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Case Report

Low-grade appendiceal mucinous adenocarcinoma with peritoneal and ovarian spread: a case report and literature review

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ABSTRACT

Appendiceal mucinous neoplasms are rare and often present with nonspecific symptoms, leading to diagnostic challenges. This case report describes an atypical presentation of a low-grade mucinous appendiceal adenocarcinoma initially misdiagnosed as an ovarian mass, highlighting the importance of a multidisciplinary diagnostic approach. This case report involves a 57-year-old postmenopausal woman presented with persistent gastroesophageal reflux disease (GERD) and was found to have a large abdominopelvic cystic mass on imaging, suggestive of ovarian origin. Further evaluation, including computed tomography (CT), magnetic resonance imaging (MRI), and histopathological analysis after exploratory laparotomy, revealed a metastatic low-grade mucinous appendiceal adenocarcinoma with peritoneal dissemination (pseudomyxoma peritonei). The patient was referred for cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC). This case illustrates the diagnostic complexity of appendiceal mucinous neoplasms, particularly when mimicking gynaecological pathology**.** A high index of suspicion, comprehensive imaging, and histopathological confirmation are crucial for accurate diagnosis. CRS and HIPEC remain the standard of care for advanced disease, emphasizing the need for early multidisciplinary involvement in management.

Keywords: Appendiceal mucinous adenocarcinoma, Pseudomyxoma peritonei, Ovarian metastasis, Cytoreductive surgery, HIPEC, Atypical presentation

INTRODUCTION

Appendiceal mucinous neoplasms are rare clinical entities that often present diagnostic challenges due to their variable and nonspecific clinical manifestations. These tumours account for less than 0.5% of gastrointestinal malignancies and are frequently discovered incidentally during imaging or surgery for unrelated conditions.[1] The spectrum of appendiceal mucinous lesions ranges from benign mucoceles to malignant adenocarcinomas, with low-grade appendiceal mucinous neoplasms (LAMNs) occupying an intermediate position.[2,3]

This case report describes a 57-year-old postmenopausal woman who initially presented with persistent gastroesophageal reflux disease (GERD) and was subsequently found to have a large abdominopelvic cystic mass of suspected ovarian origin. Imaging and histopathological evaluation ultimately revealed a low-grade mucinous appendiceal adenocarcinoma with ovarian and peritoneal metastases, consistent with pseudomyxoma peritonei (PMP).

This case highlights the importance of a multidisciplinary approach in diagnosing and managing atypical presentations of appendiceal malignancies, particularly when mimicking gynaecological pathology. Additionally, we review the current literature on appendiceal mucoceles, their diagnostic pitfalls, and the evolving role of cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC) in treatment.[4,5]

CASE REPORT

Patient information

A 57-year-old woman, gravida 5 para 5, postmenopausal for five years, with a past surgical history of inguinal hernia repair over twenty years ago, initially presented for persistent GERD and nausea. There was no history of weight loss, hematemesis, or dysphagia.

Clinical findings

On admission, the patient was hemodynamically and respiratorily stable. Abdominal examination revealed a renitent mass reaching the umbilicus. Her obesity masked any obvious abdominal distension. Gynaecological examination was unremarkable, with no abnormal discharge or cervical lesions.

Timeline of current episode

Week 0: Presentation with persistent GERD, week 1: endoscopy revealed extrinsic compression of the gastric wall, week 2: CT scan showed a large multiloculated cystic abdominopelvic mass, week 3: referral to gynaecology, week 4: MRI confirmed a suspicious ovarian mass (ORADS 4), week 5: exploratory laparotomy and adnexectomy, and week 6: histopathological diagnosis of low-grade appendiceal mucinous adenocarcinoma with ovarian and peritoneal metastases.

Diagnostic assessment

Imaging (CT and MRI) revealed a large, multiloculated cystic mass in the left adnexal region with abundant ascites, initially suggestive of a borderline ovarian tumour. Tumour markers (CA-125, CEA, AFP, CA 19-9) were all within normal ranges.

Intraoperative findings and histopathology revealed low-grade mucinous appendiceal adenocarcinoma with peritoneal and ovarian metastases, confirming pseudomyxoma peritonei.

Diagnosis

Final diagnosis included low-grade mucinous appendiceal adenocarcinoma with peritoneal and ovarian spread, consistent with pseudomyxoma peritonei.

Therapeutic interventions

A midline exploratory laparotomy was performed. Surgical procedures included a left adnexectomy (Figures 1 and 2), appendectomy, peritoneal biopsies (parieto-colic gutters, epiploic region), and contralateral ovarian sampling. The procedure lasted 40 minutes with no hemodynamic instability, postoperative complications, or need for blood transfusion.

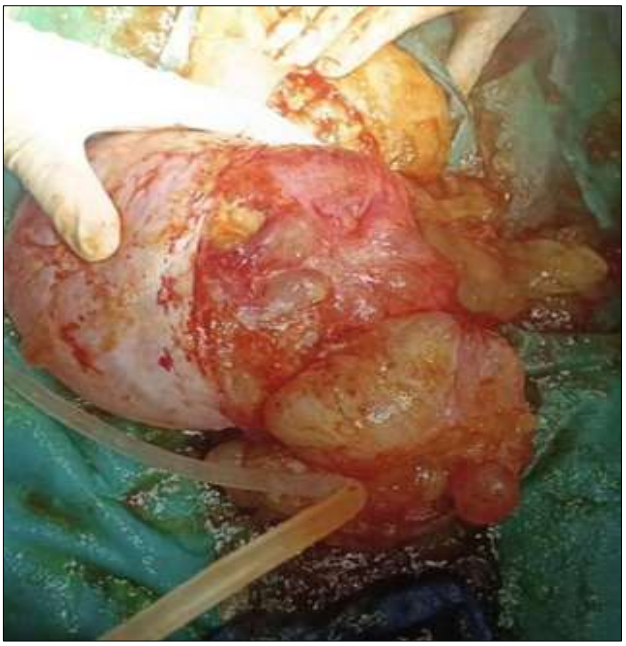

Figure 1: Ovarian tumour during the procedure.

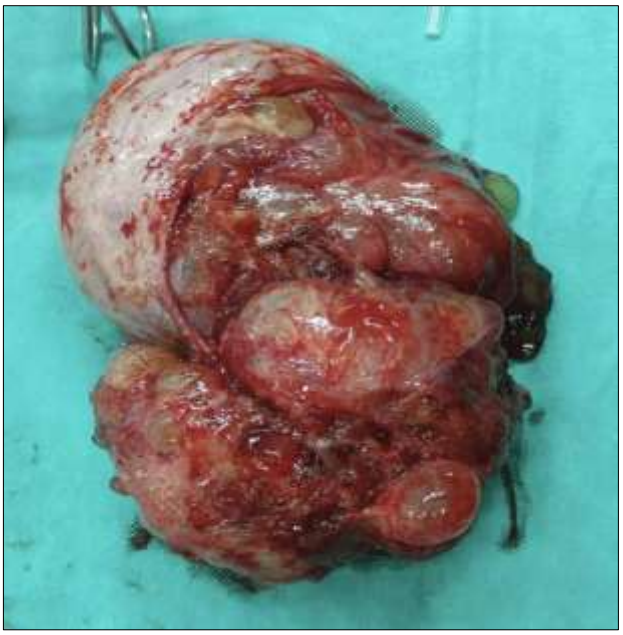

Figure 2: Surgical specimen.

Follow-up and outcomes of interventions

Postoperative recovery was uneventful. Following multidisciplinary discussion, the patient was referred to a specialized oncology center for cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC).

Patient perspective

The patient expressed relief at receiving a definitive diagnosis after an extended period of uncertainty. She was

appreciative of the thorough multidisciplinary evaluation and is optimistic regarding her treatment plan.

DISCUSSION

Appendiceal mucinous neoplasms are rare clinical entities, representing less than 0.5% of gastrointestinal malignancies. Among these, low-grade appendiceal mucinous adenocarcinomas (LAMNs) are even more uncommon, typically presenting in the fifth to sixth decades of life, with a slight female predominance.[1] These tumors may remain asymptomatic or manifest through vague abdominal discomfort. In our case, the patient presented with atypical upper gastrointestinal symptoms---specifically, gastroesophageal reflux linked to extrinsic gastric compression, a highly unusual presentation. Mucinous neoplasms may eventually cause pseudomyxoma peritonei (PMP), a potentially life-threatening condition characterized by progressive intraperitoneal accumulation of mucinous material.[2,3]

The diagnosis of appendiceal mucinous tumours is challenging due to their nonspecific presentation and radiological mimicry of ovarian masses, especially in women. On ultrasound, mucoceles may appear as cystic masses with internal echogenic layering ("onion-skin sign"). CT typically reveals a well-circumscribed, low-attenuation, tubular mass adjacent to the cecum, often with mural calcifications.[4] MRI provides superior soft-tissue contrast and may further characterize the lesion.[5]

In our patient, the mass was misinterpreted as an ovarian tumor on imaging, with an ORADS 4 classification. This underscores the diagnostic difficulty and the necessity of histopathological confirmation following surgical exploration.

The standard management of LAMNs with peritoneal dissemination is a combination of CRS and HIPEC. CRS involves extensive resection of all visible tumour deposits, including omentectomy and selective peritonectomies, while HIPEC aims to eradicate microscopic residual disease through the direct administration of heated chemotherapeutic agents into the peritoneal cavity.[6,7] In this case, the initial surgery was limited to diagnostic and staging purposes. The patient was subsequently referred to a specialized center for CRS and HIPEC, which has been shown to significantly improve survival outcomes when complete cytoreduction is achieved.[8]

This case illustrates multiple atypical aspects like an unusual initial symptom (GERD), significant obesity that masked physical signs, and a primary tumour that mimicked an adnexal mass. Such presentations highlight the importance of maintaining a broad differential diagnosis, particularly when imaging findings are equivocal. Early multidisciplinary evaluation including gynaecology, oncology, radiology, and pathology is essential to avoid misdiagnosis and ensure optimal management. Continued reporting of rare and misleading cases is crucial for improving diagnostic pathways and clinical outcomes.[9,10]

CONCLUSION

This case underscores the diagnostic complexity of appendiceal mucinous neoplasms, particularly when presenting with atypical symptoms and mimicking ovarian pathology. The patient's initial presentation with GERD and subsequent discovery of a large cystic mass highlights the need for a high index of suspicion in cases of unexplained abdominal findings, especially in the presence of ascites. The definitive diagnosis of low-grade mucinous appendiceal adenocarcinoma with peritoneal dissemination necessitated referral for specialized management, including CRS and HIPEC, which remain the cornerstone of treatment for pseudomyxoma peritonei.

This report emphasizes the importance of comprehensive imaging, histopathological correlation, and multi-disciplinary collaboration in optimizing outcomes for rare and diagnostically challenging cases. Further research is needed to refine diagnostic algorithms and therapeutic strategies for appendiceal mucinous neoplasms to improve early detection and long-term survival.

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