Case Report

Rare Cases of Low-Grade Appendiceal Mucinous Neoplasm: Two Case Reports and a Literature Review

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Keywords

Appendiceal mucocoele · Low-grade appendiceal mucinous neoplasm · Surgical resection

Abstract

Appendiceal mucocoele (AM) is rare disease found in <1% of all appendectomy specimens. AM is often misdiagnosed as appendicitis because the most frequent symptom is right lower quadrant abdominal pain. AM should be considered in the differential diagnosis of abdominal pain. Although there are pathological classifications, surgical resection is accepted as the treatment to prevent the development of peritoneal pseudomyxoma (PP); however, the optimal surgical technique that must be used is unclear. We present two cases suspected of being AM prior to surgery that were pathologically diagnosed as low-grade appendiceal mucinous neoplasms. Each case progressed without developing PP. The surgical procedures we chose are considered appropriate for each case.

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Introduction

Appendiceal mucocoele (AM) is a rare condition, accounting for 0.3–0.7% of appendiceal pathology, comprising 8% of appendiceal tumors, and is found in <1% of all appendectomy

specimens [1–3]. It is often misdiagnosed as appendicitis, pelvic mass, or retroperitoneal tumor in many cases because the most frequent symptom is right lower quadrant abdominal pain [3, 4]. AM can be diagnosed at any age and should be considered in the differential diagnosis of abdominal pain. Although there are pathological classifications, surgical resection is the accepted treatment modality for each histological type to prevent the development of peritoneal pseudomyxoma (PP) that is associated with a high mortality rate [1–4]. However, pre-operative diagnosis of AM is difficult because of the non-specific clinical manifestations of the disease. We present two cases that were thought to be AM prior to surgery but were diagnosed pathologically as low-grade appendiceal mucinous neoplasm (LAMN). Each case progressed without developing PP.

Case Reports

Case 1

An 82-year-old man was admitted to our hospital with complaints of weight loss of 10 kg over one year. Serum carcinoembryonic antigen (CEA) level was elevated above the normal range. Computed tomography (CT) revealed a cystic mass with enhancing wall nodules (40 × 48 mm) and a dilated appendix (22 mm) in the right iliac fossa (Fig. 1a, b), suspected to represent AM. Ileocecal resection and lymph node dissection were performed (Fig. 1c). On hematoxylin and eosin (HE) staining, the tumor was covered with mucin-producing cells, showing papillary growth and partially stratified nuclei, with no disturbance in polarity (Fig. 1d). The final pathologic diagnosis was LAMN with no metastasis to lymph nodes, according to the 2010 World Health Organization (WHO) classification. Eight years have passed since the operation, and tumor recurrence or PP has not been detected on CT. Serum CEA level has remained within the normal range after operation.

Case 2

A 73-year-old woman was referred to our hospital for an appendiceal tumor discovered incidentally on CT. She had no complaints; serum CEA level was within the normal range, and no abnormalities were observed on colonoscopy. CT showed a dilated appendix with enhancing wall nodules (45 × 45 mm) (Fig. 2a), suspected to be AM. Intraoperatively, the appendix proximal to the cecum was normal (Fig. 2b). Only appendectomy with laparoscopic assisted was performed (Fig. 2c).

On HE staining, the tumor was covered with mucinous producing cells, showing papillary growth and partially stratified nuclei (Fig. 2d). The final pathologic diagnosis was LAMN according to the 2010 WHO classification. Three years have passed since the operation, and tumor recurrence or PP has not been detected on CT.

Discussion

AM was first described in 1842 by Rokitsansky and is an unspecific term used to define cystic dilation of the appendix caused by accumulation of mucus secretion. AM is a rare disease found in < 1% of all appendectomy specimens and misdiagnosed as acute appendicitis or pelvic mass or retroperitoneal tumors in many cases, because the most frequent symptom is acute or chronic right lower quadrant abdominal pain [1–4]. It can be diagnosed at any age and should be considered in the differential diagnosis of acute abdominal pain at any age.

Diagnosis of AM is difficult because of non-specific clinical manifestations; however, imaging modalities such as ultrasound or CT help to make the diagnosis. In particular, CT is the ideal diagnostic modality for AM [5–7]. Cystadenoma or adenocarcinoma may be present; the tumors are usually >60 mm. Cystic dilatation of the appendix and luminal diameter >15 mm are important imaging indicators of a neoplasm, whereas <20 mm are rarely malignant [2, 8]. Wang et al. reported that irregular wall and soft-tissue thickening were features most likely to be associated with malignancy, and mural calcification of the wall may also be regarded as diagnostic clues for adenocarcinoma [7, 8]. In addition to these findings, an important advantage of CT is determination of the anatomic relationship between the elongated cystic mass and the cecum [8].

The treatment depends on the size of the appendix and on the histological type of the original lesion. Surgical resection is the treatment modality of choice, and surgical approaches either by laparoscopic or open surgery and extension by only appendectomy or right hemicolectomy are not defined clearly [3, 7]. However, it is very important to select an appropriate surgical method to prevent complications. Among complications, the worst is PP, characterized by peritoneal dissemination caused by rupture of the mucocoele. The choice of right hemicolectomy should be considered for luminal diameter of the appendix being >20 mm, extension beyond the appendix, and presence of lymph node metastasis or cystadenocarcinoma [1, 8].

Various classification systems have been proposed; we diagnosed the lesions reported here as LAMN according to the 2010 WHO classification [9]. The classification has the following 3 categories of mucinous neoplasms: mucinous adenoma, LAMN, and appendiceal adenocarcinoma. The prognosis differs and they each require different surveillance and treatment according to the histological type.

In our cases, the appendices were dilated slightly >20 mm; however, the masses were <60 mm in diameter, and the walls were regular without calcification. The tumors were unlikely to be appendiceal adenocarcinomas; therefore, ileocecal resection instead of right hemicolectomy was performed in case 1 and appendectomy was performed in case 2. As no recurrence of tumor or PP has been noted, both operations were appropriate.

There is no specific opinion regarding the ideal surgical procedure for AM; however, the surgical procedures we chose may be an effective option for LAMN.

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Statement of Ethics

Written informed consent was obtained from the patients.

Disclosure Statement

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Author Contributions

SY made substantial contribution towards the concept and design of the study and data acquisition and interpretation. HY, HT, SK, YT, and KS were involved in drafting the manuscript and critical revision for important intellectual content. SY approved the final version of the manuscript that was submitted for publication. All authors read and approved the final manuscript.

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Fig. 1. (a) A cystic lesion with enhancing wall nodules and adjacent cecum detected on CT. (b) Dilated appendix with regular wall detected on CT. (c) Resected ileocecum with the dilated appendix adjacent to the cecum. (d) The tumor was covered with mucinous producing cells, showing papillary growth and partially stratified nuclei; no disturbance in polarity was observed.

Fig. 2. (a) Appendix with enhancing wall nodules detected on CT. (b) Appendix proximal to the cecum was normal. (c) Resected appendix. (d) The tumor was covered by mucinous producing cells, showing papillary growth and partially stratified nuclei; no disturbance in polarity was observed.