A 60-Year-Old Woman with a Cecal Lesion Seen on Colonoscopy

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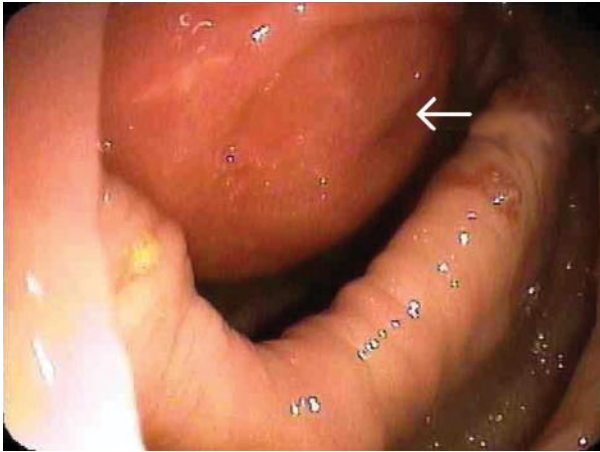
Figure 1. Endoscopic view of extrinsic compression of the cecum at the level of the appendiceal orifice (*arrow*).

Figure 2. Computed tomography scan demonstrating a fluid-filled mass (*black arrow*) extrinsically compressing the cecum (*white arrow*).

CASE PRESENTATION

A 60-year-old woman presented for routine screening colonoscopy following referral by her primary care physician. The patient was asymptomatic at the time. She was healthy with no medical conditions and took no medications. Colonoscopy was unremarkable with the exception of a submucosal lesion seen in the cecum that extrinsically compressed and partially obscured the appendiceal orifice (**Figure 1**). Multiple endoscopic biopsy specimens of the overlying cecal mucosa were taken, but these demonstrated normal colonic mucosa. Laboratory study results were normal, including a complete blood count with differential, electrolytes, liver function tests, and carcinoembryonic antigen (CEA).

The patient was then referred for a computed tomography (CT) scan of the abdomen/pelvis with oral and intravenous contrast. CT revealed an irregular, tortuous, fluid-filled, tubular-shaped structure that partially compressed the cecum. This structure measured approximately 2 cm wide in the region of the cecum to 1 cm in the region of the blind-ending tip, which descended into the pelvis (**Figure 2**). The liver, gallbladder, spleen, pancreas, small bowel, and kidneys were all normal. No lymphadenopathy was visualized.

WHAT IS YOUR DIAGNOSIS?

(A) Appendiceal adenocarcinoma
(B) Appendiceal gastrointestinal stromal tumor
(C) Appendiceal mucocele
(D) Lipoma
(E) Carcinoid tumor

WHAT IS THE NEXT APPROPRIATE STEP IN THE MANAGEMENT OF THIS LESION?

(A) Observation
(B) Percutaneous aspiration of the lesion to obtain cytology
(C) Repeat colonoscopy with biopsy
(D) Repeat CT imaging in 6 months
(E) Surgical resection

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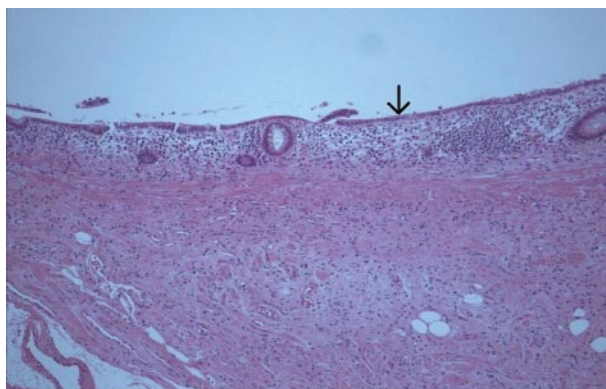
Figure 3. Pathologic image from a surgical specimen. The lining of the lesion demonstrates thinning of the mucosa with loss of glands, presumably due to dilatation and pressure atrophy (*arrow*). There are no neoplastic cells lining the dilated appendix. (Hematoxylin and eosin, 100×.)

ANSWERS

The correct answers are (C) appendiceal mucocele and (E) surgical resection.

DISCUSSION

A combination of history findings as well as imaging and laboratory studies point to the correct diagnosis of an appendiceal mucocele. The colonoscopy images reveal a submucosal lesion, which could be seen in the context of a lipoma, a gastrointestinal stromal tumor, or an appendiceal mucocele. Adenocarcinoma and carcinoid tumors are mucosal lesions and would most likely manifest with endoscopically abnormal mucosa; evaluation of biopsy samples from these lesions would result in a diagnosis. These lesions also would appear solid rather than fluid filled on CT imaging. This patient's biopsy specimens demonstrated normal colonic mucosa, which confirms the submucosal nature of the lesion. A lipoma is possible given the patient's asymptomatic presentation, and lipomas are seen as submucosal lesions; however, appendiceal lipomas would appear both solid and extremely dark on the CT scan, consistent with the usual appearance of fat on CT imaging.

The next step in management of this patient would be surgical resection. Appendiceal mucoceles have the potential for malignant transformation to mucinous adenocarcinoma; therefore, repeat imaging in 6 months or watchful waiting are not warranted in a healthy patient. In addition, malignant transformation may have already occurred and cannot be ruled out by imaging alone. Repeat colonoscopy with biopsy is unlikely to reveal any additional data as this is a submucosal lesion. Percutaneous aspiration of the lesion is rela-

tively contraindicated as the lesion requires resection in any event and there is a small risk of tumor seeding in the abdomen with such a procedure.

OUTCOME OF THE CASE

The CT findings and pathology reports prompted referral to a surgeon. The patient underwent laparoscopic appendectomy with partial cecectomy 2 weeks following her colonoscopy. Laparoscopy demonstrated no free fluid in her pelvis, and there was no sign of mucinous ascites. No lymphadenopathy or hepatic metastases were observed at the time of surgery. No other tumors were noted in the abdominal or pelvic cavities. The appendix was distended up to the base of the cecum. There was no disruption of the appendix. The patient did well postoperatively and was discharged home 2 days following the surgery. There were no complications. The final pathology specimen revealed a simple appendiceal mucocele with focal dilatation and fibrosis along with prominent lymphoid follicle formation. There was no evidence of malignancy (**Figure 3**).

APPENDICEAL MUCOCELE

Mucoceles are rare pathologic lesions of the appendix that are characterized by cystic dilatation of the lumen due to abnormal accumulation of mucus. The incidence of mucocele ranges from 0.2% to 0.3% of all appendectomies, with a higher frequency in women (4:1) and in persons older than 50 years.[1,2] This accumulation of mucus can be related to a variety of pathologic conditions. Most concerning are the mucoceles caused by mucinous cystadenomas and cystadenocarcinomas. In the latter case, rupture of the cystadenocarcinoma may result in the clinical condition pseudomyxoma peritonei. Pseudomyxoma peritonei has been variably defined in the past but generally refers to an accumulation of mucin within the abdominal cavity. The mucin may contain malignant epithelium or adenomatous epithelium with only low-grade dysplasia or may be acellular altogether.[3,4] A correct diagnosis may help avoid iatrogenic rupture during surgery, although spontaneous rupture can sometimes occur (**Figure 4**).

Differential Diagnosis

Different types of malignancies can be found in the appendix, and mucoceles (as potentially malignant lesions) should be viewed in the context of a larger differential diagnosis. A study by McGory et al[5] reviewed 2514 appendiceal cancer cases, grouping them into the following categories: mucinous adenocarcinoma (951), adenocarcinoma (646), carcinoid (435), goblet (369), and signet-ring cell (113). Five-year survival was

highest for carcinoid and lowest for signet-ring cell tumors. A review of 2117 malignancies by McCusker et al[6] found similar results.

The term *mucocele* is often used as a general descriptive term for dilatation of the appendiceal lumen by mucinous secretions. Three general subgroups of the disease can be identified according to the characteristics of the epithelium: simple or retention mucoceles, cystadenomas, and malignant cystadenocarcinomas. Simple mucoceles result from obstruction of the appendiceal outflow, usually by a fecalith (hard mass of feces), and are characterized by normal epithelium and mild luminal dilatation. Cystadenomas are the most common form, accounting for 63% to 84% of cases. Lined by adenomatous epithelium, these exhibit mostly epithelial villous adenomatous changes with some degree of epithelial atypia; they are characterized by marked distention of the lumen up to 6 cm. Malignant cystadenocarcinomas represent 11% to 20% of cases. They are distinguished from benign mucoceles by stromal invasion of the dysplastic epithelium and are therefore, by definition, malignant. The luminal distention in these lesions can often be 6 cm or greater.[7–9]

Carr et al[10] reviewed the pathology from 184 noncarcinoid appendiceal tumors surgically resected between 1976 and 1992. In this series that included simple mucoceles, hyperplastic polyps, adenomas, carcinomas, and mucinous tumors of uncertain malignant potential, 46% of the lesions were malignant and 38% were adenomas or tumors of unknown malignant potential. Only a small proportion of all lesions were nonmucinous. Simple mucoceles were small in number, accounting for only 6% to 10% of the total, respectively. There was a significant association with synchronous or metachronous carcinoma of the colon, as well as with mucinous ovarian tumors.[10]

Clinical Features

The symptomatology of appendiceal mucoceles is often nonspecific: many lesions are completely asymptomatic. Large lesions are asymptomatic in only 25% of patients. The most common presentation of all lesions is right lower quadrant pain, similar to an acute appendicitis; a palpable mass can be found in 50% of cases.[8] Elevation of tumor markers such as CEA indicates a probable neoplastic origin.[11,12] Rare complications include intestinal obstruction, frequently caused by intussusception, as well as gastrointestinal bleeding.[13]

The worst complication of appendiceal mucocele is pseudomyxoma peritonei, which is characterized by peritoneal dissemination caused by spontaneous or
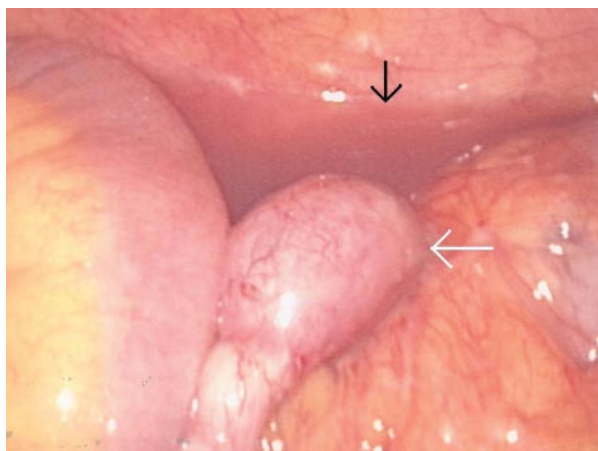

Figure 4. Representative laparoscopic image of an appendiceal mucocele that spontaneously perforated (*white arrow*) with dissemination of periappendicular free fluid (*black arrow*). (Image courtesy of Jason Schwartz, MD, University of Utah, Salt Lake City, UT.)

iatrogenic perforation of the appendix. In benign mucoceles, the tumor is confined to the periappendicular area. In malignant cases, this dissemination is considered a real metastatic entity; retroperitoneal or pleural implants in this setting have been reported.[11,12]

Treatment

Because the endoscopic evaluation of appendiceal lesion is often limited and imaging studies such as CT and MRI are not always able to differentiate between benign and malignant lesions, it is recommended that all mucoceles be managed by surgical excision. Appendectomy is often adequate, unless the involved appendiceal wall is contiguous with the cecum, in which case the latter will also require excision, as occurred in the case patient.[14] A right hemicolectomy (rather than appendectomy) is performed for all noncarcinoid appendiceal tumors or if a carcinoid tumor is larger than 2 cm. In some patients, these resections can be accomplished laparoscopically.[15] If exploration reveals a ruptured appendiceal mucocele with pseudomyxoma peritonei, the primary resection should be accompanied by removal of all gross implants.[16–18]

Postoperatively, patients with simple or benign neoplastic mucoceles have an excellent prognosis, with 5-year survival rates of 91% to 100%, even in cases with extension of mucus into the extra-appendiceal spaces. In malignant mucoceles, however, the 5-year survival rate falls to approximately 25%.[1]

In summary, patients with mucoceles can present with various symptoms, but may also be asymptomatic. Recognition of these lesions and surgical resection are

critical given the potential for malignant transformation and the possibility for rupture with the development of pseudomyxoma peritonei. **HP**

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