Mucinous Cystadenoma of the Appendix

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Abstract

An appendiceal mucocele found in approximately 0.2%-0.4% of all appendectomies [1-6]. Its rarity makes diagnosis and management challenging. Treatment options differ based on cytology, resection margins, and whether or not the mucocele has perforated. Although the average age at diagnosis is > 50 years [6], here is presented a 36 year-old female with a large appendiceal mucinous cystadenoma who underwent a laparoscopic converted to open appendectomy. Also provided is a brief review of the pathology, diagnosis, and management of mucinous cystadenoma of the appendix.

ABBREVIATIONS

CT: Computed Tomography; HIPEC: Heated Intraoperative Intraperitoneal Chemotherapy

INTRODUCTION

Although technically benign, mucinous cystadenoma of the appendix can be a devastating condition if not treated optimally. It is one of the four histologic types of appendiceal mucocele: mucinous cystadenoma, mucosal hyperplasia, simple (retention) cyst and mucinous cystadenocarcinoma. These subtypes are found in decreasing frequency of 52%, 20%, 18% and 10% respectively [7]. Classification helps to define biologic behavior rather than to direct initial management as all four types require surgical resection. However, if any type is found to be perforated (whether spontaneously or during the course of resection), the management may differ drastically, ranging from resection alone to cytoreductive surgery and intraperitoneal chemotherapy.

CASE PRESENTATION

A previously healthy 36 year-old African American female presented with a 3-day history of right lower quadrant abdominal pain associated with anorexia, nausea and vomiting. The pain had started in the periumbilical area and migrated to the right lower quadrant in the 12 hours before presentation. She denied fever or chills, but had no flatus or bowel movement during the prior 24 hours. Her medical history was unremarkable other than having had a Cesarean section. Vital signs on presentation were normal however her physical examination revealed a patient in mild distress with right lower quadrant abdominal tenderness and a suggestion of a mass in that location upon palpation. There was no guarding or rebound tenderness. Laboratory data showed a mild leukocytosis.

A CT scan of the abdomen and pelvis (Figure 1) revealed what appeared to be a dilated, fluid-filled small bowel loop measuring 12.3 x 4.8 cm in its greatest dimension, raising suspicion of a small bowel volvulus, torsed Meckel's diverticulum, or appendiceal mucocele. The patient consented to a diagnostic laparoscopy and possible laparoscopic or open resection. She was taken to the operating room where, upon laparoscopic exploration, a large cystic mass was found in the right lower quadrant (Figure 2). Further inspection revealed that the mass was a much dilated appendix with minimal inflammation that was not adherent to other structures.

Due to its size and to avoid perforation upon manipulation or extraction, the procedure was converted to a mini-laparotomy. The appendix and cecum were easily delivered into the incision. There was no involvement of the appendiceal base, cecum, or ileocecal junction. No lymphadenopathy was appreciated and there was no perforation, nor mucin in the peritoneal cavity. The ovaries appeared normal, and there was no Meckel's diverticulum. A simple appendectomy was then performed. The patient recovered uneventfully and was discharged the following day. Pathologic examination confirmed our suspicion of mucinous cystadenoma of the appendix with clear margins and no malignant features. The specimen measured 15.0 cm in length and 5.0 cm in diameter. The wall thickness ranged from 0.1 cm to 0.2 cm. By the time of her post-operative visit the patient had fully recovered without residual symptoms.

DISCUSSION

Mucinous cystadenoma is a rare but significant entity, found in approximately 0.2%-0.4% of all appendectomy specimens [1-6]. It is more prevalent in patients > 50 years of age, possibly more common in females [6]. It may be challenging to diagnose this condition preoperatively, since smaller lesions are often asymptomatic and larger lesions may mimic normal or dilated small bowel on imaging studies. Surgical treatment requires the utmost care to prevent perforation and leakage of mucin into...
the peritoneal cavity, which can lead to the often devastating pseudomyxoma peritonei.

Presenting symptoms often mimic those of acute appendicitis but on rare occasions, they include signs of obstruction from appendiceal intussusception or volvulus, gastrointestinal bleeding, hydronephrosis from right ureter obstruction, an acute abdomen from mucocele rupture, or abdominal distention from mucinous ascites. Symptoms are absent in 25% to 50% of patients, who are incidentally found to have a mucocele of the appendix on imaging or at the time of exploration [3-6]. This patient had the classical symptoms of appendicitis but in addition her examination suggested a right lower quadrant mass, which is present in approximately 14% of patients [3].

The diagnostic imaging of choice is CT, although ultrasound maybe utilized. On CT, a well-defined low attenuationmass with or without calcifications can be found, similar to findings on ultrasound [3]. Enhancing wall nodularity of the mucocele suggests but does not confirm malignancy [2,3]. Colonoscopy may be unremarkable but the appearance of an extrinsic mass that is in fact the mucocele pushing into the cecum has been reported [2]. The patient in this report did not have the typical appearance of a well-encapsulated mass but rather imaging suggested a blind ending dilated bowel loop suspicious for a Meckel’s diverticulum, though appendiceal mucocele was also in our differential.

The treatment of choice is complete resection of the mucocele. This may require partial cecectomy or ileocecectomy if the mucocele involves the base of the cecum. Both laparoscopic and open techniques have been reported. The open approach may be preferred to minimize the risk of iatrogenic perforation, which could result in mucinous carcinomatosis or pseudomyxoma peritonei. This would drastically alter the patient’s future

Figure 1 CT series of fluid filled mass indicated by white arrows.

Figure 2 Intraoperative view of appendiceal mucocele.

Figure 3 Appendix delivered through incision prior to resection.
course and even survival, especially in the case of a mucinous cystadenocarcinoma. Nonetheless, there have been numerous reports of laparoscopic approaches [5,6,8], including single-port laparoscopic resection [4]. If laparoscopy is used, the incision must be large enough to safely remove the specimen without risk of rupture or spillage. With either approach, exploration requires a thorough examination of the entire peritoneal cavity for any mucoid fluid accumulation, particularly in the retrohepatic space or deep in the pelvis. If mucinous peritoneal fluid is found, it should be sent for cytological examination for epithelial cells. Laparoscopy may be used as the initial approach to optimize complete visualization of the peritoneal cavity first, thus allowing use of the smallest incision necessary to extract the specimen. In the current report, we started laparoscopically and converted to an open procedure due to the large size of the mass and its tenseness, which caused difficulty in safely grasping it with laparoscopic instruments. The incision made was just large enough to deliver the appendix manually. There was no evidence of peritoneal implants, mucinous fluid, or abnormalities of the serosal surface of the appendix.

The extent of resection is often mentioned in discussions of appendiceal mucocoele. If there is concern for involvement of the margin of resection or if there is evidence of contiguous involvement of the cecum, cecotomy is recommended [2,3] provided that the mucocoele has not perforated and that no lymph node involvement is present. Perforation or intraperitoneal dissemination of a mucinous cystadenocarcinoma is best treated with heated intra operative intraperitoneal chemotherapy (HIPEC) in addition to cytoreductive surgery [3]. Although pseudomyxoma peritonei has been mostly associated with mucinous cystadenocarcinoma, there are reports of perforated mucinous cystadenoma resulting in mucinous carcinomatosis [2]. Even in benign conditions where no further therapy is undertaken, perforation of the lesion or presence of peritoneal mucin requires close follow up and serial imaging to allow for early intervention if mucin is accumulating without symptoms. With pain or bowel obstruction, peritoneal implants should be suspected. The reader is referred to the clinical pathway presented by Dhage-Ivatury and Sugarbaker [3] for a more thorough presentation of treatment options.

The prognosis of non-perforated mucinous cystadenoma is excellent with resection alone and long-term follow up is not necessary [3]. However, survival decreases sharply for perforated cystadenocarcinomas or even cystadenomas. Cytoreductive surgery and HIPEC may improve survival [9] despite these treatments, 5-year survival ranges from 25% to 96% [2-3]. Since appendiceal mucocele can be associated with concurrent malignancy elsewhere in the abdomen, this finding should prompt further evaluation, particularly of the colon and adnexa [6].

In summary, although mucinous cystadenomas of the appendix are a rare and mostly benign entity, careful initial management is paramount. Whether treated by laparoscopic or open resection, the primary goals are to achieve complete resection and to maintain the integrity of the specimen without leakage. Evidence based guidelines are lacking due to the low incidence of this condition. When rupture of the cystic lesion is known or suspected preoperatively, specially consultation with a surgical oncologist is advised prior to any surgical intervention.

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REFERENCES