal Junction and Mucin Deposits in the Pelvis.

The Histopathology images are shown in figure 2.



Fig. 2 H&E-Stained Section Showing Dilated Appendix with Acellular Mucin Reaching Serosal Surface (LAMN, pT4a).

The follow up whole body PET-CT is shown in Figure 3.

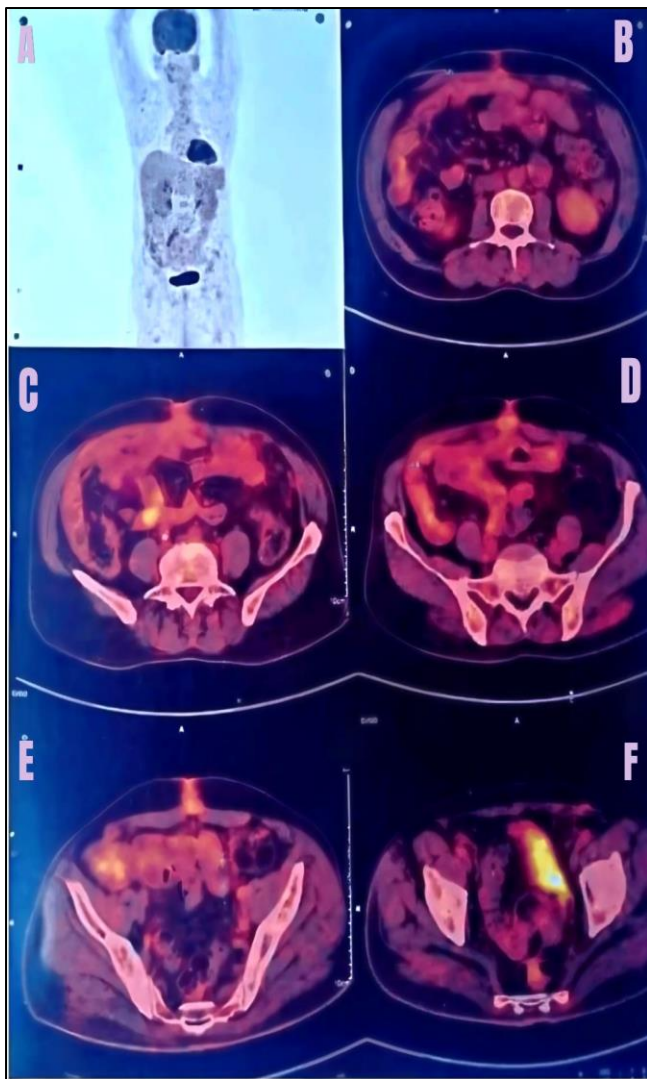


Fig 3 Follow-up PET-CT Showing no Abnormal FDG Uptake or Recurrence.

- 3(A): Whole-body PET maximum intensity projection (MIP) image showing physiological  $^{18}\text{F}$ -Fluorodeoxyglucose (FDG) uptake in the brain, myocardium, kidneys, urinary bladder, and bowel. No abnormal FDG-avid focus is seen elsewhere.
- 3(B): Axial PET-CT fusion at upper abdominal level showing homogeneous tracer distribution within the liver, spleen, kidneys, and bowel loops without focal hypermetabolic lesions.
- 3(C): Axial PET-CT section at mid-abdomen showing normal metabolic activity in bowel loops and mesenteric fat with no FDG-avid nodal or soft-tissue deposits.
- 3(D): Axial PET-CT at right iliac fossa level (previous disease site) showing no abnormal tracer uptake, suggesting complete metabolic resolution.
- 3(E): Axial PET-CT at pelvic inlet showing iliac vessels and pelvic musculature without FDG-avid lesions.
- 3(F): Axial PET-CT through the true pelvis showing physiological tracer accumulation in urinary bladder and rectum, with no abnormal pelvic uptake.

### III. DISCUSSION

Low-grade appendiceal mucinous neoplasms (LAMNs) are uncommon epithelial tumors, accounting for less than 0.3% of appendectomy specimens. They are part of a spectrum of appendiceal mucinous tumors ranging from benign retention cysts and mucocoeles to invasive adenocarcinomas. LAMNs are characterized histologically by villous or flat mucinous epithelium with low-grade atypia and "pushing" rather than infiltrative invasion. These tumors produce abundant mucin, which can dissect through the appendiceal wall and accumulate within the peritoneal cavity.

Clinically, patients often present with right lower quadrant pain resembling acute appendicitis or an appendicular lump. Occasionally, a palpable mass, ascites, or mucinous deposits are found incidentally during surgery. Imaging plays a crucial role; ultrasound or CT scan typically demonstrates a cystic, fluid-filled appendix or mucocoele with mural calcifications. However, the definitive diagnosis is histopathological.

#### ➤ Histopathological and Staging Considerations

LAMN differs from invasive adenocarcinoma by its non-destructive, pushing pattern of growth and absence of infiltrative invasion. The AJCC 8th Edition TNM classification recognizes LAMN as a unique entity. Tumors confined to the mucosa are staged as *pTis(LAMN)*, those extending into the subserosa as *pT3*, and those involving the serosal surface by mucin or mucinous epithelium as *pT4a*. When adjacent organs are invaded, the stage is *pT4b*.

In this case, acellular mucin was identified on the serosal surface without epithelial cells, consistent with *pT4a* disease, which carries a much better prognosis than cellular mucinous dissemination (*pM1b*).

#### ➤ Risk of Pseudomyxoma Peritonei (PMP)

One of the most serious complications of LAMN is pseudomyxoma peritonei, a condition characterized by progressive intraperitoneal accumulation of mucinous material that may contain neoplastic epithelial cells. The risk of PMP correlates with appendiceal perforation and the presence of cellular mucin beyond the appendix. Acellular mucin on the serosa, as in our patient, represents a lower-risk scenario.

Guner et al. (2023) reported recurrence in 7.5% of LAMN cases, primarily in patients with serosal perforation or cellular mucin. Similarly, Ballentine et al. (2021) found that acellular mucin was associated with favorable outcomes compared to cases with cellular peritoneal implants. Thus, the presence or absence of neoplastic epithelium in the peritoneal mucin is the key prognostic determinant.

#### ➤ Management Approach

The optimal management of LAMN remains debated. The American Society of Colon and Rectal Surgeons (ASCRS) guidelines recommend appendectomy alone for LAMNs with negative margins and no evidence of

peritoneal implants. Right hemicolectomy is reserved for tumors involving the appendiceal base, high-grade dysplasia, lymphovascular invasion, or positive margins. In our patient, appendectomy with peritoneal lavage was adequate since the resection margin was clear, and no cellular mucin was observed.

When cellular mucin or peritoneal involvement is present, cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC) is indicated to remove mucinous deposits and eradicate microscopic disease. This aggressive approach improves survival in PMP but carries significant morbidity, and hence is not justified for acellular mucin alone.

#### ➤ Follow-up and Prognosis

Surveillance is crucial due to the potential for late recurrence. Regular monitoring with serum tumor markers (CEA, CA 19-9) and cross-sectional imaging (CT or MRI) every 6–12 months for at least 5 years is recommended. Most recurrences occur within the first 2–3 years but late cases have been reported up to a decade later.

Our patient's postoperative PET-CT showed no residual disease, and tumor markers remained stable during follow-up. His prognosis is excellent, as studies indicate 5-year disease-free survival approaching 95% in patients with pT4a LAMN and acellular mucin only.

Table 2 AJCC (8th Edition) TNM Staging

| Stage Component   | Description                                                                              |
|-------------------|------------------------------------------------------------------------------------------|
| <b>Tis (LAMN)</b> | Tumor confined to mucosa with “pushing” invasion, no invasion beyond muscularis propria. |
| <b>T4a</b>        | Tumor invades through visceral peritoneum or acellular mucin involving serosa.           |
| <b>N0</b>         | No regional lymph node metastasis.                                                       |
| <b>M1a</b>        | Intraperitoneal acellular mucin without tumor cells.                                     |
| <b>M1b</b>        | Intraperitoneal metastasis with mucinous epithelium (PMP).                               |

#### IV. CONCLUSION

Low-grade appendiceal mucinous neoplasm is a rare entity often misdiagnosed as appendicitis. Accurate histopathologic evaluation is essential to guide management. Patients with acellular mucin (pT4a) and negative margins have excellent prognosis with appendectomy alone. Long-term follow-up is crucial to identify recurrence or progression to pseudomyxoma peritonei.

#### ACKNOWLEDGMENT

The authors thank the Department of Surgical Pathology and Surgical Oncology, SCB Medical College, for support.

#### REFERENCES

- [1]. Carr NJ, et al. A consensus for classification and pathologic reporting of pseudomyxoma peritonei. *Am J Surg Pathol*. 2016;40(1):14-26.
- [2]. Shaib WL, Assi R, Shamseddine A, et al. Low-grade appendiceal mucinous neoplasms: A case series. *J Surg Case Rep*. 2025;214.
- [3]. Guner M, Aydin C. Low-grade appendiceal mucinous neoplasm: What is the best treatment? *Cureus*. 2023;15:e46591.
- [4]. Fournier KF, Rafeeq S. Low-grade appendiceal mucinous neoplasm of uncertain malignant potential. *Ann Surg Oncol*. 2016;24:187-193.
- [5]. Glasgow SC, Gaertner W. ASCRS clinical practice guidelines for appendiceal neoplasms. *Dis Colon Rectum*. 2019;62:1425-1438.
- [6]. Ballentine SJ, Carr J, Bekhor EY. Updated staging and patient outcomes in low-grade appendiceal mucinous neoplasms. *Mod Pathol*. 2021;34:104-115.