Accidental discovery of appendiceal carcinoma in gynecological surgery: A case report

Wang L et al. Mucinous adenocarcinoma of the appendix

Lin Wang, Yan Dong, Ya-Hui Chen, Ya-Nan Wang, Lin Sun
Abstract

BACKGROUND
Malignant tumors of the appendix are extremely rare, constituting about 1% of all gastrointestinal tumors. Generally, pathology identifies these tumors during or after appendectomy because they are difficult to detect at the preoperative stage. This case report aims to introduce the definitive diagnosis and treatment of mucous adenocarcinoma of the appendix (MAA).

CASE SUMMARY
A 49-year-old female patient came to our hospital with right lower abdominal pain, nausea, and vomiting for three days. There was no change in the menstrual cycle. Gynecological ultrasound showed a cystic, solid mass in the right adnexa. Abdominal enhanced CT showed a thick appendix. Cancer was found in the exploration of the appendix during gynecological surgery. The right colon was removed. After surgery, the patient received chemotherapy and is recovering well.

CONCLUSION
Appendiceal carcinoma is frequently found during or after the operation. Preoperative examination and early evaluation of clinical manifestations are extremely important.

Key Words: Abdominal pain; Pelvic mass; Appendix carcinoma; Mucous adenocarcinoma; Case report

Core Tip: Mucinous adenocarcinoma of the appendix (MAA) has a low incidence rate and is relatively rare. The increase of tumor markers in patients has certain guiding significance. Imaging examination can suggest that the appendix is thickened, and its diagnosis depends on histopathology.
INTRODUCTION
The appendix epithelial malignant tumors can be divided into three categories: mucous adenocarcinoma of the appendix (MAA)\(^1\), intestinal type of adenocarcinoma, and signet ring cell carcinoma. Among these, MAA is the most prevalent histological form. Its occurrence might be linked to chronic inflammatory infiltration of the appendix. Here, we report a case of mucinous appendiceal adenocarcinoma, accidentally found during an operation for the right lower abdominal pain and a pelvic mass. We also reviewed available relevant literature.

CASE PRESENTATION

Chief complaints
A 49-year-old female patient came to our hospital with right lower abdominal pain, nausea, and vomiting.

History of present illness
Patient’s symptoms started three days ago with right lower abdominal pain, nausea, and vomiting.

History of past illness
There was no change in the menstrual cycle. The patient had a free previous medical history.

Personal and family history
The patient is in good health and has no family genetic diseases.

Physical examination
Gynecologic examination suggested normal vulvar development, a smooth vagina, little vaginal discharge, a soft cervix, a uterus of average size, no tenderness. There was no abnormality in the left accessory, and a cystic solid tumor with a size of about 6 cm could be touched in the right accessory.

**Laboratory examinations**

HE4, CEA, AFP were normal on May 14, 2021. The patient’s CA125 was 392.9 U/mL and CA199 was 88.27 U/mL on May 14, 2021.

**Imaging examinations**

Gynecological ultrasound on May 15, 2021, showed a cystic, solid mass in the right adnexa area of approximately 6.2 cm × 5.6 cm × 5.8 cm in size, unclear right ovary. On May 18, 2021, a complete abdominal CT showed hypocrine shadow in the right adnexa area with a visible compartment inside and appendiceal thickening with a maximal thickness of around 12 mm (Figure 1). The mass in the pelvic cavity remained unidentified.

**FINAL DIAGNOSIS**

The patient had an exploratory laparotomy on May 21, 2021, during which the right accessory and cystic mass were removed, thickening of the appendix (~6 cm in length and ~1 cm in diameter) was noted, with hard texture and edema, attached to the posterior wall of the ascending colon. Appendiceal malignancy could not be excluded, which was revealed to be true by rapid intraoperative pathology. The final diagnosis of the presented case is MAA.

**TREATMENT**

Gastrointestinal surgery facilitated the right hemicolecotomy and peripheral lymph node dissection. The residual intestine, stomach, liver, greater omentum, and
peritoneum surface were explored at the end of the surgery, and no evidence of abnormality was found.

OUTCOME AND FOLLOW-UP
Postoperative pathology confirmed mucous adenocarcinoma of the appendix with partial signet-ring cell carcinoma. Immunohistochemistry was performed using markers: CDX-2 (+), CK7 (-), CK20 (+), CA125 (-), CD56 (-), Syn (-), Pax-8 (-), WT-1 (-), SATB2 (+) (Figure 2). The patient began to receive chemotherapy 45 days after the operation. By now, six courses of XELOX chemotherapy regimen (Oxaliplatin + Capecitabine) have been completed, and there was no evidence of recurrence. MAA with signet-ring cell features is considered more invasive and has a worse prognosis. The patient requires regular follow-up every four months for three years, every six months for the next two years, and then every year for the next 15 years, following initial therapy[3].

DISCUSSION
MAA is a rare disease characterized by elevated CA199 and CEA levels. C.PABLO et al. showed that tumor markers CEA and CA199 have high clinical value in diagnosing MAA[3-4]. Moreover, the increasing level of CA125 while maintaining normal HE4 helps discriminate between benign and malignant ovarian tumors. The patient, in this case, showed elevated CA199 and CA125 levels, which can help diagnose appendiceal lesions[5]. Michelle Moh et al. used immunohistochemistry to identify SATB1 in ovarian mucinous tumors. The presence of SATB2 (+) and CDX-2 (+) highly suggests that the tumor originates from the colon or appendix[4]. Imaging is a useful diagnostic tool for MAA. Ultrasound observations revealed a cystic mass in the appendix, heterogeneous echogenicity, hypocoistic or tubular lesions in the appendix, and irregular thickening. CT scans rule out appendiceal inflammation and abscess, and all above findings help diagnose mucinous cystadenoma[5-6]. MAA is difficult to diagnose due to the non-specific nature of early symptoms, including lower abdominal
pain, weight loss, nausea, vomiting, a palpable mass, and acute appendicitis, frequently misdiagnosed as a gynecological condition such as right adnexal mass\textsuperscript{[7]}. The bladder may also be affected, having symptoms of bladder irritation or the formation of hematuria, leading to a misdiagnosis of urinary tract infection or bladder cancer\textsuperscript{[8-10]}. In most cases, appendiceal malignancy is detected accidentally by abdominal CT or surgery for appendicitis due to other reasons. Appendiceal cancer is difficult to be found even by preoperative colonoscopy\textsuperscript{[11]}. In the case of submucosal lesions of the cecum near the mouth of the appendix, mucus is flowing out of the mouth of the appendix; thus, the treating physician should be highly vigilant against appendiceal lesions. Mucous adenocarcinoma of the appendix is associated with a high risk of peritoneal seeding along with hematogenous and lymph node metastasis. As a surgical treatment for mucous adenocarcinoma, simultaneous surgical removal of the appendix and right hemicolecctiony with peripheral lymph node dissection is preferred\textsuperscript{[12]}. Laparotomy is superior to laparoscopic surgery as it facilitates exploring other organs involvement. It is better to protect from the incision, thereby avoiding mass rupture, leading to intra-peritoneal dissemination and affecting prognosis.

**CONCLUSION**

MAA is remarkably rare, difficult to diagnose and distinguish from other tumors. Preoperative laboratory, imaging examinations, a well-planned diagnostic and treatment strategy are essential. In mucous neoplasms, we do believe that a right hemicolecctiony should definitely be performed if required for tumor clearance because a complete cytoreduction of mucous tumors of the appendix is associated with improved survival\textsuperscript{[12, 13]}. The patient described here presented with common clinical symptoms of MAA. For patients with the appearance of an abnormal appendix in the preoperative examination, it is necessary to explore the appendix at the same time to avoid missing diagnosis and misdiagnosis.
**ORIGINALITY REPORT**

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