—Case Reports—

Giant Appendiceal Mucocele: Report of a Case

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Abstract

Mucoceles of the appendix are a group of mucus-filled lesions causing obstructive dilation of the ileocecal appendix. We report a rare case of giant appendiceal mucocele. A 48-year-old woman, with no discomfort, was admitted to our hospital after a mass was detected in the right lower quadrant of the abdomen. The patient underwent right hemicolectomy on the basis of the clinical diagnosis of a possible appendiceal tumor. The final pathologic diagnosis was mucocele of the appendix.
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Key word: appendiceal mucocele

Introduction

Mucoceles of the appendix are a group of mucus-filled lesions causing obstructive dilation of the ileocecal appendix. They are infrequent and represent only 0.25% of indications for appendectomy and 8% of appendiceal tumors[1]. Mucoceles of the appendix can be asymptomatic and discovered incidentally with radiological or endoscopic tests or at laparotomy or laparoscopy performed for other reasons[2,3]; thus more than 50% of cases present with pain in right iliac fossa suggestive of acute appendicitis. Mucoceles of the appendix may be a benign or malignant and, thus, require individualized treatment[4]. Ten percent to 15% of mucoceles progress to pseudomyxoma peritonei, changing completely the outcome. We report a rare case of giant appendiceal mucocele and also review the literature about the clinical, radiologic, and diagnostic characteristics of this rare entity.

Case Report

A 48-year-old woman, with no discomfort, was admitted to our hospital after a mass was detected in the right lower quadrant of the abdomen. She denied any family history of malignancy. Her husband's medical, surgical, and family histories were likewise unremarkable. Palpable mass was exhibited in the right lower quadrant but rebound pain was not noted. A computed tomographic (CT) scan with contrast enhancement demonstrated a 13-cm diameter, low-density, well-encapsulated mass with wall calcification extending below the inferior wall of the cecum **(Fig. 1)**. The mass was medial to the cecum and extended to the right lateral aspect

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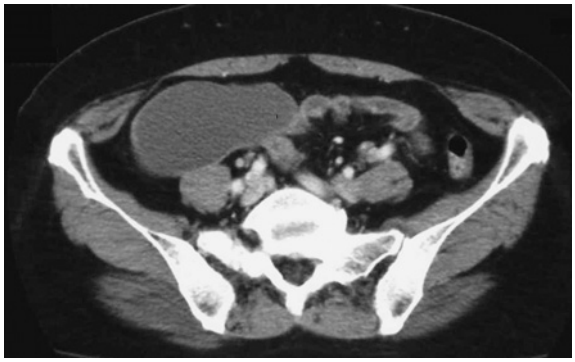

Fig. 1 An abdominopelvic CT scan demonstrated a 13-cm diameter, low-density, well-encapsulated mass with the presence of wall calcification extending below the inferior wall of the cecum.

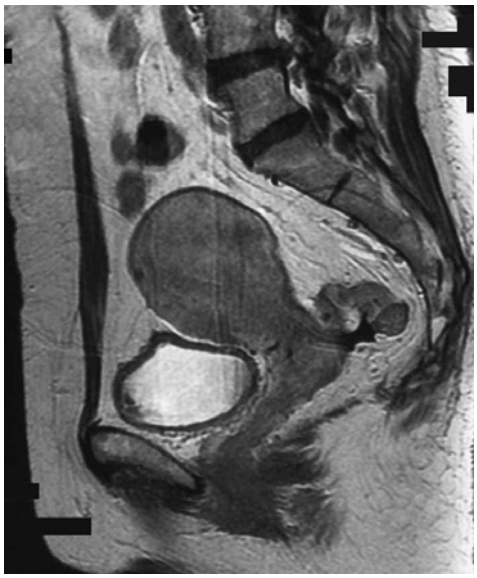

Fig. 2 T1-weighted MRI showed a well-circumscribed mass with intermediate signal intensity which extended inferiorly to the pelvis for several centimeters.

of the uterus. There was no surrounding inflammation or fluid. A T1-weighted magnetic resonance imaging (MRI) scan showed a well-circumscribed mass with intermediate signal intensity extending inferiorly to the pelvis for several centimeters **(Fig. 2)**. Results of laboratory tests, including measurements of carcinoembryonic antigen and CA 19-9, were unremarkable. Plain abdominal radiographs did not show dilated bowel loops. The liver and other solid organs were normal. In the absence of altered bowel habits, stool samples were not collected preoperatively.

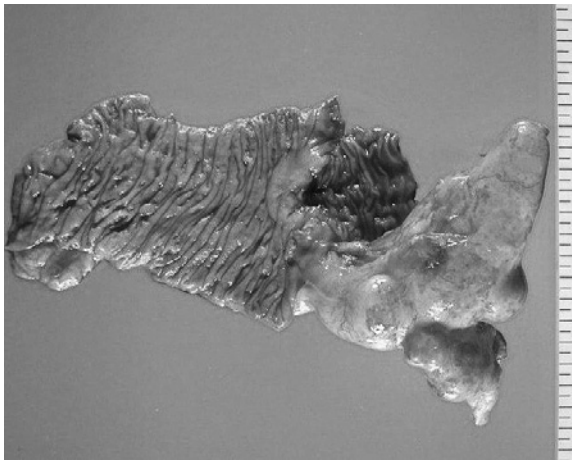

Fig. 3 At pathologic examination, the mass, which measured 13 cm in length and 10 cm in diameter, was identified as a cystic appendix.

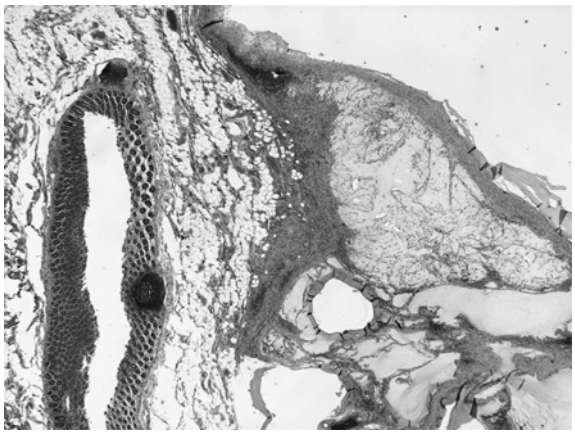

Fig. 4 On microscopic examination, patches of adenomatous mucosa were identified without evidence of invasive carcinoma.

Laparotomy revealed an appendiceal mass, but lymph nodes were not palpable along the draining vessels. Right hemicolectomy was performed on the basis of a clinical diagnosis of a possible appendiceal tumor. The reproductive organs, specifically the ovaries, were inspected intraoperatively and found to be grossly normal.

At pathologic examination, the mass, which measured 13 cm in length and 10 cm in diameter, was identified as a cystic appendix **(Fig. 3)**. There was extensive organization of intraluminal mucin against the mucosal surface. The serosal surface was smooth and pale and without implants. On microscopic examination, patches of adenomatous mucosa were identified without evidence of invasive

carcinoma (**Fig. 4**). The final pathologic diagnosis was mucocele of the appendix. The patient's postoperative course was uneventful.

Discussion

Appendiceal adenomas are divided into the diffuse, circumferential type and the extremely rare, localized, nodular type. The former tends to produce huge, thin-walled, mucus-filled cysts and is hence named mucinous cystadenoma or mucocele. However, the term "mucocele" is often used as a general descriptive term for dilatation of the appendiceal lumen by mucinous secretions. Mucoceles are found at 0.2% to 0.3% of appendectomies and autopsies, but most are smaller than the mucocele in the present patient[5]. Mucoceles are divided into 4 groups on the basis of the characteristics of their lining epithelium[5]. The first group are simple or retention mucoceles resulting from obstruction of the appendiceal outflow, usually by a fecalith, and are characterized by normal epithelium and mild luminal dilatation (≤1 cm). The second group are mucoceles with hyperplastic epithelium and mild luminal dilatation; these constitute 5% to 25% of mucoceles[5]. The third group are benign mucinous cystadenomas, the most common form, accounting for 63% to 84% of mucoceles. These exhibit mostly epithelial villous adenomatous changes with some degree of epithelial atypia and are characterized by marked distention of the lumen (≤6 cm)[5]. The mucocele in our patient belongs to this group. The fourth group are malignant mucinous cystadenocarcinomas, representing 11% to 20% of mucoceles. They are distinguished from the previous group by their glandular stromal invasion or presence of epithelial cells in the peritoneal implants or both.

Unlike the symptoms in the present case, the symptom most often associated with appendiceal mucoceles is acute or chronic abdominal pain in the right lower quadrant, which occurs in two-thirds of patients[5]. Occasionally, patients present with intermittent, colicky pain caused by intussusception of the mucocele into the cecum[6,7], gastrointestinal bleeding due to intussusception or sigmoid invasion[7,8], ureteral obstruction[9], small-bowel obstruction due to volvulus[10], or acute abdomen due to rupture and infection[9].

Mucoceles are rarely diagnosed before operation because symptoms are either absent or nonspecific. The lesion might be detected be via radiologic, sonographic, or endoscopic means. On CT, mucoceles usually appear as masses with near-water density in the right lower quadrant, with or without calcification or septation. These masses may be seen to arise from, or indent, the cecum. Attenuation values may range from near-water density to soft-tissue density[7,11]. Takahashi et al. have described MRI findings of a case of mucocele of the tip of the appendix[12]. The mucocele showed intermediate signal intensity on T1-weighted MRI because of its high protein content. Zissin et al. have reported similar findings, including absence of enhancement with gadolinium[13].

In the present case the presumptive diagnosis, before histologic examination, was an appendiceal or cecal tumor, in view of the mass lesion. At the time of operation, recognition and resection of appendiceal mucoceles are important because some are cystadenocarcinomas, which can rupture and lead to pseudomyxoma peritonei. Because of the possibility of cancer, right hemicolectomy was indicated in our case. At operation, a search should be made for coexisting tumors of the ovary and gastrointestinal tract. If exploration reveals a ruptured appendiceal mucocele, the primary resection should be accompanied by removal of all gross implants. Postoperatively, patients with simple or benign neoplastic mucoceles have an excellent prognosis, with 5-year survival rates of 91% to 100%. Fortunately, the histologic examination showed the present mucocele to be benign. Malignant mucoceles, however, have a 5-year survival rate of only 25% because of the complications of pseudomyxoma peritonei[5].

The present case shows us that, although the diagnosis of mucocele of the appendix is frequently incidental, a thorough physical examination, CT scan, and MRI may suggest the diagnosis and help in the choice of operation. Therefore, preoperative recognition with a carefully planned resection to

remove the mass is required[14,15].

Conflict of Interest: The authors declare no conflict of interest.

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