

When Lightning Strikes Twice: Synchronous Appendiceal Neoplasms

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Introduction

Cancer of the appendix, though rare, is cause for concern given that it can mimic appendicitis. Neuroendocrine tumors happen to be the most common appendicular cancers, accounting for 7 per 100,000 in prevalence. In comparison, low-grade appendiceal mucinous neoplasms (LAMN) make up 0.2-0.3% of appendectomy specimens. We report a rare case of a large appendicular mucocoele, measuring 10.7cm in length and 7.0cm in diameter, with final pathologies of both a LAMN and a neuroendocrine tumor.

Case Presentation

A 55-year-old male presented to the ED with three days of nondescript RLQ abdominal pain concerning for appendicitis. His history was notable for benign polyps on colonoscopy one year prior. Laboratory studies revealed a WBC 9.1, and CEA 12.5. CT Abdomen/Pelvis demonstrated a large cystic structure in the RLQ measuring 15x10cm, concerning for a dilated appendix secondary to appendiceal mucocoele. The patient was taken to the OR. Upon entering the abdomen, a large, smooth mass was encountered in the RLQ. After the soft tissue attachments encapsulating and anchoring the mass to the pelvis were bluntly dissected, a right hemicolectomy with ileotransverse anastomosis was performed. Pathology demonstrated a well-differentiated neuroendocrine tumor (carcinoid tumor) of the appendix tip measuring 1.6cm with vascular invasion and negative surgical margins. Additionally, a LAMN measuring 10.7cm involving the remainder of the appendix was noted with negative margins. Postoperatively, the patient did well and continues surveillance with medical oncology with yearly CT scans.

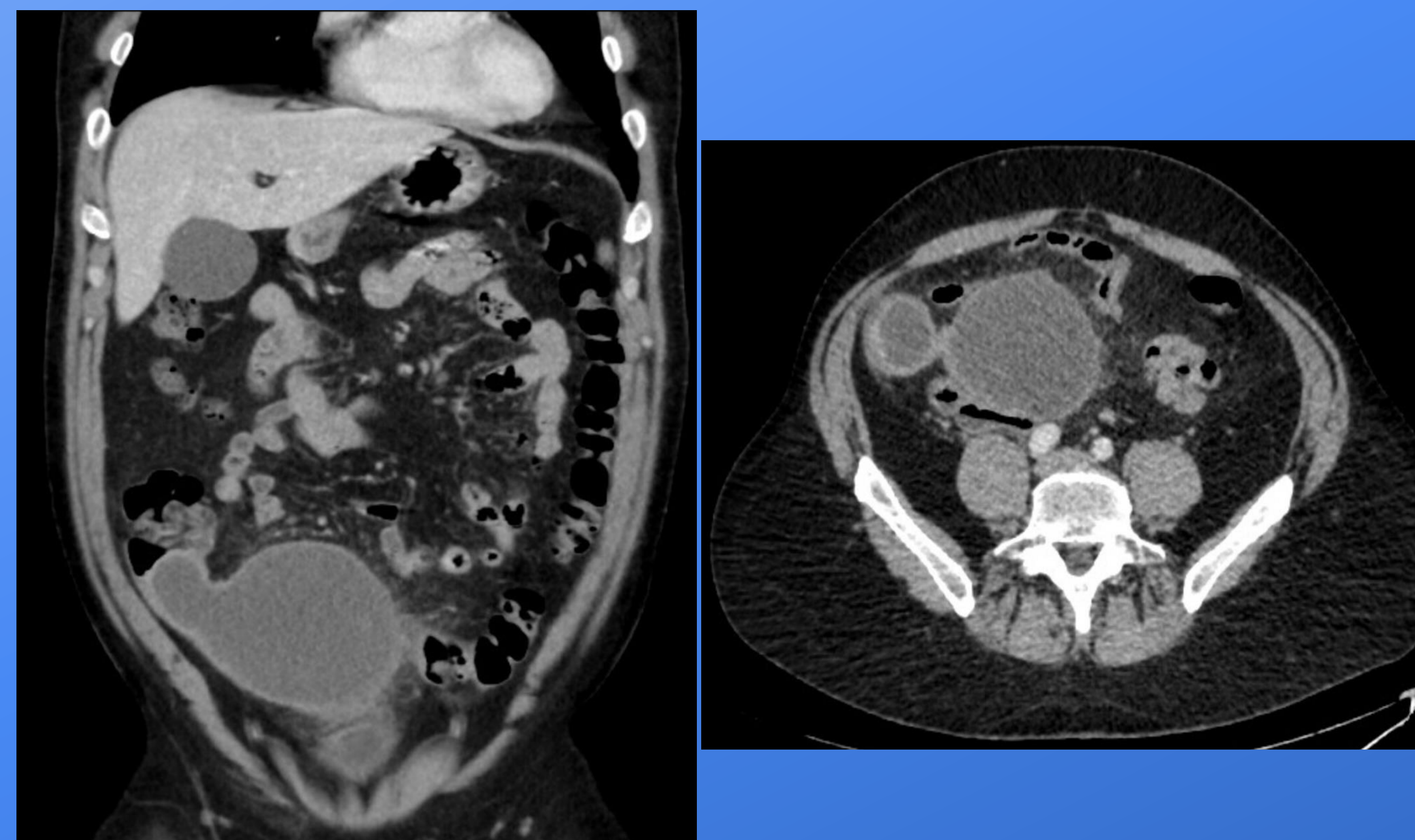


Figure 1. CT AP demonstrating a pear shaped, cystic mass believed to be a distended appendix secondary to appendiceal mucocoele (15x10cm).



Figure 2. Right hemicolectomy with mucocoele of the appendix.

Results

- Pathology showed 2 synchronous neoplasms:
 - A 1.6cm well-differentiated neuroendocrine (carcinoid) tumor with lymphatic invasion at the appendiceal tip
 - A low-grade appendiceal mucinous neoplasm involving the remaining appendix
- 13 lymph nodes, no carcinoma was identified (0/13).
- All final surgical margins were negative

Discussion / Conclusion

- Mucinous neoplasms are clinically significant due to malignant potential and risk of rupture, which may result in pseudomyxoma peritonei or peritoneal mucinosis
- Primary appendiceal tumors are rare, representing approximately 0.5% of gastrointestinal malignancies
- Neuroendocrine tumors are the most common appendiceal neoplasm (0.9 per 100,000 person-years)
- Low-grade appendiceal mucinous neoplasms (LAMNs) are less common (0.11 per 100,000 person-years) but account for ~54% of appendiceal mucocoeles in recent reports
- This case is notable for a large LAMN despite a benign colonoscopy one year prior and for the presence of two distinct neoplasms
- Appendiceal neoplasms should be considered in the differential diagnosis of suspected acute appendicitis

References

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