Case Report: Appendiceal Mucocele, an Uncommon Answer to Common Symptoms

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abstract
A 49-year-old man presented with abdominal pain, fatigue, tachycardia, and anemia three weeks after sphenopalatine artery ligation for massive epistaxis. In light of the recent bleed, this constellation of symptoms was non-specific and therefore difficult to interpret. They were consistent with peptic ulcer disease, a very common diagnosis that can generally be worked up as an outpatient. This resulted in a delay of several months in the diagnosis and treatment of a large appendiceal mucocele, an uncommon and often benign entity involving the accumulation of mucous within the appendix with a potential for malignant spread. Although this might have been an incidental finding, all symptoms have resolved since surgical removal of the growth. This case illustrates the importance of timely diagnosis in patients with vague yet persistent symptoms. An uncommon condition can have potentially serious consequences if missed.

case
After being discharged home, the patient returned to care three times over the following month with episodes of epigastric discomfort, tachycardia, and malaise. During each visit, he had epigastric tenderness, but no definitive diagnosis was achieved. An esophagogastroduodenoscopy (EGD) was planned, but it was not prioritized due to the negative results of the Helicobacter pylori breath test. A 24-hour urine sample was collected upon suspicion that a pheochromocytoma might explain his episodic symptoms, but the catecholamines and metabolite assays were normal. Other laboratory investigations were normal, including TSH, liver enzymes, liver function, renal/electrolytes, lipase, CEA, cortisol, and CRP. Absent an anatomical diagnosis, we considered psychosomatic abdominal pain due to anxiety or stress. However, the patient was mystified, given his previous perfectly normal health and enjoyment of frequent vigorous exercise.

While he awaited endoscopy, the abdominal pain worsened, and he lost 7 kg (approximately 10%) of his body weight. An abdominal CT scan was ordered two months after the initial presentation. This revealed early mesenteric panniculitis of the jejunum and massive distension of the appendix, which protruded into the cecal lumen and contained low-density debris consistent with an appendiceal mucocele. The patient was referred to general surgery.

Prior to surgery, the EGD was normal. Colonoscopy demonstrated a mass adjacent to the ileocecal valve originating from the cecal pole, and an ultrasound also confirmed appendiceal mucocele. More than three months after first seeking medical attention, the patient underwent a laparoscopic hand-assisted right hemicolectomy. There was no evidence of carcinomatosis or pseudomyxoma peritonei. However, an inflammatory process was observed in the right upper quadrant with adhesions, which were lysed intraoperatively.

Pathological analysis of the appendiceal specimen showed a 6 by 1.5 cm low-grade mucinous neoplasm confined to the appendix, without evidence of rupture or invasion into the adjacent cecum. Thirteen lymph nodes were negative for metastatic cells. The patient was symptom-free after discharge and at six-month follow-up.
Appendiceal mucoceles are present in 0.2 to 0.3% of all appendectomy specimens and in 8% of appendiceal tumours. A mucocele refers to an appendix that has dilated due to progressive accumulation of mucus within its lumen. They can be considered benign or malignant and can be histologically subdivided into four subtypes. Mucoceles are often found incidentally on imaging or during surgery and are usually seen in patients over the age of 50.

Diagnosis is confirmed by pathology. The first three histological types are considered benign, and the last is malignant. Type I appendiceal mucoceles are the result of proximal obstruction in the lumen (commonly a fecolith), causing a retention mucocele less than 1 cm in diameter with normal epithelium. If the appendix has mild dilatation and normal but hyperplastic epithelium, it is considered to be a type II mucocele (5-25% of all appendiceal mucoceles). Type III or mucinous cystadenomas are the most common (63-84% of all appendiceal mucoceles), and are generally less than 6 cm in diameter and lined by columnar epithelium with atypia. Type IV mucoceles (11-20% of all appendiceal mucoceles) are malignant adenocarcinomas that show severe dilatation, glandular stromal invasion, and/or peritoneal implants.

The signs and symptoms of appendiceal mucoceles are non-specific. They include abdominal pain and mass, nausea, vomiting, and weight loss (Table 1). In one case series of 19 patients over 20 years, 11 presented clinically as appendicitis until imaging suggested a mucocele. Of 135 patients identified over 25 years at the Mayo Clinic, only 65 had symptoms attributable to appendiceal pathology.

While asymptomatic patients are difficult to diagnose, the literature shows that it is symptomatic patients who are more likely to have malignant disease. Certain symptoms and findings more commonly found in patients with adenocarcinomas include abdominal pain (56%), weight loss (77%), abdominal mass (86%), mucocele extravasation (83%), and diffuse intra-peritoneal spread (95%).

While the definitive diagnosis depends on pathology, a presumptive diagnosis can be made from imaging and direct visualization. Low-attenuation mass adjacent to the cecum on CT (Figure 1) or an ultrasonographic image of a through-transmitting mass with echogenic content in the correct location is suggestive of the diagnosis. Colonoscopy can reveal a soft submucosal lesion in

<table>
<thead>
<tr>
<th>Symptom/sign</th>
<th>Prevalence</th>
<th>Sources</th>
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<tbody>
<tr>
<td>Abdominal pain</td>
<td>27-100%</td>
<td>Papziogas et al. 2007, Stocchi et al. 2003</td>
</tr>
<tr>
<td>Abdominal mass</td>
<td>16-50%</td>
<td>Rampone et al. 2005, Stocchi et al. 2003</td>
</tr>
<tr>
<td>Weight loss</td>
<td>5-10%</td>
<td>Papziogas et al. 2007, Stocchi et al. 2003</td>
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<tr>
<td>Nausea/vomiting</td>
<td>5-9%</td>
<td>Papziogas et al. 2007, Stocchi et al. 2003</td>
</tr>
<tr>
<td>Unexplained anemia</td>
<td>5%</td>
<td>Papziogas et al. 2007, Stocchi et al. 2003</td>
</tr>
<tr>
<td>Acute appendicitis</td>
<td>8-57%</td>
<td>Papziogas et al. 2007, Stocchi et al. 2003</td>
</tr>
<tr>
<td>Asymptomatic or symptoms not reasonably attributable to the appendix</td>
<td>25-51%</td>
<td>Papziogas et al. 2007, Rampone et al. 2005, Stocchi et al. 2003</td>
</tr>
</tbody>
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Table 1: Prevalence of presenting symptoms in patients with appendiceal mucocele.

Figure 1: Sagittal section of CT abdomen pelvis. Appendiceal mucocele (arrow) with measurements of 6 x 1.5 cm. Max diameter 2.6 cm.
the cecum with a central impression (appendiceal orifice) from which mucous can be visible as a “volcano sign.” Pre-operative colonoscopy is used to screen for concurrent colon tumors, reportedly found in up to 29% of cases, which can alter the surgical approach. While colorectal tumors are the most common tumor associated with appendiceal mucocles, ovarian, breast, and kidney tumors have also been linked.

The major complications from untreated appendiceal mucocles include intestinal obstruction or bleeding, intussusception, fistulization, and volvulus. One of the most serious is the development of pseudomyxoma peritonei, the accumulation of thick gelatinous mucin containing neoplastic cells in the peritoneal cavity. This can occur through the natural progression of the neoplasm by invasion through the thinned appendiceal wall or via spontaneous or iatrogenic rupture. If the tumor is malignant, the material can be generalized throughout the peritoneum rather than localized to the peri-appendicular space as seen with benign mucocles.

Surgical resection is preferred to observation due to the potential for malignant transformation and dissemination. A recent literature review recommended that an open approach is generally preferred over laparoscopy to reduce the risk of iatrogenic perforation. Accordingly, if the mucocoele is discovered incidentally during a laparoscopic surgery, it is recommended that the surgeon convert to an open procedure. Simple appendectomy typically results in cure if the mucocoele is small and confined to the appendix. When there is evidence of perforation, positive cytology, lymph nodes, or margins on pathology, a right hemicolectomy is recommended with post-operative intraperitoneal chemotherapy and long-term follow-up.

The prognosis of a benign appendiceal mucocoele after treatment is good, with a 5-year survival of 91-100%. For malignant lesions, this drops to 25% at five years, usually due to complications from pseudomyxoma peritonei.

Another rare condition that might have played a role in our patient was mesenteric panniculitis, a chronic inflammatory condition of mesenteric adipose tissue of unknown etiology. A large case series of patients with CT findings of mesenteric panniculitis found that 45 of 118 (38%) had concurrent malignancy, mostly colorectal, lymphoma, and urogenital tumors. Mesenteric panniculitis can also have no identifiable cause, yet it can produce a variety of symptoms, including abdominal pain. It is unclear whether our patient’s mucocoele and panniculitis were linked, as the mesenteric panniculitis might have caused his symptoms, and the mucocoele may have been only an incidental finding from abdominal CT.

Our patient experienced a delay of definitive diagnosis of a potentially serious condition after he presented with persistent abdominal pain. Barriers to accurate diagnosis and definitive management included a recent but apparently unrelated emergent condition (life-threatening epistaxis) and a lack of striking physical or laboratory findings. The persistent complaint of unexplained abdominal pain in an otherwise well-looking patient ultimately led to CT imaging, yielding unexpected but significant findings.

In an era when we should question the overuse of ionizing radiation, this patient’s persistent resolve that something was wrong was the catalyst to the diagnosis and treatment of uncommon conditions and to resolution of his symptoms. Sir William Osler taught, “If you listen, the patient will tell you the diagnosis.” In this case, the patient gave us a needed push in the right direction.

Subjective:
- The patient is a 49-year-old man with a history of peptic ulcer disease presenting with fatigue, malaise, and recurrent episodes of mild epigastric abdominal pain after a recent massive epistaxis requiring sphenopalatine artery ligation.

Objective:
- Heart rate is 100 beats/min, blood pressure is 118/84 mmHg without orthostatic change, and the abdomen is diffusely tender without localized pain or peritoneal signs. The hemoglobin is 118 g/L (135-170), the MCV 85 fL (82-98), and his stool tests positive for occult blood. Other laboratory tests are normal, including TSH, CRP, urine catecholamines, and Helicobacter pylori breath testing.
-Computed tomography of the abdomen reveals massive distention of the appendix containing low-density debris, consistent with an appendiceal mucocoele.

Assessment:
- The patient is a 49-year-old man with nonspecific abdominal complaints, normocytic anemia, and a history consistent with peptic ulcer disease. After failing to respond to empiric proton-pump inhibitor therapy, he has been found to have an appendiceal mucocoele on CT imaging of the abdomen.

Plan:
- Referred to general surgery for surgical management of the lesion, for assessment of the extent of disease, and to rule out concurrent malignancy in the colon via colonoscopy.

Figure 2: SOAP note.

recesses