Case Report

Apppendiceal Mucocele with Cystadenoma — A Case Report and Literature Review

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Appendiceal mucocele is a rare entity ,encompassing various kinds of pathology, and correct preoperative diagnosis is infrequently achieved in cases presenting as acute right lower quadrant abdominal pain. We report a 76 -year old female with the chief complaint of acute right lower quadrant pain, whose preoperative diagnosis was acute appendicitis. Marked enlargement of the appendix and adhesion to the cecum were noted during operation, and right hemicolectomy was done with the impression of appendiceal mucocele of possible malignant etiology. The postoperative course was uneventful. Pathological examination revealed an appendiceal mucocele with cystadenoma of the appendix. [*J Soc Colon Rectal Surgeon (Taiwan) 2002;13:121-124*]

Appendiceal mucocele, first described by Rokitansky in 1842,[1] indicates the gross enlargement of the appendix due to accumulation of mucoid substance within the lumen. It is the sequela of several kinds of benign and malignant neoplasms. About 23-50% of cases with appendiceal mucocele are asymptomatic, with their incidental identification during surgery, radiological studies, or endoscopic performance for lesions other than the mucocele.[2] Symptoms and signs, if present, are frequently non-specific; together with its rarity, they contribute to the difficulty of achieving correct preoperative diagnosis in cases presenting with acute right lower quadrant pain of abdomen.

Case Report

A 76-year old woman came to the emergency room with the complaint of abdominal pain at right lower quadrant for hours. She had myasthenia gravis with steroid and neostigmine medication for ten years. Abdominal total hysterectomy with bilateral salpingo-oophorectomy had been performed for uterine leiomyoma twelve years ago. No anorexia, body weight loss, bowel habit change, change in stool caliber, or hematochezia were noted in recent months. Physical examination revealed tenderness and rebound pain at right lower quadrant of the abdomen. The hematogram showed marked leukocytosis with neutrophil dominance. Result of urinalysis was unremarkable. Plain standard abdominal radiography showed only some air-filled small bowel loops in the pelvic cavity.

Abdominal exploration was performed under the impression of acute appendicitis. A firm, movable mass with the size of about 7 × 6 cm^2 was palpated at right lower quadrant after general anesthesia. After laparotomy, marked enlargement of the appendix with adhesion to cecal wall was noted. Grossly, there was no mucinous implant or regional lymphadenopathy observed. Right hemicolectomy was performed due to the

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suspicion of appendiceal mucocele with malignant etiology (Fig. 1). No intraoperative appendiceal perforation occurred. The postoperative course was uneventful.

On postoperative pathological examination, the appendix was measured as $9.5 \times 9.0 \times 9.0$ cm^3, with

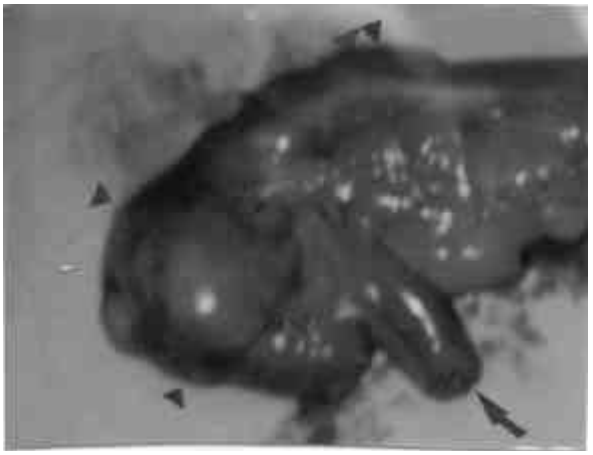

Fig. 1. The resected specimen. The four arrowheads indicate the circumfernce of the mucocele; the arrow indicates terminal ileum, and the double arrowheads indicate the ascending colon.

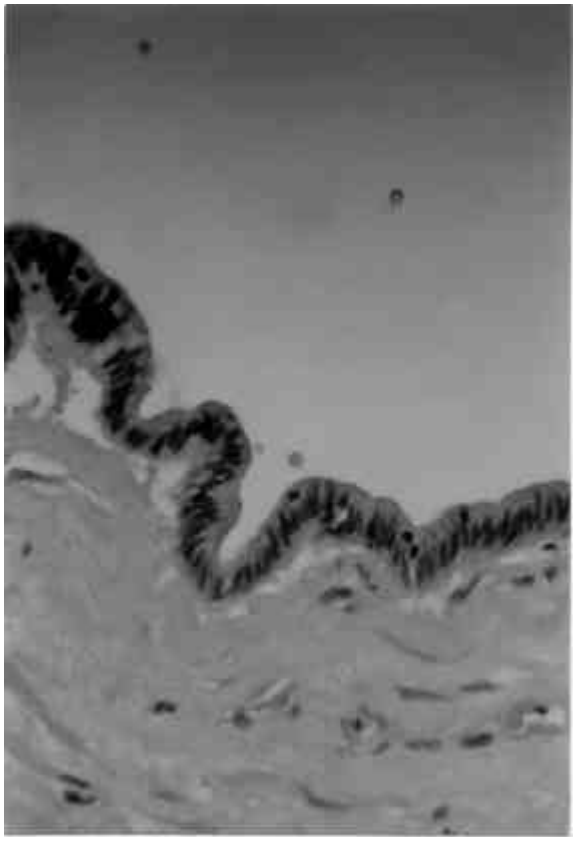

Fig. 2. Microscopically, the appendiceal mucosa showed mild degree of cellular atypia (H&E stain, 400x).

Fig. 3. The arrow indicates luminal mucin. There were calcification spots (arrowheads) occupying some area of epithelial lining. Note also the outer wall heavily infiltrated with inflammatory cells (H&E stain, 100x).

serosal congestion. The appendiceal orifice was edematous but without obstruction. The lumen was severely dilated and contained abundant mucus. The mucosa was extensively coated with fibrinoid exudate. There was no gross tumor in the appendix. Microscopically, the ulcerated mucosal surface was lined with a single layer of columnar or flattened epithelium with mild degree of cellular atypia (Fig. 2). Neither papillary growth nor signs of malignancy were noted. The appendiceal lumen contained mucin and inflammatory exudates (Fig. 3), and the wall was heavily infiltrated with acute and chronic inflammatory cells. The resected colon and terminal ileum showed no remarkable finding. None of the dissected lymph nodes revealed evidence of metastasis. The pathological diagnoses were (1) appendiceal mucocele with mucinous cystadenoma of the appendix and (2) acute suppurative appendicitis.

Discussion

As a rare lesion, appendiceal mucocele accounts for 0.2-0.3% of appendectomies and autopsy series[3]. According to the review study of 60 cases by Aho et al., the mean age of presentation is 55 years (range: 2nd to 9th decade) with a female:male ratio of 4:1.[4] It can be classified into four groups depending on the underlying pathology:[5,6] (1) simple or retention mucoceles resulting from obstruction of the appendiceal outflow. The cause of obstruction includes fecalith, scarring from previous

in flam ma tion, or less com mon, endometriosis. (2) Mucoceles re lated with ep i the lial hy per pla sia. (3) Mucinous cystadenomas of the ap pen dix, which rep re sent the most com mon form of mucoceles, com pris ing 63-84% of the entity.[3] Histologically, they ex hibit mostly villous adenomatous changes with some degree of atypia and marked di la ta tion of the lu men.[6] The pa thol ogy of our pa tient be longed to this group. In ad di tion, twenty percent of cases of mucinous cystadenomas are as so ci ated with perforation.[3] (4) Mucinous cystadenocarcinomas, which ac count for 11-20% of mucoceles and of which 6% are as so ci ated with per fo ra tion.[3,7] If per fo ra tion occurs, it re sults in dis sem i nated intraperitoneal mucinous implants, so-called pseudomyxoma peritonei. There have been reports regarding the association between other tu mors and appendiceal cystadenoma, with adenocarcinoma of the co lon be ing the most com mon, with an in ci dence of about 20%.[6] The in ci dence of as so ci ated ovar ian neo plasm ranges between2-24%.[4] No co lonic or ovar ian tu mor was de tected in our case dur ing op er a tion.

The most com mon symp toms and signs are acute or chronic abdominal right lower quadrant pain (64%)[4] and pal pa ble ab dom i nal mass (50%).[5] In our case, the mass was pal pated only af ter the pa tient was an es the tized and no more mus cle guard ing hin dered thor ough ex am i na tion. If the mass could have been pal pated dur ing the ER stay, sub se quent im age stud ies might have dis closed its na ture be fore op er a tion. The only im age study we per formed was plain ab dom i nal ra di og ra phy. Plain ab dom i nal ra di og ra phy may re veal a soft tis sue shadow and curvilinear cal ci fi ca tion in the right iliac fossa.[8]

When these le sions are iden ti fied pre op er a tively or incidentally during operation for other reasons, they should be re moved be cause of the pos si bil ity of ma lig nancy or per fo ra tion with sub se quent pseudomyxoma peritonei. Ex tremely rarely, pseudomyxoma peritonei also oc curs in cases of per fo ra tion of mucinous cystadenoma, with a better prognosis than those of ma lig nant per fo ra tion.[9] In most pa tients, simple ap pen dec tomy to gether with its mesentery suf fices for an un com pli cated, unruptured mucocele.[3] Nei ther hematogenous nor lymphatic spread has been reported in cases of cystadenocarcinoma without mesenteric or adjacent organ involvement.[3] Right hemicolectomy is sug gested if the in volved appendiceal wall ad heres to or shows signs of in vad ing the ce cum, il eum, or mesentery, as in our case.[4,6] When pseudomyxoma peritonei ex ists, it is im por tant to remove as many gross implants as possible.[10] If the mucocele should be identified during laparoscopic op er a tion for other rea sons, it re mains an is sue of debate con cern ing lap aro scopic re moval of the tu mor due to reports of occurrence of pseudomyxoma peritonei af ter re sec tion of a nonperforated mucinous cystadenoma by lap aro scopic op er a tion.[11,12]

The post op er a tive prog no sis var ies de pend ing on the underlying pathology.[3] For pa tients with be nign neo plastic mucoceles, the 5-year survival rate approaches 91-100%. The 5-year sur vival rate for pa tients with malig nant mucoceles, how ever, de creases to about 25% due to the com pli ca tion of pseudo myxoma peritonei.

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