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Case Report and Review of the Literature

Laparoscopic Hand-Assisted Right Colectomy for Appendiceal Mucinous Cystadenoma: A Case Report and Literature Review

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ABSTRACT

Background: A mucocoele of the appendix is defined as a cystic luminal distention of the appendix caused by mucin. Mucocoeles are an uncommon pathology and account for 0.2 to 0.4% of appendectomy specimens. Cystadenomas are the most common cause of mucocoeles, representing up to 84% of the cases. The average age of presentation for mucocoeles is 50 years. The clinical presentation for cystadenomas usually includes chronic or acute right lower abdominal pain, although they can be asymptomatic in 25 to 50% of the patients. The treatment for mucocoeles is surgical excision, whether an appendectomy or a right colectomy is needed depends on histopathological examination and the cecum's involvement. The 5-year survival rate for benign mucocoeles is 90 to 100%. The most fearful complication for both benign and malignant mucocoeles is pseudomixoma peritonei.

Case report: A 57-year-old male patient presented with the chief complaint of transanal bleeding and anal pain associated with defecation. We performed a colonoscopy due to the patient's age and observed a submucosal tumor of approximately 3cm located in the cecum at the level of the appendiceal orifice. Biopsies of the region affected by the tumor revealed normal colonic mucosa. A CT scan showed a 2cm lesion dependent of the cecum. A laparoscopic hand-assisted right colectomy with oncological technique and a side-to-side ileotransverse mechanical anastomosis was performed. The final pathology report revealed an appendiceal mucinous cystadenoma with extension to the cecum. No further surgical intervention or adjuvant therapy was recommended.

Highlights

A mucocoele is a cystic luminal distention of the appendix caused by mucin.

Mucocoeles account for 0.2 to 0.4% of appendectomy specimens.

Surgical treatment depends on the dimensions, histology, and clinical presentation.

Open surgery was indicated, nevertheless the laparoscopic approach is now accepted.

Pseudomixoma peritonei is the most fearful complication for mucocoeles.

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Introduction

The term mucocoele of the appendix implies a cystic luminal distention of the appendix caused by the accumulation of mucin [1]. Mucocoeles can be caused by cystadenomas, cystadenocarcinomas, mucosal hyperplasia,

or retention cysts [1-4]. Mucocoeles account for 8 to 10% of appendiceal tumors [2-5]. Appendectomy specimens show mucocoeles in 0.2 to 0.4% of the cases [1-13]. Cystadenomas are responsible for 63 to 84% of mucocoeles while cystadenocarcinomas account for 11 to 20% [1, 6, 11]. Appendiceal mucocoeles present with synchronous colon cancers in 19.5

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to 21.4% of the patients [8]. The mean age of presentation for mucocoeles is 50-years [2, 4, 8, 10]. In 25 to 50% of the cases cystadenomas are asymptomatic and diagnosed as an incidental finding during radiological examination or surgical intervention [2-4, 6, 8, 11-15]. The usual clinical presentation includes acute or chronic right lower abdominal pain (2,6,11,14). The treatment of choice is appendectomy or right colectomy depending on the histopathological diagnosis and the tumor's extension [2, 7, 16]. Benign mucocoeles have a 5-year survival rate of 90 to 100% [2, 7, 8, 10, 16]. Pseudomixoma peritonei (PMP) is the most fearful complication for benign and malign mucocoeles and results from the spillage of mucin secondary to appendiceal perforation [1, 9, 14]. The 5-year survival rate for PMP is 25% [3, 7].

Case Report

A 57-year-old male patient presented with the chief complaint of transanal bleeding and anal pain associated with defecation. On physical examination the patient was well hydrated, the abdomen was flat, with normal bowel sounds, and perianal inspection showed hemorrhoidal disease grade II and III. Rubber band ligation was performed without complications. We performed a colonoscopy and observed a submucosal tumor of approximately 3cm located in the cecum at the level of the appendiceal orifice (Fig. 1). Several biopsies of the affected region were taken nevertheless the pathology report revealed normal colonic mucosa. A CT scan of the abdomen and pelvis with oral, intravenous, and rectal contrast revealed a focal heterogeneous lesion, dependent of the cecum with a diameter of 2cm and well-defined borders (Fig. 2). Blood count, blood chemistry, carcinoembryonic antigen (CEA), 5-hydroxyindoleacetic acid (5-HIAA), and chromogranin A were within normal parameters.

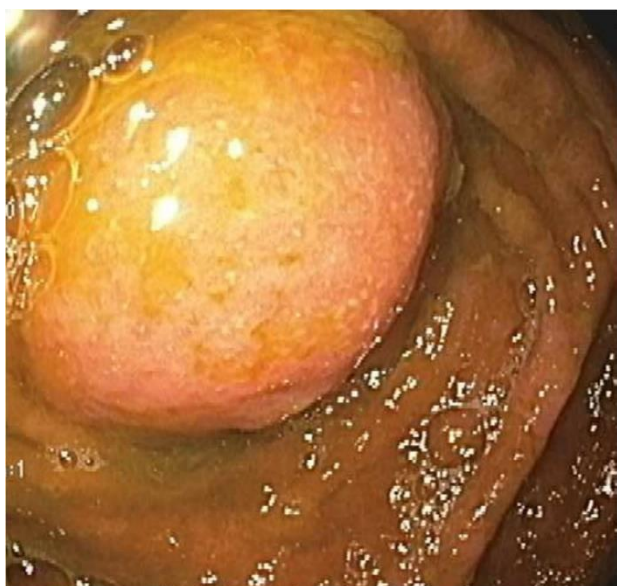


Figure 1: Colonoscopy showing a submucosal tumor in the cecum at the level of the appendiceal orifice.

An exploratory laparoscopy was performed, in which we identified a small amount of periappendiceal free serous fluid (Fig. 3 & Fig. 4). No hepatic or peritoneal implants were observed. Intraoperative cytologic examination of the fluid was negative for epithelial cells. We performed a laparoscopic hand-assisted right colectomy with oncological technique

(Fig. 5) and a side-to-side ileotransverse mechanical anastomosis. The patient had a benign postoperative course and was discharged at the sixth day after surgery. Final pathology report revealed an appendiceal mucinous cystadenoma with extension to the cecum (Fig. 6). No further surgical intervention or adjuvant therapy was recommended.



Figure 2: CT scan of the abdomen and pelvis with oral, intravenous, and rectal contrast showing a focal heterogeneous lesion dependent of the cecum. (A) Axial. (B) Sagittal.

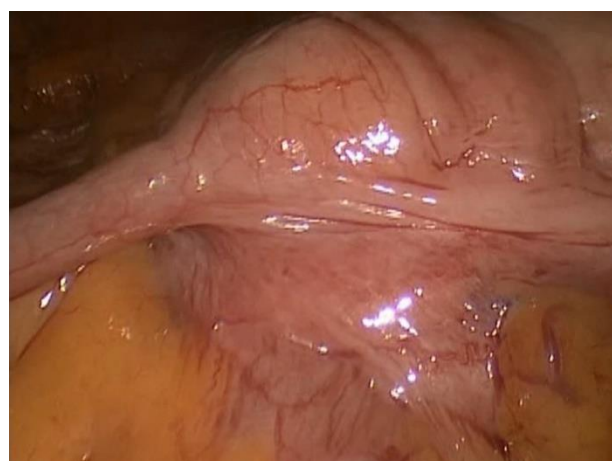


Figure 3: Laparoscopic image of the cecum and the appendix.



Figure 4: Laparoscopic image showing a small amount of periappendiceal free serous fluid.

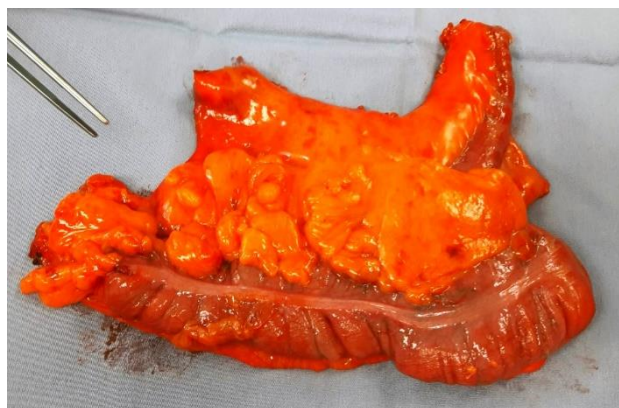


Figure 5: Right colectomy with oncological technique specimen.



Figure 6: Appendiceal mucinous cystadenoma with extension to the cecum.

Discussion

Appendiceal tumors represent 0.5% of all gastrointestinal neoplasms and are found in 1% of appendectomy specimens [5]. Mucocoeles account for 8 to 10% of appendiceal tumors, representing between 0.2 to 0.4% of all appendectomies [1-13]. Appendiceal mucocoele was first described by Rokitsky in 1842 and was formally named by Feren in 1876 [3, 4, 12, 17]. A mucocoele is defined as an appendiceal cystic luminal distention due to the accumulation of mucin [1, 3, 12]. Mucocoeles can be secondary to mucinous cystadenoma, cystadenocarcinoma, mucosal hyperplasia, or retention cysts [1-4, 12]. Mucinous cystadenomas account for 63 to 84% of mucocoeles and are characterized by a glandular or papillary pattern with mucus production and dysplastic epithelium similar to colon adenomatous polyps [1-3, 12]. Microscopically, mucinous cystadenomas are sessile lesions that circumferentially involve the appendiceal mucosa, composed of mucin-rich epithelium [15]. Appendiceal mucinous cystadenomas can cause luminal distention of up to 6cm and present spontaneous perforation in 20% of the cases [13, 15]. Mucosal hyperplasia and retention cysts represent 5 to 25% of mucocoeles. Mucinous cystadenocarcinomas are responsible for 11 to 20% of mucocoeles and differ from cystadenomas in the presence of glandular and stromal invasion [4,12].

The mean age of occurrence for mucocoeles is 50-years and appear to be more frequent in women [2, 4, 8, 10]. Cystadenomas may be asymptomatic in 25 to 50% of the cases [2-4, 6, 8, 11-15]. The most common clinical manifestation is acute or chronic right lower abdominal pain [4, 6, 11, 14]. Abdominal pain, similar to acute appendicitis, is present in 30 to 64% of the cases [11]. A palpable mass located in the right iliac fossa is found in approximately 50% of the patients [2, 6, 11, 14]. Less common clinical manifestations include nausea, vomiting, intussusception, lower gastrointestinal bleeding, change in bowel habits, weight loss, intestinal obstruction, unexplained anemia, generalized peritonitis, sepsis, and urologic symptoms due to right ureteral obstruction [2, 3, 5, 8, 11, 13, 14, 15, 18].

Approximately 40% of mucocoeles are diagnosed during surgery for suspected acute appendicitis [3]. Mucinous cystadenomas are diagnosed before surgery in only 15 to 29% of the patients [8]. Abdominal CT scan is the study of choice for the diagnosis of mucocoele [2, 5]. The typical finding is a well circumscribed cystic thin walled mass that communicates directly with the cecum [3, 4, 12]. Abdominal CT scan may show curvilinear mural calcifications, suggestive of mucocoele, in 50% of the patients [3, 4, 12, 13]. The differentiation between benign and malign mucocoeles with CT findings has low reliability but wall irregularity and soft-tissue thickening suggest malignancy [4]. Abdominal ultrasound may identify multiple echogenic layers along a dilated appendix resembling "onion skin" circles, which are pathognomonic for mucocoele [2, 3, 8, 12, 13]. Colonoscopy may show the "volcano sign", pathognomonic for mucocoele, consisting of a soft erythematous mass with the appendiceal orifice as a central crater showing mucin discharge [3, 8, 12]. Synchronous colonic neoplastic lesions are found in 20% of the patients [2, 12, 16].

Surgical treatment depends on the dimensions, histology, and clinical presentation [16]. The standard treatment used to be right colectomy [13, 18, 19]. Benign mucocoeles smaller than 2cm can be treated with simple appendectomy [16]. Indications for right hemicolectomy are necessity to clear the tumor or achieve complete cytoreduction, appendiceal or ileocolic lymph node involvement, or a non-mucinous neoplasm [19]. If the extension of the tumor is in doubt, cryostat sectioning of the surgical margin is indicated [13]. Oncological resection is indicated in case of appendiceal perforation or extravasation of mucin [18]. Historically, laparoscopic surgery conversion to open surgery was indicated, nevertheless the laparoscopic approach is now accepted [1, 2, 5, 18]. Benign mucocoeles reach a 10-year survival rate of 91% [3, 7, 16]. Cystadenocarcinomas have a 10-year survival rate of less than 10% [16].

PMP refers to the accumulation of thick, gelatinous material on intraperitoneal surfaces either because of the localized rupture of a benign mucocoele or secondary to the diffuse proliferation of viable neoplastic cells in the peritoneum [2, 15]. PMP is estimated to develop in 2% of appendiceal mucocoeles and 20% of appendiceal mucinous adenocarcinomas [17]. This condition has a male: female ratio of 1:3 and occurs in 2 out of 10,000 laparotomies [15]. Approximately 3 to 10% of PMP cases have ovarian origin while 82% are of appendiceal origin [17]. Any liquid found outside the appendix should be examined in search for epithelial cells to diagnose PMP [13]. In case of positive cytology for epithelial cells cytoreductive surgery with hyperthermic intraperitoneal chemotherapy (HIPEC) is indicated [12]. HIPEC is performed with mitomycin or oxaliplatin heated to 42°C for 60 to 120 minutes [12].

Patients with PMP limited to the peritoneum and absence of distant metastases who are treated with cytoreductive surgery associated with HIPEC reach a 5-year survival rate of 50 to 96% [12, 15]. The 5-year survival rate for PMP in patients that are not candidates for curative resection is 25% [3, 7].

The follow-up of patients with diagnosis of mucocele is dictated by the final pathology report [2, 12]. In case of simple or hyperplastic mucoceles there is no need for follow-up, cystadenomas are followed-up as colonic adenomatous polyps, with serial colonoscopies, and cystadenocarcinomas as colonic adenocarcinomas with serial colonoscopies and CEA [2, 12]. CEA and CA 19-9 can be used for follow-up in patients diagnosed with PMP [20]. The true positive rate for recurrent PMP is 56.1% for CEA, 67.1% for CA 19-9, and 78.8% if a positive test is defined as either an elevated CEA or CA 19-9 [20].

Conclusion

Our patient presented with the chief complaint of transanal bleeding and anal pain associated with defecation. We advised him to get a colonoscopy due to his age and the presence of transanal bleeding. During the colonoscopy we excluded the possibility of a synchronic colonic neoplasm. We performed a CT scan with oral, intravenous, and rectal contrast which revealed a focal heterogeneous lesion, dependent of the cecum with a diameter of 2cm and well-defined borders. CT scan has a low reliability differentiating between benign and malign mucoceles, so we planned an oncological resection. We performed an exploratory laparoscopy to verify the absence of hepatic and peritoneal implants and identified free fluid. The cytological examination of the free liquid was negative for epithelial cells, so we ruled out the diagnosis of PMP. We decided to perform a laparoscopic hand-assisted right colectomy with oncological technique because the tumor invaded the base of the appendix. Final pathology report revealed an appendicular mucinous cystadenoma with extension to the cecum so there was no need for further treatment. We will perform annual colonoscopies as follow-up.

Conflict of interest statement

The authors declare no potential conflict of interests.

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Author's Contributions

All authors discussed the results and contributed to the final manuscript.

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