Appendiceal Mucoceles

Luise Pernar, Harvard Medical School IV
Gillian Lieberman, MD
Patient 1: JH

- 68 yo woman with history of myelodysplastic syndrome
- Admitted to BIDMC for induction of chemotherapy for acute myelogenous leukemia
- During course of induction patient developed fever and neutropenia
- CT scan was performed to search for possible site of infection
Patient 1: JH - CT

- Cecum
- Terminal Ileum
- Cyst (27 HU)
- Focal wall calcification

PACS, BIDMC - Courtesy of Karen Lee, MD
Patient 1: JH - CT

• Findings summary:
  – Cystic structure adjacent to cecum near ileocecal junction; appendix not seen separately
  – Density ~ 27HU
  – Suggestion of rim-enhancement of cystic wall with focal calcification
  – No surrounding stranding suggestive of inflammation

➤ Officially read as: ‘likely an appendiceal mucocele’
Patient 2: LC

- 54 yo woman with history of breast cancer diagnosed in 2000; treated with L mastectomy, chest wall radiation therapy, tamoxifen
- Presented with complaints of abdominal distention and mild abdominal pain
- Paracentesis yielded 1.5L of ascites fluid with malignant cells
- CT scan was performed to determine possible source
Patient 2: LC - CT

Cecum

Terminal Ileum

Rim-enhancement

Omental caking

Cyst (34 HU)

Ascites fluid

PACS, BIDMC - Courtesy of Karen Lee, MD
Patient 2: LC - CT

- **Findings summary:**
  - Cystic structure adjacent to cecum and ileum; appendix not seen separately
  - Density ~ 34HU
  - Suggestion of rim-enhancement of cystic wall
  - No surrounding stranding suggestive of inflammation
  - Omental caking and ascites

➤ Officially read as: ‘*could represent an appendix mucocele*’
Appendiceal Mucocele - Definition

- Appendiceal lesion characterized by
  - Appendiceal lumen dilation
  - Mucosal lining alteration
  - Hypersecretion of mucus
  - Potential for extension outside the appendix

➤ This definition is problematic since it is descriptive and does not convey information about the primary underlying disease

Higa et.al. Cancer 1973
Appendiceal Mucocele - Definition

• Histologically mucoceles can be divided into
  – Mucosal hyperplasia (25%)
  – Mucinous cystadenoma (63%)
  – Mucinous cystadenocarcinoma (12%)
  – Retention cysts have also been described

• Malignancy of mucoceles has been variably defined by
  – Histologic type of epithelial cells in resected specimen
  – Dissection of the appendiceal wall by mucin
  – Presence of epithelial cells in mucin if there has been egress into the peritoneal cavity

Lo and Sarr Hepatogastroenterology 2003; Higa et.al. Cancer 1973
Incidence and Diagnosis

- Frequency: 0.1% - 0.4% of all appendectomy specimens show findings consistent with mucocele
- F:M = 1.2-4:1
- Age at diagnosis 50’s-60’s
- 49% symptomatic
  - Malignant appendiceal mucoceles more frequently become symptomatic
  - Common presenting complaints are
    - abdominal pain (27%)
    - palpable abdominal mass (16-25%)
    - abdominal distention (14%)
    - weight loss (10%)
- Laboratory analysis may show elevated CEA, WBC, and ESR

Differential Diagnosis

• Intraperitoneal
  – Other appendiceal neoplasm (lipoma, fibroma, neuroma, carcinoid, lymphoma)
  – Appendicitis
  – Cyst (ovarian, mesenteric, omental)
  – Mesenteric hematoma or tumor
  – Abdominal abscess
  – Hydrosalpinx

• Retroperitoneal
  – Inflammation
  – Tumor
  – Hemorrhage

Horgan et.al. AJR 1984
Imaging of Mucoceles

• Correct preoperative diagnosis is key for appropriate surgical intervention (more on this later)

• Diagnostic imaging modalities used in preoperative diagnosis include
  – US
  – X-ray
  – Barium enema
  – Endoscopy
  – CT
Trans-abdominal US showing:

- Elongated, unilocular cystic structure with internal echoes
- Enhanced through-transmission suggested cyst is fluid-filled
- Indistinct cystic wall

Pickhardt et.al. RadioGraphics 2003
Mucocele on US – Companion Patient 2

Trans-abdominal US showing:
- Elongated, unilocular cystic mass (M) with internal echos
- No distinct cyst wall
- No posterior or lateral shadowing

Sasaki et.al. Abdom Imaging 2003
Mucocele on US – Companion Patient 3

Trans-abdominal US showing:

- Cystic mass with echogenic layers ‘onion-skin’ sign

Caspi et.al. J Ultrasound Med 2004
Mucocele on X-ray – Companion Patient 4

Coned-down plain film of RLQ showing:

- Round mass (Arrowheads)
- Curvy-linear calcifications (White arrows)
Mucocele on X-ray – Companion Patient 5

Plain film of RLQ showing:

• Rounded mass suggested by wall calcifications (Arrows)

Higa et.al. Cancer 1973
Mucocele on Barium Enema – Companion Patient 6

Single contrast barium enema showing:

- Smooth, broad-based filling defect (Arrowhead) in the medial cecum adjacent to the ileocelecal valve

Pickhardt et.al. RadioGraphics 2003
Mucocele on Barium Enema – Companion Patient 7

Air-barium double contrast enema showing:

- Smooth, submucosal filling defect (M) in the medial cecum

Pickhardt et.al. RadioGraphics 2003
Mucocele on Colonoscopy – Companion Patient 6

Colonoscopy, performed on patient 6 seen previously, showing:

- Bulbous, smooth submucosal lesion (M) protruding into the cecum
- Mass’s movement with respiration is thought classic for a mucocele

Pickhardt et.al. RadioGraphics 2003
Mucocele on Colonoscopy – Companion Patient 8

Colonoscopy showing:

- Bulbous, smooth submucosal lesion protruding into the cecum at the site of the appendiceal orifice
- Appendiceal orifice seen at the center of the mound is the ‘volcano’ sign considered classic for a mucocele

Zanati et.al. Gastrointest Endosc 2005
CT scan, of patient 6 seen previously, showing:

- Cystic lesion (Arrowhead) adjacent to cecum extending into the peritoneal cavity (Arrow)
- Density range for mucoceles seen on CT ~ 10-45HU
- Note absence of peri-appendiceal inflammation or abscess

Pickhardt et.al. RadioGraphics 2003
Mucocele on CT – Companion Patient 9

CT scan showing:

• Low-density, well-capsulated mass (Arrow) adjacent to the cecum in the expected location of the appendix

Sasaki et.al. Abdom Imaging 2003
Imaging of Mucoceles – Summary of Findings

- **US** – Elongated, unilocular cyst-like mass with internal echos; indistinct wall; ‘onion-skin’ sign may be pathognomonic
- **X-ray** – RLQ rounded mass with curvilinear calcification
- **Barium enema** – Smooth, broad-based filling defect in the cecum
- **Endoscopy** – Bulbous, smooth, submucosal lesion protruding into the cecum near site of the appendiceal orifice; ‘volcano sign’ and movement of the mass with respirations are considered classic for appendiceal mucocele
- **CT** – RLQ mass adjacent to the cecum with low-attenuating content (0-45HU) and wall calcification

Why Pre-op Diagnosis?

- Feared complication of appendiceal mucocele, due to any cause, is **PSEUDOMYXOMA PERITONEI**
  - Diffuse, gelatinous, cellular ascites
  - Origin thought to be
    - Dissemination of mucinous cells from appendiceal mucocele due to rupture of appendix or metastatic spread OR
    - Neoplastic transformation of peritoneum following mucinous metaplasia of mesothelium
  - Often fatal without treatment as it causes intestinal obstruction

Pseudomyxoma Peritonei on US – Companion Patient 10

Rectal ultrasound showing:

- Thick, gelatinous fluid (F) in the pouch of Douglas

Khan et al. Ultrasound Obstet Gynecol 2002
Pseudomyxoma Peritonei on CT – Companion Patient 11

CT scan showing:
- Diffuse intraperitoneal locules with mass effect on adjacent bowel
- Bowels do not float centrally
- Omental caking is present

Pickhardt et.al. RadioGraphics 2003
Pseudomyxoma Peritonei on CT – Companion Case 12

CT scan showing:

- Scalloping of solid organs by mucinous implants
- Septal calcifications in mucinous fluid (Arrowheads)

Pickhardt et al. RadioGraphics 2003
Imaging of Pseudomyxoma Peritonei – Summary of Findings

• US – Thick gelatinous fluid; usually not mobile with maneuvers; fluid septations may be seen
• CT – Increased abdominal girth; diffuse intraperitoneal locules; mass-effect and distortion of bowel; scalloping of surfaces of solid organs

Treatment

**Mucocele Diagnosed Pre-operatively?**

<table>
<thead>
<tr>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
</table>
| Perform laparotomy  
- To prevent mucocele rupture  
- To allow thorough examination of peritoneal cavity for mucinous fluid | If employing laparoscopic approach, convert to laparotomy |

- Appendectomy typically sufficient  
- If fluid is present  
  - Harvest all mucinous fluid  
  - Submit to pathology for examination for epithelial cells
  
If epithelial cells are present  
- Diagnose pseudomyxoma peritonei  
- Refer patient for  
  - debulking surgery (complete resection of gelatinous masses, greater omentum, major viscera, as appropriate)  
  - Intraperitoneal chemotherapy (mitomycin C +/- 5-fluorouracil)

---

Outcomes

• 91-100% survival after resection of mucocele due to mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma if not complicated by pseudomyxoma peritonei

• 25-33% survival in presence of pseudomyxoma peritonei

• Debulking surgery with intraperitoneal chemotherapy yields 3 year survival between 61-86% with an associated 35% risk of morbidity including bowel perforation, fistula formation and anastomotic leak

Summary

- Appendiceal mucoceles are rare and are often found incidentally; incorrect intraoperative handling may lead to major complications.
- Suggestive and characteristic imaging findings can help establish the pre-operative diagnosis of mucoceles highlighting the role radiologists play in pre-operative planning and in ensuring good patient outcomes.
Bibliography

• Dachman AH; Lichtenstein JE; Friedman AC. Mucocele of the appendix and pseudomyxoma peritonei. AJR (1985) 144: 923-929.
• Higa E; Rosai J; Pizzimbono CA; Wise L. Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix; a re-evaluation of the appendiceal mucocele. Cancer (1973) 32: 1525-1541.
• Horgan JG; Chow PP; Richter JO; Rosenfield AT; Taylor KJW. CT and sonography in the recognition of mucocele of the appendix. AJR (1984) 143: 959-962.
• Landen S; Bertrand C; Maddern GJ; Herman D; Pourbaix A; deNeve A; Schmitz A. Appendiceal mucolceles and pseudomyxoma peritonei. Surg Gynecol Obstet (1992) 175: 401-404.
• Lo NS; Sarr MG. Mucinous cystadenocarcinoma of the appendix; the controversy persists: a review. Hepatogastroenterology (2003) 50: 432-437.
• Sasaki K; Komatsuda T; Suzuki T; Konno K; Ohtaka M; Sato M; Ishida J; Sakai T; Watanabe S. Appendiceal mucocele: sonographic findings. Abdom Imaging (2003) 28: 15-18.
• Zanati SA; Martin JA; Baker JP; Streutker CJ; Marcon NE. Colonoscopic diagnosis of the mucocele of the appendix. Gastrointest Endosc (2005) 62: 452-456.
Acknowledgement

Thank you:

• Karen Lee, MD – for giving me great cases
• Gillian Lieberman, MD – for directing and teaching a rotation I wish I had taken earlier
• Pamela Lebkowski – for outstanding support and organization
• Larry Barbaras