

Low-grade appendiceal mucinous neoplasm

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Abstract

Low-grade appendiceal mucinous neoplasm (LAMN) is extremely rare and easily misdiagnosed before surgery. Classically, LAMN can range from non-neoplastic to neoplastic tumors of the appendix which are incidentally found in up to 1% of appendectomy specimens. Appendiceal mucinous lesions are usually discovered incidentally during radiologic or endoscopic evaluation for unrelated symptoms. Though LAMN can be asymptomatic and stable over long periods of time, patients can present with abdominal pain, intussusception and obstruction. LAMN is an enigmatic tumor that lacks the capacity for classic invasion but can dissect through the appendiceal wall leading to pseudomyxoma peritonei (PMP). Therefore, early identification and treatment are crucial to reduce the risk of seeding of mucin and neoplastic epithelium into the peritoneum.

Introduction

LAMN is a rare indolent mucinous neoplasm of the appendix. In the past, there was considerable controversy regarding histological findings and clinical behavior of appendiceal mucinous neoplasms. In 2003, they were classified by Misdraji *et al* into low-grade appendiceal mucinous neoplasms LAMNs and mucinous adenocarcinoma¹. Though 25% of LAMN cases are asymptomatic², some patients present with symptoms similar to acute appendicitis which can include abdominal pain, nausea, vomiting and bloating³. Other patients present with peritoneal metastases after rupture or transmural invasion of the primary tumor.

Macroscopically, the appendix appears grossly distended. The lumen is filled with gelatinous mucinous material.

Microscopically, dysplastic epithelium produces abundant mucin and characteristically exhibits expansile growth with a "pushing" border, which may or may not cause loss of the muscular components of the appendicular wall⁴.

Here, we present a case of a 52-year-old man who came for a colonoscopy screening for ulcerative colitis. A glossy, rounded protruding mass (8× 4 × 2cm), arising from the

appendiceal orifice was detected. It was treated by a laparoscopic appendectomy and partial caecectomy, that was histologically LAMN. We follow our case presentation by a review of the relevant literature.

Case report

A 52-year-old male had a colonoscopy as surveillance for ulcerative colitis. The patient did not complain of any symptoms related to the appendix. The colonoscopy went smoothly up to the cecum. In the site of the appendicular orifice, a smooth bulbous submucosal lesion of the cecum with an impression formed by the appendiceal orifice was observed [Figure 1]. The lesion was confirmed to be a LAMN on histological examination. A surgical team was consulted and the patient underwent appendectomy with the removal of 3 cm of the cecum, the picture of the appendix is shown in [Figures 2 and 3]. The appendix was sent for histopathology analysis and the result revealed a feature of LAMN [Figures 4 and 5]. The patient's medical history is significant for ulcerative colitis diagnosed in 2015 when he presented with abdominal cramps and diarrhea. He has a history of five hospital admissions due to flare-ups. He was commenced on infliximab in 2018 every 8 weeks, then shifted to 4 weekly. He also takes mesalamine 2400 mg daily. He had a colonoscopy in 2019 which revealed a normal appendicular orifice. Prior to surgery, a CT scan was done and reported a distended appendix measuring 2 cm in maximum diameter with a smooth appendiceal wall with no evidence of calcification or peritoneal deposit [Figures 6 and 7]. The patient recovered well and was discharged home with no complications.

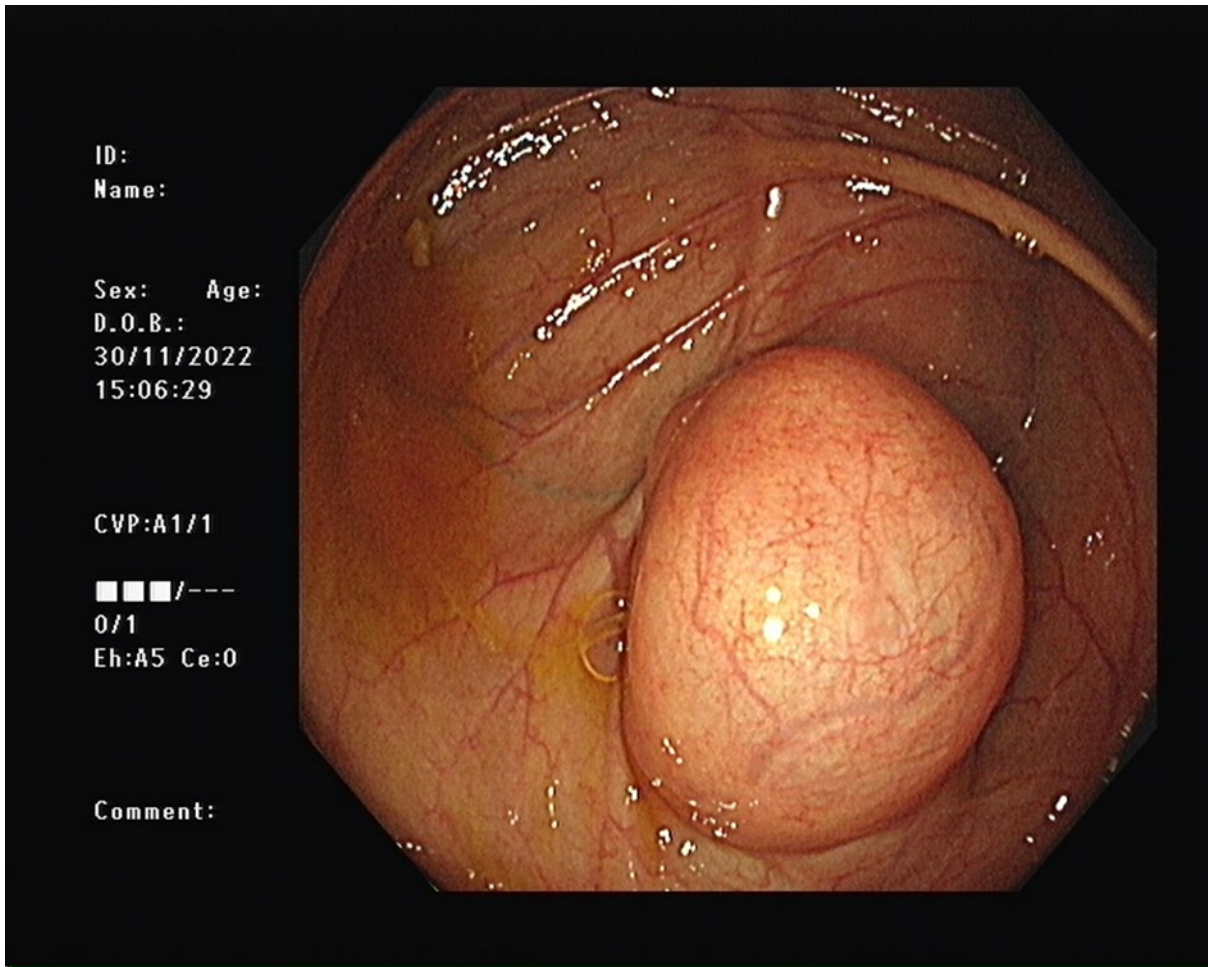


Figure 1: A smooth-surfaced rounded shape submucosal mass of the cecum with the appendiceal orifice in the center.

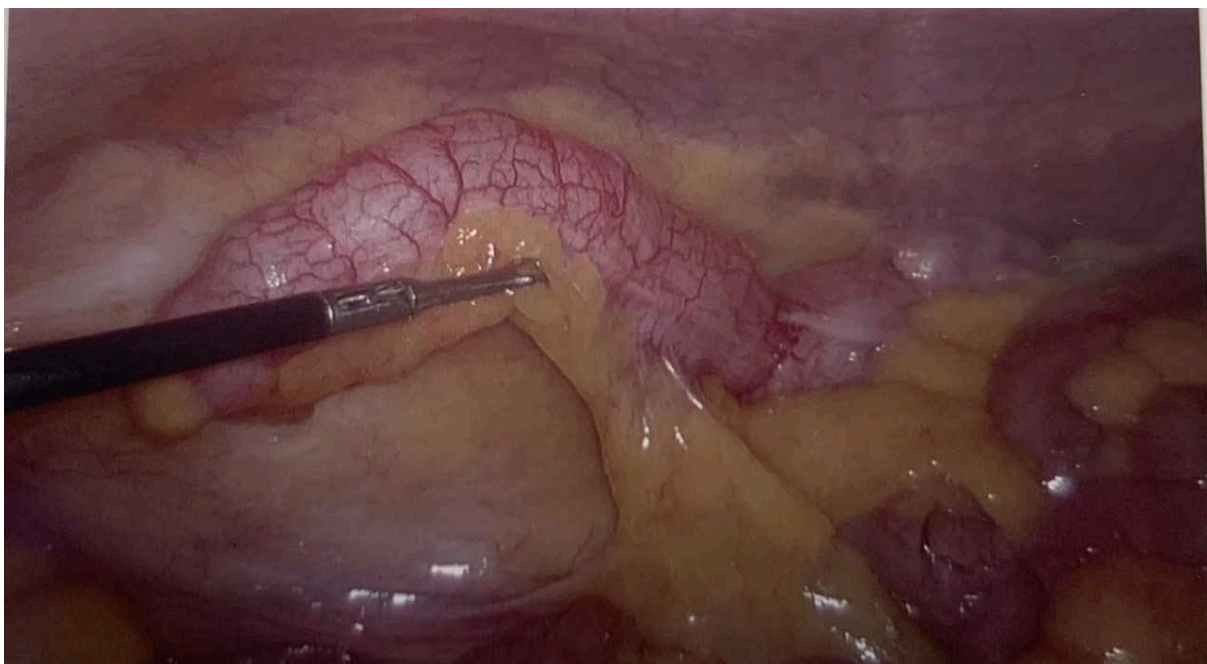


Figure 2: Intra-operative photo taken at the time of laparotomy showing distended appendix.



Figure 3: Gross appendiceal specimen.

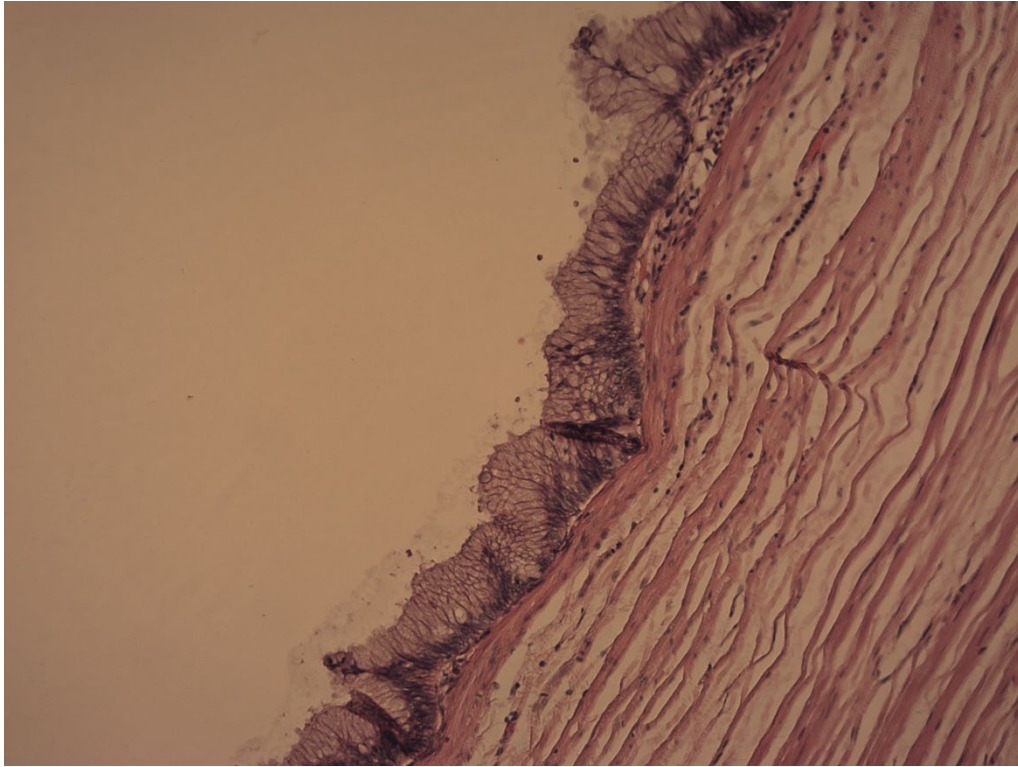


Figure 4: It shows the lumen of the appendix which is lined by predominantly flat glandular epithelium with low-grade cytological atypia (nuclear elongation and hyperchromasia). Normal lamina propria is absent and the epithelium is in direct contact with attenuated muscularis mucosa. No invasive glands were seen.

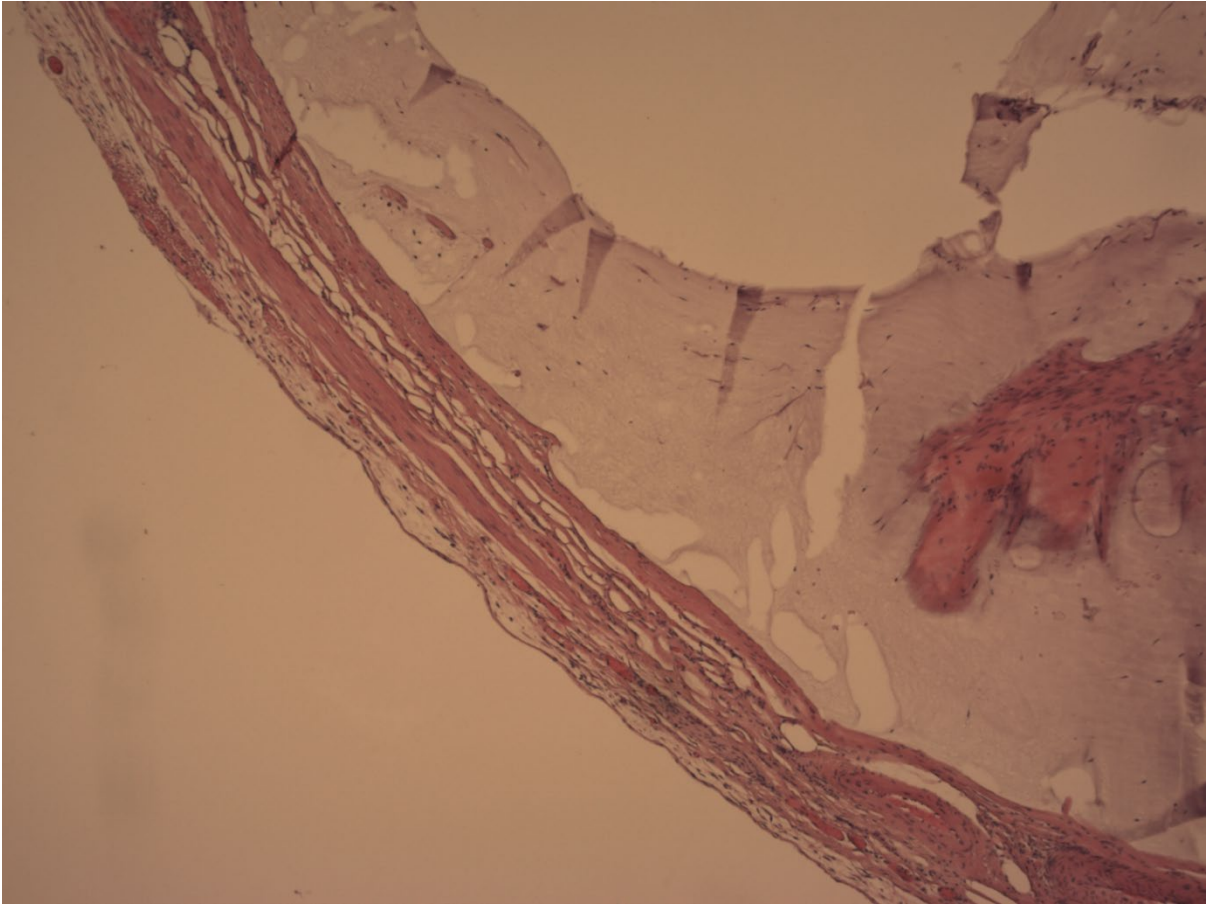


Figure 5: Appendiceal lumen distended by mucin

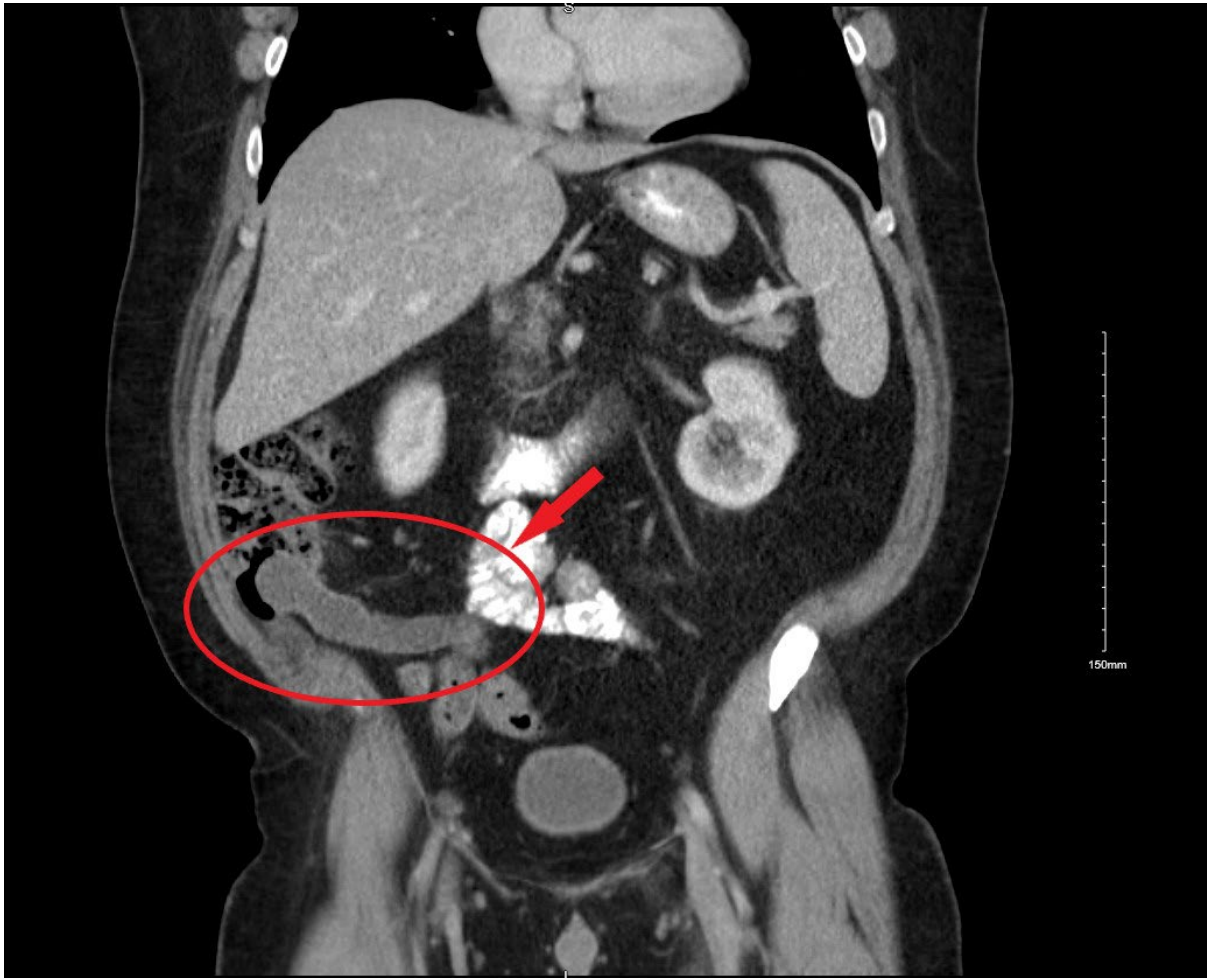


Figure 6: Intravenous and oral contrast-enhanced CT abdomen and pelvis – Coronal image

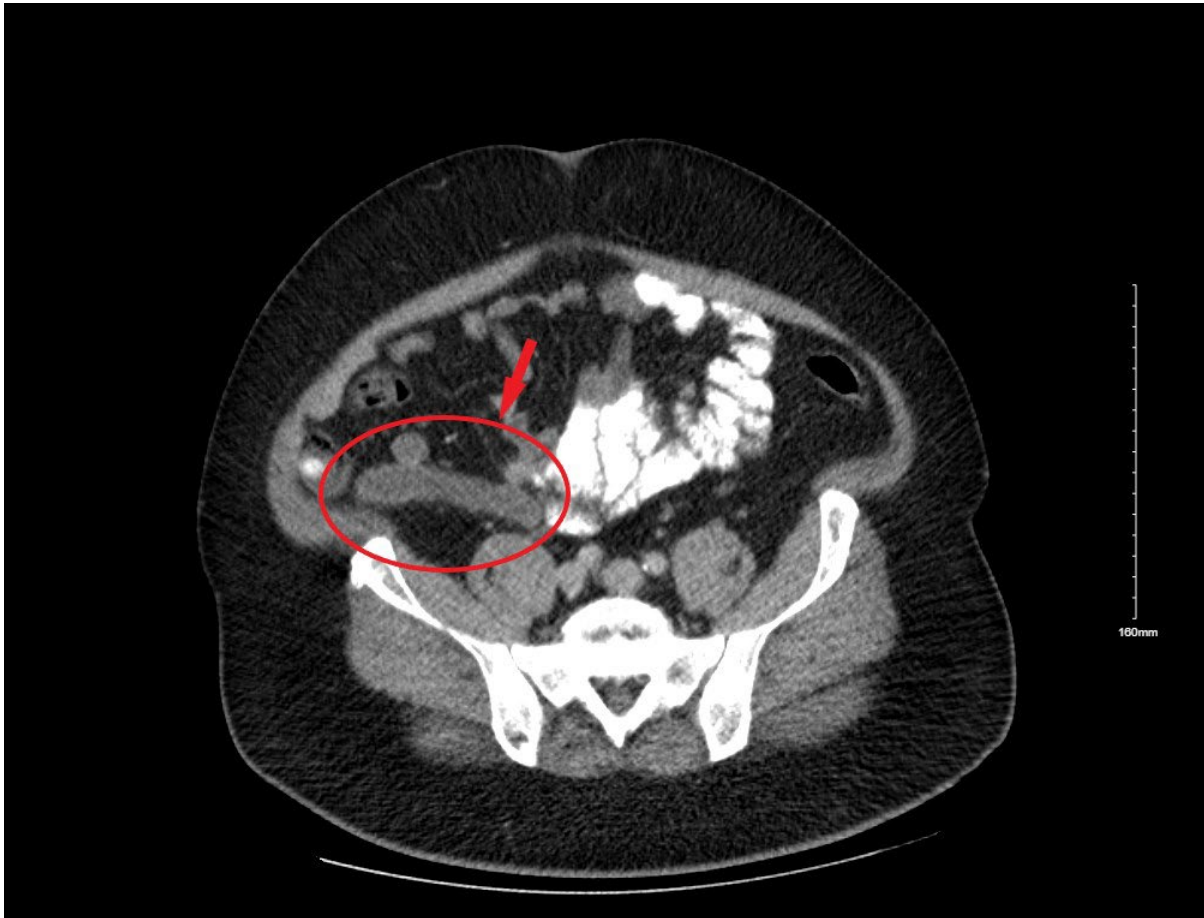


Figure 7: Intravenous and oral contrast-enhanced CT abdomen and pelvis – Axial image

Discussion

Older literature on appendiceal mucocoeles did not distinguish between benign and neoplastic lesions, the classification of appendiceal mucinous lesions has undergone significant clarification over the last 8 years⁵. The confusion surrounding diagnostic terminology has been resolved by Surface Oncology Group International (PSOGI In 2012⁶. According to (PSOGI) classification, appendicular mucocoele can be nonneoplastic such as simple mucocoeles, or neoplastic which includes serrated polyps of the appendix, mucinous appendiceal neoplasms, and mucinous adenocarcinomas of the appendix⁷. In 2016 the term “mucinous cystadenomas” was replaced with the new term “low-grade appendiceal mucinous neoplasm—LAMN⁶.”

It is difficult to make an appropriate early diagnosis of appendicular mucinous neoplasms because of the nonspecific clinical presentation unless the patient presented with features of appendicitis or pseudomyxoma peritonei when appendiceal neoplasms may perforate and spread to the peritoneal cavity. Most patients with LAMN are asymptomatic at the time of

diagnosis, with right lower quadrant pain being the most commonly reported symptom but, seen only in fewer than 25% percent of affected individuals⁸.

Radiologic studies in particular contrast-enhanced abdominal computed tomography (CT) and ultrasound are important diagnostic tools. They are usually done for right lower abdominal pain or mass or unrelated symptoms. However, imaging findings cannot definitively distinguish between non-neoplastic and neoplastic appendiceal mucocele⁹.

Colonoscopy — Appendiceal mucinous lesions may be found incidentally on colonoscopy performed for colorectal cancer screening or evaluation of other symptoms. Appendiceal mucoceles may be recognized at colonoscopy as a smooth bulbous submucosal lesion of the cecum with an impression formed by the appendiceal orifice.

Prognosis

patients with LAMNs confined to the appendix have no risk of disease progression or recurrence. This includes LAMNs with extension into the muscularis propria (pTis) as well as those with neoplastic epithelium or acellular mucin in the subserosal soft tissue (pT3), without the involvement of the serosal surface¹⁰.

Our case is 52 years old male who underwent a screening colonoscopy for ulcerative colitis. He denied any gastrointestinal symptoms related to his ulcerative colitis. The physical exam was unremarkable. The colonoscopy exam was normal except for a glossy, rounded, protruding mass arising from the appendiceal orifice. The overlying mucosa was normal. The mass didn't flatten with air insufflations [figure 5]. The patient was diagnosed with appendiceal mucocele on oral and intravenous enhanced CT. He underwent a laparoscopic appendectomy and removal of 3 cm from the cecum. He recovered well and was discharged home with no complications. Pathological examination revealed a LAMN confined to the appendix with uninvolved margins.

On a 2-month follow-up, the patient was not complaining of active complaints related to surgery or his controlled ulcerative colitis. Since the diagnosis proved to be a LAMN with negative margins, no further treatment was needed.

To the best of our knowledge, total excision of LAMNs confined to the appendix is associated with a favorable outcome.

Finally, on an extensive literature review, we were unable to find any report of concomitant IBD and of low-grade appendiceal mucinous neoplasm of the appendix in adult patients. However, in a study of 192 patients with LAMN of the appendix carried out in Mount Sinai, New York by Samuel J et al, 4.7% of the patients in the study had a concurrent diagnosis of IBD. This figure may be explained by the fact that the center is a major referral center for the diagnosis, treatment, and surveillance of patients with IBD¹¹.

Declarations of Conflicts of Interest:

None declared.

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