Mucinous Appendiceal Neoplasm: Minimally Invasive Treatment as a First Step. Case series

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Case Series

Internal Medicine

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Background:
Mucinous appendiceal neoplasms (MANs) are rare epithelial tumors, representing <1% of appendectomy specimens. They are often discovered incidentally during surgery for appendicitis or imaging studies, with nonspecific symptoms like abdominal pain and distension. This study presents three cases of mucinous appendiceal neoplasms treated over the last decade with varied presentations, including acute abdomen, chronic diarrhea, and ureterolithiasis. All patients underwent laparoscopic appendectomy as the initial treatment, revealing low-grade mucinous neoplasms with negative margins, and none required further oncological treatment. The diagnosis of MANs relies on imaging techniques such as CT and MRI, along with tumor markers. Surgical resection, preferably appendectomy, is the standard treatment for low-grade lesions confined to the appendix, while more extensive procedures like right hemicolectomy may be indicated for invasive cases. Proper surgical handling is essential to prevent pseudomyxoma peritonei. This series highlights the importance of minimally invasive surgery for initial management and the need for a multidisciplinary approach in these rare tumors.

Keywords: Mucinous appendiceal neoplasms.

Primary tumors of the appendix are an uncommon condition, with an incidence of 1.2 cases per 100,000 people per year in the United States. No well-established risk factors are known. They often present with the classic symptoms of acute appendicitis and are incidentally diagnosed during postoperative histopathological examination. This organ can develop a wide variety of tumors. The differential diagnosis includes adenocarcinoma, neuroendocrine tumors, and mixed neoplasms. (12)

Case report

We present the case of three patients with different clinical presentations of mucinous appendiceal pathology, treated in the last 10 years:

A) A 44-year-old male with no significant history, presenting with acute abdomen suggestive of an appendiceal process. CT scan reported acute inflammatory appendiceal disease.

B) A 37-year-old male with no significant history, presenting with a clinical picture of diarrhea lasting one month. Infectious, allergic, and functional causes were ruled out. Colonoscopy revealed mucinous discharge from the appendiceal orifice. A CT scan showed a probable appendiceal tumor in the distal third of the appendix.

C) A 29-year-old male with no significant history, presenting with clinical features of ureteral lithiasis. CT scan revealed a stone in the mid-right ureter and a probable mucinous tumor in the middle third of the appendix.

In all three cases, laparoscopic appendectomy was performed as the initial treatment, with special attention to proper dissection of the appendiceal base (fig. 1). The specimens in all three cases demonstrated low-grade mucinous appendiceal neoplasms with negative margins (fig. 2). The patients did not require close follow-up given the described histological findings and absence of intraperitoneal findings.

Discussion

Mucinous appendiceal neoplasms (AMNs) are rare tumors, representing only 1% of appendectomy specimens. Due to their low incidence and atypical clinical presentation, these tumors are often misdiagnosed in clinical practice. They are commonly identified incidentally during surgery for appendicitis or through CT imaging. (11)

Preoperative diagnosis can be achieved using the following tools, especially in incidental cases rather than acute appendicitis:
A) **CT scan:** Presence of high-density mucin,

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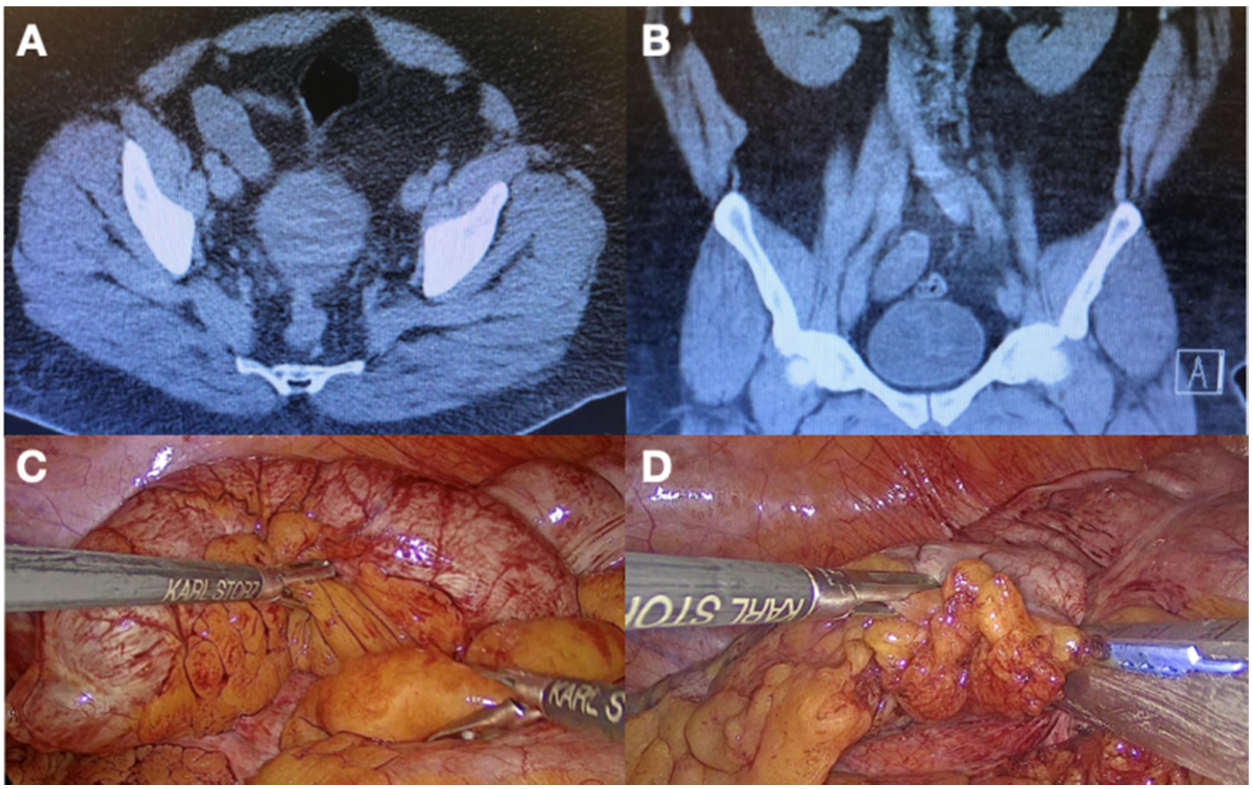

Figure 1. A) Simple abdominal CT scan, coronal cuts showing a dilated cecal appendix with semi-liquid content. B) Simple abdominal CT scan, axial cuts showing appendix dilation up to 28 mm. C) Laparoscopic approach showing a dilated appendix without evidence of perforation or mucinous implants. D) Laparoscopic appendectomy with a stapler at the appendiceal base.

characterization of lesion extension for surgical planning, and prognosis.
B) **MRI:** Useful for tumor localization, with T1 and T2 sequences being more sensitive for mucin distinction.
C) **Tumor markers:** Especially CA125, CA19-9, and CEA to avoid misdiagnoses.

All mucinous appendiceal lesions should be resected for diagnosis and treatment. The optimal treatment for low-grade neoplasms in a surgical context or the role of chemotherapy is still undefined. Most literature suggests that appendectomy alone is sufficient for lesions confined to the appendix. Some

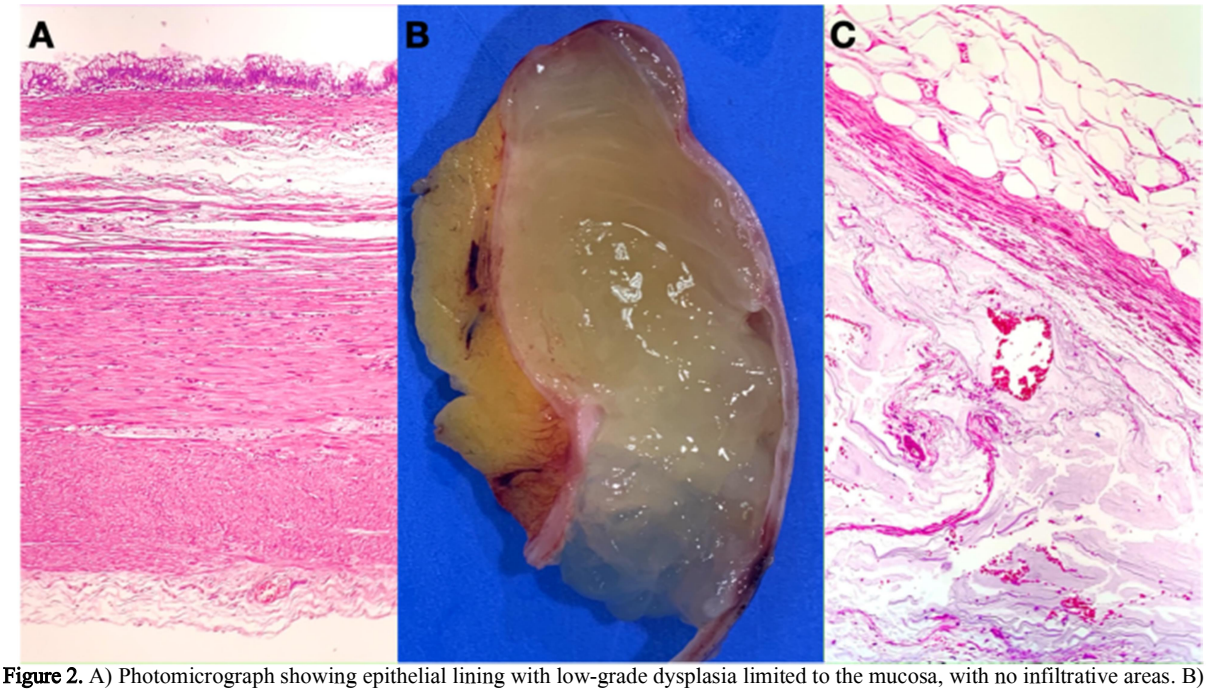

Figure 2. A) Photomicrograph showing epithelial lining with low-grade dysplasia limited to the mucosa, with no infiltrative areas. B) Cross-section of the cecal appendix showing mucosal flattening with luminal dilation filled with mucinous material and intramural mucin at the appendiceal apex. C) Subserosal adipose tissue with acellular mucin pools.

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recommendations suggest right hemicolectomy, along with peritoneal washing and inspection for suspicious lesions. Surgical removal should be performed carefully, avoiding perforation or mucin spillage to prevent pseudomyxoma peritonei.

The term **appendiceal mucocele (AM)** describes mucinous intraluminal distension of the appendix, regardless of underlying pathology. Its incidence ranges from 0.2% to 0.4% of appendectomy specimens.

In 2016, the Peritoneal Oncology Group issued a consensus on mucinous neoplasm classification:

- **Low-grade mucinous appendiceal neoplasms:** Extension through the mucosa into the appendiceal wall without infiltration and with low-grade atypia.
- **High-grade mucinous appendiceal neoplasms:** Extension through the mucosa into the appendiceal wall without infiltration but with high-grade atypia.
- **Mucinous adenocarcinoma:** Infiltrative invasion of the appendiceal wall with poor or moderate differentiation.
- **Mucinous adenocarcinoma with signet-ring cells:** Poorly differentiated with signet-ring cells.
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The histology of appendiceal neoplasms predicts biological behavior and dissemination. Regardless of atypia grade, both low- and high-grade neoplasms can have transmural extension or rupture, posing a risk for pseudomyxoma peritonei.

Conclusion

Appendiceal neoplasms are rare and can present as either a mucocele or an aggressive tumor with peritoneal dissemination. A multidisciplinary team is essential for proper diagnosis and treatment. Surgeons should be familiar with the clinical presentations, imaging limitations, and the use of minimally invasive surgery for treatment.

Conflicts of interests

There was no conflict of interest during the study, and it was not funded by any organization.

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