Clinical Case

Low-grade Appendiceal Mucinous Neoplasm and Pseudomyxoma Peritonei: A Case of Appendiceal Mucocele Autoamputation

Neoplasia Mucinosa Apendicular de Baixo Grau e Pseudomixoma Peritoneal: Um Caso de Autoamputação de Mucocelo Apendicular

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ABSTRACT

Low-grade appendiceal mucinous neoplasms (LAMNs) are rare mucin-producing lesions of the appendix, accounting for less than 1% of all gastrointestinal neoplasms. These lesions typically present as a mucocele — a dilated, mucin-filled appendix — either causing an acute appendicitis or detected incidentally on imaging. Perforation of the mucocele can lead to peritoneal mucin deposits, clinically referred to as pseudomyxoma peritonei (PMP). We report a case of a 31-year-old woman, with no significant personal medical history, presenting with lower abdominal pain of several weeks' duration, whose imaging was suspicious for an appendiceal mucocele. The patient underwent a laparoscopic exploration, and an appendiceal mucocele completely amputated from the cecum was identified, along with mucin deposits on the appendiceal serosa, and free mucin within the peritoneal cavity, with a Peritoneal

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Cancer Index (PCI) >3. Cytological analysis of the mucin was negative for malignant cells, with histological findings compatible with a LAMN. The patient underwent cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC). Appendiceal auto-amputation represents an exceptionally rare clinical entity, with the literature limited to a small number of reported cases and is even more uncommon when associated with concomitant PMP and LAMN. In the management of these lesions, it is essential to thoroughly inspect the entire peritoneal cavity to exclude or confirm the presence of PMP, as this influences future therapeutic decisions and the overall prognosis.

Keywords: Appendiceal Neoplasms; Mucocele; Pseudomyxoma Peritonei

RESUMO

As neoplasias mucinosas do apêndice com displasia de baixo grau (LAMN) são lesões produtoras de mucina e representam menos de 1% de todas as neoplasias gastrointestinais. Estas lesões originam frequentemente o denominado mucocelo apendicular, apêndice dilatado e preenchido por mucina. Pode apresentar-se na forma de apendicite aguda ou ser detetado incidentalmente em exames de imagem. A perfuração do mucocelo pode levar à deposição peritoneal de mucina, clinicamente designado de pseudomixoma peritoneal (PMP). Relatamos o caso de uma mulher de 31 anos, sem antecedentes pessoais relevantes, com dor abdominal nos quadrantes inferiores com semanas de evolução, e exames de imagem sugestivos de mucocelo apendicular. Foi realizada uma laparoscopia exploradora, identificando-se um mucocelo apendicular completamente amputado do cego, associado a depósitos de mucina na serosa apendicular e na cavidade peritoneal, com um Índice de Carcinomatose Peritoneal (PCI) >3. A análise citológica da mucina foi negativa para células malignas e os achados histológicos compatíveis com LAMN. A doente foi submetida a cirurgia citorredutora e quimioterapia intraperitoneal hipertérmica. Os casos de autoamputação apendicular descritos na literatura são raros, e ainda menos comuns quando associados a LAMN e PMP. Na abordagem destas neoplasias, é essencial inspecionar adequadamente toda a cavidade peritoneal de forma a excluir ou confirmar a presença de PMP, que influencia a decisão terapêutica futura e o prognóstico da doença.

Palavras-chave: Neoplasias do Apêndice; Mucocelo; Pseudomixoma Peritoneal

INTRODUCTION

The 5th edition of the World Health Organization (WHO) Classification of Digestive System Tumors' (2019) categorizes non-neuroendocrine epithelial tumors of the appendix into serrated lesions and polyps (hyperplastic polyps, serrated lesions with and without dysplasia), appendiceal mucinous neoplasms (low-grade appendiceal mucinous neoplasms [LAMNs] and high-grade appendiceal mucinous neoplasms [HAMNs]), mucinous adenocarcinoma and non-mucinous adenocarcinoma.[1]

Low-grade appendiceal mucinous neoplasms (LAMNs) are rare mucin-producing malignant lesions, accounting for less than 1% of all gastrointestinal tumors.[2] Histologically characterized by low-grade cytologic atypia and features such as loss of the muscularis mucosae and submucosal fibrosis, without infiltrative growth.[2-4] The staging of LAMNs has been more recently described by the AJCC (American Joint Committee on Cancer), with a dedicated TNM classification.[5,6]

These lesions may present clinically as acute appendicitis due to appendiceal obstruction, or they may be incidentally detected on imaging studies performed for the evaluation of abdominal pain. They typically present, either intraoperatively or on imaging, as a mucocele (a dilated, mucin-filled appendix). Perforation of the mucocele may occur spontaneously or intraoperatively, leading to the spread of mucin and epithelial cells throughout the peritoneal cavity, resulting in pseudomyxoma peritonei, which can be life-threatening. Lymphatic and hematogenous spread is essentially restricted to mucinous adenocarcinoma; recent multi-institutional data confirm the absence of nodal involvement in both LAMN and HAMN, with lymph node metastases identified almost exclusively in appendiceal adenocarcinoma cases.[2-10]

Autoamputation of the appendix, which refers to the complete separation of any part of the appendix (tip, body or root) without surgical intervention, is a rare finding in clinical practice and is thought to result from full-thickness

necrosis of a portion of the appendix due to ischemia. It is typically associated with diffuse peritonitis and even intestinal fistulas.[7]

The Peritoneal Surface Oncology Group International (PSOGI) established a consensus (2020) on the management of appendiceal neoplasms and pseudomyxoma peritonei (PMP). According to these recommendations, low-grade appendiceal mucinous neoplasms (LAMNs) without evidence of perforation should be managed with appendectomy followed by clinical surveillance.[8,9] which have been more recently supported by NCCN guidelines for Appendiceal Neoplasms and Cancers.[10] In contrast, in cases of mucocele perforation, patients are almost invariably candidates for cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC), particularly when cellular mucin or established pseudomyxoma peritonei is present; the Peritoneal Cancer Index (PCI) is used for prognostication and to assess feasibility of complete cytoreduction, rather than as a rigid threshold for treatment indication.[9,10] Current evidence indicates that LAMNs and HAMNs rarely metastasize via the lymphatic system; the recent German S2k-guideline (2024) explicitly states that oncological resections such as right hemicolectomy and lymphadenectomy are not justified in these specimens.[10] According to the more recent NCCN guidelines for Appendiceal Neoplasms and Cancers, right hemicolectomy should therefore be reserved for selected cases, such as suspicion of mucinous adenocarcinoma or when R0 resection cannot be achieved by appendectomy alone.[10]

It is important to be aware of the complications associated with these neoplasms when approaching them, in order to avoid disease dissemination or unnecessary procedures.

CASE REPORT

A 31-year-old female patient, with no significant past medical history, was referred for a Colorectal Surgery consultation following the incidental finding of an appendiceal mucocele during imaging performed as part of an abdominal pain work-up in Gynecology. She reported lower abdominal pain of several weeks' duration and a perceived reduction in bowel movement regularity. She denied fever, weight loss, or fatigue. Physical examination revealed tenderness on palpation of the lower abdominal quadrants.

Laboratory findings were unremarkable (WBC 6800/µL; CRP 8.3 mg/L). Computed tomography (CT) revealed a tubular structure consistent with an appendiceal mucocele (Fig. 1). Tumour markers (CA 19.9 – 26 U/mL; CEA – 1.1 ng/mL) and a staging chest CT were without abnormalities.

After discussion within the Colorectal Surgery team and exclusion of surgical contraindications, the patient underwent a laparoscopic exploration. After induction of general anesthesia, a pneumoperitoneum was established using a conventional three-port technique, and the abdominal cavity was explored. Intraoperatively, an appendiceal mucocele completely amputated from the cecum was identified, measuring 6 cm in length and 2 cm in diameter (Fig. 2), along

Figure 1: Preoperative computed tomography (CT) revealing a dilated and filled appendix (2 cm in diameter) compatible with an appendicular mucocele.

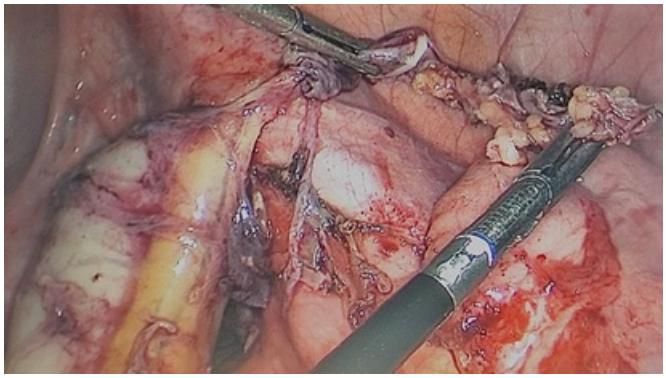

Figure 2: Laparoscopic exploration revealed an appendix amputated from the cecum, with a thin layer of mesoappendix preserved, along with mucin deposits in the peritoneal cavity, predominantly in the pelvis.

with mucin deposits on the appendiceal serosa (Fig. 3) and free mucin within the peritoneal cavity, predominantly in the pelvis. The autoamputated appendix was removed, and peritoneal fluid was collected, for histologic and cytological evaluation, respectively.

Figure 3: Surgical specimen: the appendicular mucocele, with 6 cm in length and 2 cm in diameter, and mucin on it's serosa. Pathological diagnosis: low-grade appendiceal mucinous neoplasm (LAMN). Cytological analyses: acellular mucin.

The postoperative histopathology confirmed a low-grade appendiceal mucinous neoplasm (LAMN) with mucin involving the serosal surface. Cytological analysis of the peritoneal fluid was negative for malignant cells. Based on the AJCC classification, the lesion was staged as an LAMN T4a M1a. The case was subsequently reviewed at a multidisciplinary tumor board. The patient was referred to the Portuguese Institute of Oncology for further assessment and underwent cytoreductive surgery (CRS) – with bilateral oophorectomy, omentectomy and selective peritonectomies - combined with hyperthermic intraperitoneal chemotherapy (HIPEC).

DISCUSSION

Around 50% of low-grade appendiceal mucinous neoplasms clinically present as acute appendicitis, while the remaining cases are asymptomatic and typically identified incidentally.[2]

Appendiceal mucinous neoplasms are recognized as one of the principal causes of appendiceal perforation and constitute the most frequent origin of pseudomyxoma peritonei. Conversely, around 20% of mucinous appendiceal neoplasms progress to PMP. Interestingly, in our case, the appendix was amputated from the cecum and only partially tethered to the mesoappendix with preserved blood supply. According to the scarce studies available on this topic, the presence of appendiceal amputation is confirmed intraoperatively, as it is extremely difficult to diagnose through non-invasive examinations.[7] Following an extended review, no cases of appendiceal autoamputation associated with low-grade appendiceal mucinous neoplasm (LAMN) and appendiceal mucocele were identified in the available published literature. Only a few authors have reported cases of appendiceal autoamputation. Wang *et al*[7] described this entity in the context of an appendectomy for acute appendicitis, in the absence of an appendiceal mucocele. In contrast, in an earlier report, Markey CM *et al*[12] described a case of an autoamputated appendix secondary to a chronic adnexal abscess.

The presence of acellular mucin on the appendiceal serosa and the deposits within the peritoneum warrant classification as a LAMN T4a M1a according to the AJCC staging system, with peritoneal involvement graded M1a.[5,6] Histopathological examination demonstrated the microscopic features required to confirm true serosal involvement, namely a granulation tissue-like response with neovascularisation around the serosal mucin deposits, thereby excluding luminal mucin contamination as the source — a distinction that is crucial for accurate pT4a designation. While LAMNs confined to the appendix have an excellent prognosis with a low recurrence rate, patients with pseudomyxoma peritonei exhibit a markedly variable risk of recurrence: approximately 3%–7% when mucin is acellular, and 33%–77% when mucin contains neoplastic cells.[1]

In the present case, the appendiceal perforation and peritoneal mucin deposits supported referral to a specialized peritoneal malignancy center (Portuguese Institute of Oncology) due to the indication for cytoreductive surgery (CRS) and HIPEC, in line with the PSOGI/EURACAN clinical practice guidelines (Govaerts *et al*, 2021)[9] and more recently, NCCN guidelines for Appendiceal Neoplasms and Cancers.[10]

Autoamputated appendiceal mucoceles are exceptionally rare and represent a scarcely reported condition in the literature. It becomes even rarer when associated with pseudomyxoma peritonei and a low-grade appendiceal mucinous neoplasm.

TAKE HOME MESSAGES

– Appendiceal neoplasms, including low-grade appendiceal mucinous neoplasms (LAMNs), represent an important differential diagnosis in patients presenting with abdominal pain, right iliac fossa mass and bowel obstruction;

– LAMNs staging is essential, the integrity of the appendix, mucin deposits and cytology are relevant variables to have in mind as staging plays a key role in both prognostic and therapeutic planning.

– A careful and comprehensive exploration of the peritoneal cavity should be performed and the Peritoneal Cancer Index (PCI) should be systematically calculated to assess the extent of peritoneal disease. Atypical presentations may occur as above reported.

– During the surgical management of appendiceal mucocele, emphasis should be placed on undertaking the minimum essential procedure to avoid anatomy disruption which can influence future treatments.

PRÉMIOS E APRESENTAÇÕES ANTERIORES/AWARDS AND PREVIOUS PRESENTATIONS

Caso clínico apresentado no Congresso da Sociedade Portuguesa de Cirurgia em Março de 2026 – premiado com 3º lugar na categoria de melhor póster

ETHICAL CONSIDERATIONS

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CL: Writing the original draft. Conceptualization and supervision.
AGM and CAM: Conceptualization and supervision.
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CL: Redação do rascunho original. Conceitualização e supervisão.
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