

Appendiceal mucocele

Contraindication to laparoscopic appendectomy

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Abstract. Indications and contraindications to laparoscopic surgery continue to be refined. Laparoscopic appendectomy for acute appendicitis is frequently selected by patients and surgeons, and clinical studies show it to be a reasonable alternative. In this case study, laparoscopic surgery was used to resect an appendiceal mucocele caused by a non-perforated mucinous adenocarcinoma. Implants of mucinous tumor were found widely disseminated on peritoneal surfaces at laparotomy 9 months later. As a result of this case study, the authors suggest that when an appendiceal mucinous tumor is encountered at laparoscopy, a special situation requiring totally atraumatic appendectomy is indicated. This clinical situation should be considered an indication for conversion to open appendectomy. All appendiceal tumors, including the most benign-appearing adenomas, can result in diffuse peritoneal implantation. This is the first report of an appendiceal mucinous tumor resected by laparoscopy associated with subsequent diffuse peritoneal carcinomatosis. This patient presentation reaffirms that dissemination of cancer may be associated with laparoscopic resection of structures containing a malignancy.

Key words: Appendix — Appendicitis — Mucocele — Pseudomyxoma peritonei — Peritoneal carcinomatosis — Intraperitoneal chemotherapy

As laparoscopic surgical procedures have been developed and integrated into surgical practice, they have gradually replaced their open surgery counterparts because of surgeon and patient preference. Results from many randomized studies confirm several advantages for the laparoscopic pro-

cedures, but have not led to restrictive recommendations regarding optimal utilization [1, 19, 23, 31].

As studies show, laparoscopic appendectomy is a treatment option for excision of an acute appendix [5, 30]. Although mucocele of the appendix is encountered in only 0.2 to 0.4% of appendectomies, it may present a special precaution to the laparoscopic surgeon. In this case report, a patient with an appendix mucocele who underwent laparoscopic resection developed widespread mucinous peritoneal carcinomatosis. Manipulation of the mucus-filled appendix or spillage of its contents may have caused peritoneal surface contamination leading to mucinous peritoneal carcinomatosis. When a mucinous appendiceal tumor is encountered, special precautions are required to remove this structure without trauma, especially without grasping the appendix. The authors suggest conversion to an open appendectomy.

Case report

A 37-year-old female presented with a 3-day history of right lower quadrant abdominal pain without fever or nausea. Observation was recommended, but the following day her pain worsened, and she was admitted to the hospital with the working diagnosis of acute appendicitis. Laparoscopy was elected to explore her abdomen.

A 12-mm umbilical and two 5-mm suprapubic trocars were placed. A phlegmon surrounding the right colon was found, with no free fluid present in the peritoneal cavity. Mobilization of the cecum revealed an indurated, bulbous appendix in a retrocecal location, with no signs of perforation. The appendix was mobilized, and then an endoscopic stapling device was used to transect its base. The specimen was retrieved from the abdominal cavity in a bag through the umbilical port. The abdomen was copiously irrigated with saline before the trocars were removed. Gross examination of the specimen showed a 2.3-cm bulge in the central portion of the appendix, with no evidence of perforation. Histopathologic study showed mucinous adenocarcinoma infiltrating the wall of the appendix, appearing to arise in a villous adenoma. The mucus dissected up to the serosa of the appendix, filled its lumen, and contained signet-ring cells (Fig. 1). Infiltrating mucinous adenocarcinoma penetrating into, but not through, the serosal surface was the final pathologic diagnosis.

On the fourth postoperative day, the patient was taken back to the operating room for exploratory laparotomy. There were no ascites, free

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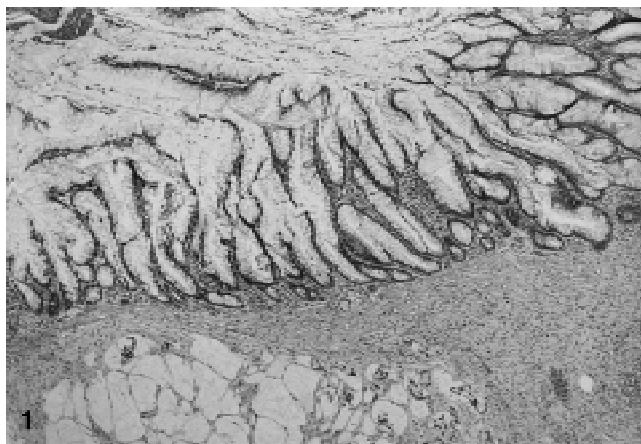


Fig. 1. Appendix showing mucin-filled lumen lined by villous adenoma. The muscular wall reveals an infiltrating mucinous adenocarcinoma characterized by pools of dissecting mucin containing malignant glands and signet-ring cells (H&E, $\times 100$).

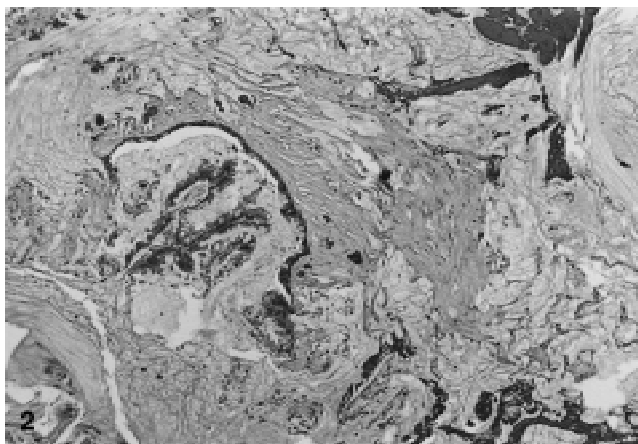


Fig. 2. Peritoneal implant revealing dissecting mucin with atypical mucinous epithelium forming complex glands. This is a characteristic feature of mucinous peritoneal carcinomatosis (H&E, $\times 100$).

mucin, peritoneal implants, or visceral masses. Right colectomy was followed by an ileocolic anastomosis, excision of trocar sites and umbilicus, cytology washings, and peritoneal biopsies. All specimens were reported negative for malignant cells.

Eleven months later a rise in serum carcinoembryonic antigen to 27 ng/ml was noted. A computed tomography (CT) scan of the abdomen showed perihepatic fluid, globular masses in the right gutter, and a large pelvic mass. Physical examination was unremarkable except for globular masses palpated on either side of the uterine cervix during rectal digital exam. A midline laparotomy was performed, excising the previous laparotomy scar. On entering the abdomen, diffuse mucinous tumor was observed throughout the peritoneal cavity, excluding the left upper quadrant (Fig. 2). Complete cytoreduction could not be achieved because of extensive small bowel disease. Intraperitoneal chemotherapy was begun with 19 mg of mitomycin C on the first postoperative day, then 5-fluorouracil (1,050 mg/day) on the following 5 days. All chemotherapy was delivered intraperitoneally in 1,500 cc of 5% dextrose peritoneal dialysis solution. The patient was discharged on the 21st postoperative day. The patient, currently being treated with systemic chemotherapy, is a candidate for second-look surgery 9 months after cytoreduction.

Discussion

Mucocele of the appendix is an infrequent entity, with a reported incidence ranging from 0.07 to 0.3% of all appendectomies [15, 18] and a female-to-male ratio of 3–4 to 1 [15, 10]. The external appearance is gross enlargement of an appendix whose lumen is distended by mucin. The causes are heterogeneous and include retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma [15, 27]. In a recent review, Ronnett and colleagues [27] indicated that all such lesions, except for the rare acellular retention cyst, if given access to peritoneal surfaces, may cause disseminated peritoneal tumors containing epithelial cells. The crucial issue in this clinical setting is that all appendiceal neoplasias, both histologically benign and malignant, can result in diffuse mucinous peritoneal tumors. These mucinous peritoneal tumors are usually lethal even with extensive treatment [8]. It must be concluded that all reasonable surgical maneuvers are necessary to prevent peritoneal dissemination of epithelial cells from within an appendiceal mucinous tumor.

Laparoscopic dissection, grasping of the appendix

specimen, pneumoperitoneum, and transport of the specimen through the abdominal wall may contribute to peritoneal dissemination of an appendiceal mucinous tumor [12]. Spillage of the mucinous contents during removal of the specimen may not have a role in this case because the appendix was isolated in a specimen bag before its transport through the abdominal wall. It is likely that an open surgical approach in this specific case, keeping in mind the retrocecal location of the appendix, would have resulted in a less traumatic dissection and less contact of a tumor-infiltrated serosa with the peritoneal surfaces. Peritoneal seeding should be preventable with atraumatic handling of the cancer specimen. Peritoneal dissemination from a mucocele of the appendix has not been regarded as a problem with open appendectomy. The incidence of disseminated peritoneal mucinous tumors from an open appendectomy must be extremely low or no reports could be discovered from a literature review. It is likely that open appendectomy would have prevented the mucinous peritoneal carcinomatosis and greatly changed the probable outcome for this patient.

Scientific studies comparing curative laparoscopic and open surgery for gastrointestinal malignancies have not been completed as yet [21]. Substantial evidence of unexpected complications from laparoscopic surgery for abdominal and pelvic cancer, namely intra-abdominal and abdominal wall tumor seeding, has been widely reported in recent years [2, 3, 7, 11, 20, 22, 24, 25]. This has led to recommendations restricting laparoscopic colonic resections for curable colorectal carcinoma to prospective studies [29] and even to strongly contraindicating laparoscopic surgery for suspected ovarian [7] and gallbladder carcinoma. [16, 17, 24, 26]. Although many authors have advised that malignant gastrointestinal lesions should not be removed laparoscopically, we have not found any reported case of peritoneal or abdominal wall tumor seeding after laparoscopic appendectomy in the biomedical literature to date.

It would be unwise to draw conclusions regarding the advisability of laparoscopic appendectomy based solely on the reported case, given the low incidence of appendiceal mucinous tumors in the general population compared with

that of true acute appendicitis. Nevertheless, whenever an atypical clinical presentation for acute appendicitis and certain well-described diagnostic image tests [9, 10, 14] cause the surgeon to entertain a preoperative diagnosis of appendiceal mucocele, we concur with Landen [16] that laparotomy should be the surgical approach of choice. In a review concerning appendiceal malignancies, Rutledge and Alexander [28] stated that mucinous cystadenocarcinoma is one of the few instances of malignant appendix neoplasms that can be diagnosed before surgery. Alternatively, if distention of the appendix lumen is observed, laparoscopically suggesting mucocele as a possible diagnosis, conversion to open appendectomy may demonstrate the most prudent surgical judgment.

This case report contributes to the continuing discussion regarding the claimed superiority of laparoscopic appendectomy to open appendectomy. Several studies have found no significant differences in clinical results between both procedures [4, 31] and others have described a higher incidence of postoperative intra-abdominal infections [13], readmissions [29], or prolonged hospital stays after laparoscopic appendectomy compared with open appendectomy. However, laparoscopic appendectomy may provide a better cosmetic result and a shorter convalescence. A recent European consensus conference on laparoscopic appendectomy [6] concluded that surgeons should be aware of this technique's potential dangers when using it. Laparoscopic resection of appendiceal mucinous lesions should be added to this list.

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