

An Incidental Discovery of Low-Grade Appendiceal Mucinous Neoplasm

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A 65-year-old man with a history of hyperplastic polyps underwent a surveillance colonoscopy, which revealed a large, smooth cystic bulge at the appendicular orifice (Figure 1). Subsequently, a computed tomography (CT) of the abdomen with contrast revealed an appendiceal mucocele measuring 13.3 x 4.5 cm (Figures 2a and 2b). Because of the abnormal imaging findings, an elective laparoscopic appendectomy and cecectomy was performed with minimal spillage. A gross specimen measuring 9 cm in length and 3 cm in diameter was collected with the appendiceal lumen, which was filled with yellow mucoid material. Histopathologic evaluation of the appendix revealed a low-grade appendiceal mucinous neoplasm (LAMN)-I (Figure 3). The patient had an uneventful recovery and was doing well at the time of writing. Repeat CT of the abdomen and pelvis was scheduled at 6 months



Figure 1. Colonoscopy image showing a large cystic lesion at the appendicular orifice.

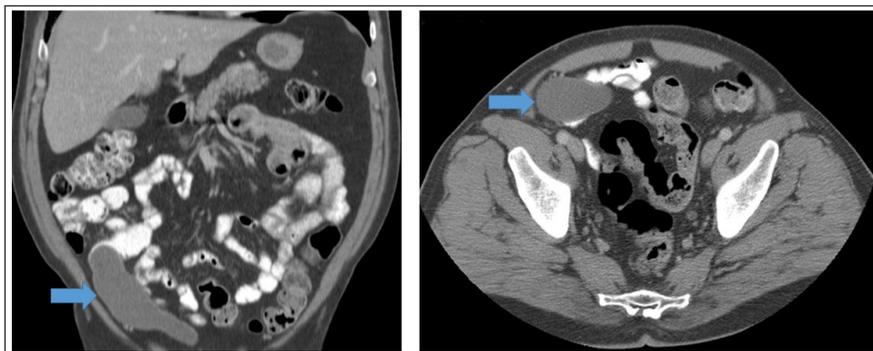


Figure 2a (left) and 2b (right). Computed tomography (coronal [2a] and transverse [2b]) with contrast, demonstrating a large appendix with diffuse wall thickening indicative of appendiceal mucocele neoplasm.

and 12 months, and a colonoscopy at 1 year after surgery.

On endoscopic visualization of this 65-year-old patient, lipoma or mucocele were considered the differentials. On the basis of the CT of the abdomen and of the endoscopy, we considered LAMN, with the rare possibility of pseudomyoma and peritoneal metastasis.

LAMN is a rare entity with an incidence ranging from 0.2% to 0.7% of all excised appendixes.¹ Approximately 25% to 50% of LAMN are incidental findings with the initial discovery during radiologic or endoscopic examinations, or during surgery.² Initial presentation can range from asymptomatic to right lower quadrant abdominal pain or a palpable abdominal mass. Initial clinical differentials include acute appendicitis, diverticulitis, ovarian mass (in women), large cecal mass, peritoneal carcinomatosis originating from

colon cancer, peritoneal sarcomatosis, peritoneal mesothelioma, disseminated peritoneal fungal infections, pseudomyxoma peritonei, or retroperitoneal cyst.^{1,3}

LAMN is divided into two major classes. LAMN-I is found in a younger group of patients, with the tumor confined to the appendix lumen.⁴ LAMN-I is rarely progressive, so a “wait-and-watch” policy is recommended with measurement of tumor markers, CT of the abdomen and pelvis at six months, and an annual work-up.⁴ LAMN-II is usually found in older patients, with mucin and/or neoplastic epithelium in the submucosa, the intestinal wall, or in the area around the appendix with or without perforation.^{1,4} Recommended treatment for LAMN-II includes hyperthermic intraperitoneal chemotherapy, prophylactic cytoreductive surgery, greater omentectomy and splenectomy, left upper quadrant peritonectomy, right upper

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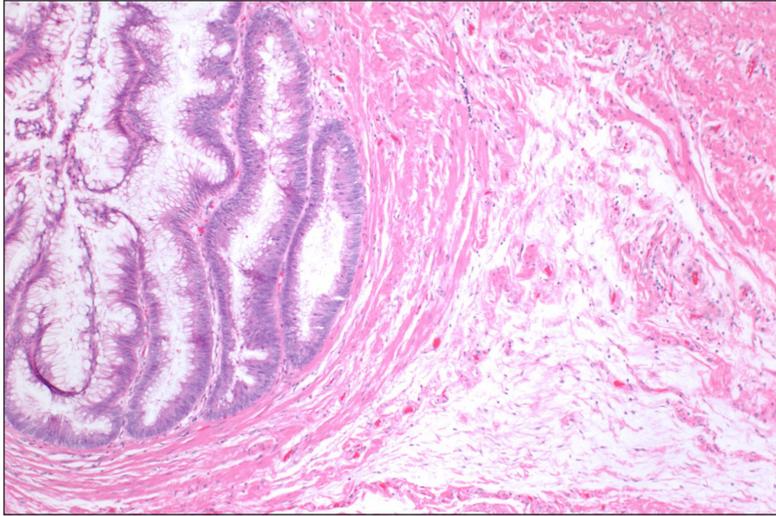


Figure 3. Histopathologic image (10x magnification) demonstrating villiform mucinous epithelium, glandular epithelium with tall columnar mucinous cells, and pseudostratified nuclei at the base.

quadrant peritonectomy, lesser omentectomy with cholecystectomy, pelvic peritonectomy with rectosigmoid resection, and antrectomy with a more aggressive follow-up.^{3,5} In women with concomitant peritoneum, ovarian, or appendix tumors, many authors believe that the appendix is the primary site with secondary involve-

ment of ovary and peritoneum, whereas others believe that each tumor has an independent origin.⁵ The topic continues to be debated. Since intact mucocoeles are benign, the worst outcomes are perforation or spillage of the mucocoeles into the peritoneal cavity, causing pseudomyxoma peritonei to develop.³ ❖

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

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