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RARE CASE OF LOW-GRADE MUCINOUS NEOPLASIA OF THE VERMIFORM APPENDIX: CASE STUDY

CASO RARO DE NEOPLASIA MUCINOSA DE BAIXO GRAU DO APÊNDICE VERMIFORME: ESTUDO DE CASO

CASO RARO DE NEOPLASIA MUCINOSA DE BAJO GRADO DEL APÉNDICE VERMIFORME: ESTUDIO DE CASO

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ABSTRACT: Mucinous neoplasms of the vermiform appendix constitute a rare and heterogeneous group of tumors characterized by abnormal mucin production and variable biological behavior. This is a descriptive and retrospective case study reporting a rare case of Low-Grade Mucinous Neoplasm of the Vermiform Appendix, treated at a general surgery referral center. The patient presented with a clinical course compatible with an initially nonspecific acute abdominal condition, which progressed within 24 hours to localized pain in the right iliac fossa, associated with clinical signs suggestive of surgical pathology. Clinical evaluation and physical examination, with a positive Blumberg sign, were decisive for the indication of surgical intervention. The findings reinforce the fundamental role of histopathological analysis for diagnostic confirmation, as well as the relevance of imaging studies in assessing the extent of the disease and in planning surgical treatment. The choice of individualized therapeutic approach proved essential for the patient's favorable clinical evolution, highlighting the need for careful management to prevent complications such as peritoneal dissemination.

KEYWORDS: Pseudomyxoma peritonei. Appendiceal neoplasms. Case report. Surgical treatment.

RESUMO: *As neoplasias mucinosas do apêndice vermiforme constituem um grupo raro e heterogêneo de tumores caracterizados pela produção anormal de mucina e comportamento biológico variável. Trata-se de um estudo de caso, de natureza descritiva e retrospectiva, que relata um caso raro de Neoplasia Mucinosa de Baixo Grau do Apêndice Vermiforme, atendido em um serviço de referência em cirurgia geral. O paciente apresentou evolução clínica compatível com quadro abdominal agudo inicialmente inespecífico, que progrediu em um período de 24 horas para dor localizada em fossa ilíaca direita, associada a sinais clínicos sugestivos de patologia cirúrgica. A avaliação clínica e o exame físico, com presença de sinal de Blumberg positivo, foram determinantes para a indicação de abordagem cirúrgica. Os achados reforçam o papel fundamental da análise histopatológica para a confirmação diagnóstica, bem como a relevância dos exames de imagem na avaliação da extensão da doença e no planejamento do tratamento cirúrgico. A escolha da conduta terapêutica individualizada mostrou-se essencial para a evolução clínica favorável do paciente, destacando a necessidade de manejo cuidadoso para prevenir complicações, como a disseminação peritoneal.*

PALAVRAS-CHAVE: *Pseudomixoma peritoneal. Neoplasias apendiculares. Relato de caso. Tratamento cirúrgico.*

RESUMEN: *Las neoplasias mucinosas del apéndice vermiforme constituyen un grupo raro y heterogéneo de tumores caracterizados por la producción anormal de mucina y un comportamiento biológico variable. Se trata de un estudio de caso, de naturaleza descriptiva y retrospectiva, que describe un caso raro de Neoplasia Mucinosa de Bajo Grado del Apéndice Vermiforme, atendido en un servicio de referencia en cirugía general. El paciente presentó una evolución clínica compatible con un cuadro abdominal agudo inicialmente inespecífico, que progresó en un período de 24 horas a dolor localizado en la fosa ilíaca derecha, asociado a signos clínicos sugestivos de patología quirúrgica. La evaluación clínica y el examen físico, con presencia de signo de Blumberg positivo, fueron determinantes para la indicación del abordaje quirúrgico. Los hallazgos refuerzan el papel fundamental del análisis histopatológico para la confirmación diagnóstica, así como la relevancia de los estudios de imagen en la evaluación de la extensión de la enfermedad y en la planificación del tratamiento quirúrgico. La elección de una conducta terapéutica individualizada resultó esencial para la evolución clínica favorable del paciente, destacando la necesidad de un manejo cuidadoso para prevenir complicaciones, como la diseminación peritoneal.*

PALABRAS CLAVE: *Seudomixoma peritoneal. Neoplasias apendiculares. Reporte de caso. Tratamiento quirúrgico.*

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INTRODUCTION

Mucinous neoplasms of the vermiform appendix constitute a rare and heterogeneous group of tumors characterized by abnormal mucin production and variable biological behavior (Carr & Arends, 2024; Constantin et al., 2023). Although they represent a minimal fraction of gastrointestinal tumors, their recognition has significant clinical importance, particularly due to the risk of rupture and subsequent mucinous dissemination into the peritoneal cavity, resulting in peritoneal pseudomyxoma (D'Amata et al., 2024; Matias-García et al., 2021). The current histopathological classification, based on World Health Organization (2019) guidelines, distinguishes these lesions according to the degree of cellular atypia and invasive potential (Ahadi et al., 2021), with low-grade appendiceal mucinous neoplasm (LAMN) being one of the most frequently diagnosed forms (Doll et al., 2024; Guner & Aydın, 2023).

The vermiform appendix is susceptible to various inflammatory and neoplastic processes, many of which mimic the clinical presentation of acute appendicitis (Bandyopadhyay et al., 2022). In many cases, LAMN is identified incidentally during abdominal surgeries performed for other reasons, reinforcing the need for careful histopathological evaluation of all resected appendiceal specimens (Li et al., 2022). This incidental finding has contributed to a better understanding of the morphological characteristics and clinical behavior of these lesions, enabling the standardization of diagnostic and therapeutic approaches (González Bayón et al., 2023).

Histologically, LAMN is characterized by neoplastic mucinous epithelium with mild cytological atypia, villous or serrated growth, and intraluminal mucin accumulation (Carr & Arends, 2024; Polydorides & Wen, 2022). Recent studies demonstrate that these neoplasms are frequently associated with mutations in the KRAS and GNAS genes, suggesting an important genetic role in their pathogenesis (Doll et al., 2024; Miller & Votanopoulos, 2023; Munari et al., 2021; Yanai et al., 2021). The integrity of the appendix wall is a key prognostic factor, as rupture or leakage of mucin containing neoplastic cells can result in peritoneal pseudomyxoma, a condition that is difficult to manage and associated with high morbidity (González Bayón et al., 2023).

Clinically, LAMN presents non-specific symptoms, commonly mistaken for acute appendicitis, including right lower quadrant abdominal pain, nausea, fever, and mild leukocytosis (Fatima et al., 2022). In other cases, patients may remain asymptomatic for long periods, with the diagnosis made incidentally on imaging studies or during surgical procedures. Computed tomography and magnetic resonance imaging are valuable tools, as they allow for the identification of appendicular dilation, mucin accumulation, and the absence of typical signs of acute inflammation (Mannarini et al., 2025; Yao et al., 2024).

Treatment for LAMN is primarily surgical and can range from simple appendectomy to

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more extensive procedures, depending on the extent of the disease and the integrity of the appendiceal wall (Lu et al., 2025; Ma et al., 2025). In cases confined to the appendix, the prognosis tends to be favorable, but requires close follow-up due to the possibility of recurrence or progression to peritoneal pseudomyxoma (Hissong & Yantiss, 2022). Therapeutic management should be individualized, taking into account clinical, histopathological, and surgical factors to ensure the safest and most effective approach (Mouawad et al., 2024).

In light of the above, the present study aims to report a rare case of low-grade mucinous neoplasm of the vermiform appendix, highlighting the clinical, radiological, and histopathological findings, as well as discussing the diagnostic and therapeutic challenges related to this entity. Thus, the study aims to contribute to the medical literature by reinforcing the importance of early diagnosis and appropriate surgical management in preventing complications such as peritoneal pseudomyxoma and improving the prognosis of affected patients.

METHODS

This is a descriptive, retrospective case report describing a rare case of Low-Grade Mucinous Neoplasm of the Vermiform Appendix treated at a referral center for general surgery.

Data collection was performed through the analysis of medical records, including information such as clinical data, clinical manifestations, laboratory and imaging tests, therapeutic management, intraoperative findings, histopathological results, clinical course, and postoperative outcomes.

The diagnosis was established based on histopathological findings obtained from tissue samples, following the diagnostic criteria recommended by the World Health Organization (2019) for neoplasms of the gastrointestinal tract.

Imaging studies consisted mainly of abdominal computed tomography (CT), whose reports were analyzed to characterize the appendicular lesion and verify the presence of signs of peritoneal dissemination.

The therapeutic approach was determined based on the patient's clinical presentation and intraoperative findings, in accordance with current recommendations for the management of low-grade appendiceal mucinous neoplasms.

The data obtained were organized and analyzed descriptively, without the use of statistical methods, given the nature of the study.

This study was conducted in accordance with ethical principles; the patient was duly informed about the study's objectives and signed the Informed Consent Form (ICF). The study was submitted for review to the Research Ethics Committee of the FACVEST University Center and was approved under Decision No. 7.953.455, in accordance with the provisions of Resolu-

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tion No. 466/2012 of the National Health Council.

CASE REPORT

A male adult patient sought medical care complaining of moderate abdominal pain in the lower abdomen, accompanied by nausea. He was initially seen in the emergency department of a hospital, where, following a clinical evaluation, no findings suggestive of an acute condition requiring immediate medical or surgical treatment were identified; consequently, the patient was discharged after being seen.

Approximately 24 hours later, the patient's condition worsened, characterized by intensified abdominal pain, now localized in the lower abdomen and right iliac fossa, accompanied by nausea and loss of appetite. The patient remained afebrile, with vital signs within normal parameters. Given the progression of symptoms, he sought care at a private clinic, where the clinical evaluation indicated the presence of a surgical condition requiring urgent intervention.

On physical examination, the patient presented with abdominal tenderness on deep palpation in the right iliac fossa, along with a positive Blumberg's sign, but without hemodynamic abnormalities or fever. Based on these clinical findings, the patient was admitted to the hospital for surgical treatment.

During the surgical procedure, intraoperatively, an extremely dilated vermiform appendix was identified, with significant edema and local inflammatory signs. An appendectomy was performed using a conventional technique, and the surgical specimen was duly sent for histopathological examination.

Histopathological analysis revealed findings consistent with low-grade mucinous neoplasia of the vermiform appendix, confirming the diagnosis of the pathology addressed in this study. In the postoperative period, the patient showed satisfactory clinical progress, without complications, with adequate recovery and was discharged from the hospital in good general condition.

RESULTS

The male patient presented with a clinical course consistent with an initially nonspecific acute abdominal condition, which progressed over a 24-hour period to localized pain in the right iliac fossa, associated with clinical signs suggestive of a surgical condition. The clinical evaluation and physical examination, which revealed a positive Blumberg's sign, were decisive in indicating a surgical approach.

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During the operative procedure, macroscopic findings revealed a significantly dilated vermiform appendix, with marked edema and local inflammatory signs, suggesting structural changes beyond the pattern observed in conventional acute appendicitis. The appendectomy performed using the classic technique allowed for complete removal of the affected structure and referral of the surgical specimen for histopathological analysis.

Histopathological examination confirmed the diagnosis of a low-grade mucinous neoplasm of the vermiform appendix, corroborating the suspicion of a rare condition that is difficult to diagnose clinically prior to surgery. Postoperatively, the patient had a satisfactory course, with no immediate complications, adequate clinical recovery, and was discharged from the hospital in good general condition.

DISCUSSION

Mucinous neoplasms of the vermiform appendix constitute a rare group of gastrointestinal tract tumors, accounting for less than 1% of appendiceal neoplasms, as widely described in the literature (Ma et al., 2025; Zhou et al., 2024). This low frequency contributes to healthcare professionals' limited familiarity with the condition, which can result in diagnostic delays or inappropriate therapeutic management (Zhou et al., 2024). The case presented confirms this pattern of rarity and reinforces the diagnostic challenge associated with this pathology, since its clinical presentation can be nonspecific and similar to more prevalent abdominal conditions (Faro et al., 2025; Zhou et al., 2024).

Similar to what has been described in previous studies, the patient presented with nonspecific clinical manifestations, which corroborates the difficulty reported by several authors in establishing a preoperative diagnosis of appendiceal neoplasms (Çakar et al., 2023; Pereira et al., 2021). The clinical presentation of these neoplasms is often nonspecific and may mimic acute appendicitis, chronic abdominal pain, or, in some cases, remain asymptomatic (D'Amata et al., 2024; Zambrano-Lechuga et al., 2025). In the literature, most cases are identified incidentally during surgical procedures or after pathological analysis of the surgical specimen, a situation also observed in the present study, reinforcing the need to consider this pathology in the differential diagnosis of atypical appendicular conditions (Çakar et al., 2023; Dohner et al., 2024).

With regard to imaging studies, abdominal computed tomography is described in the literature as the most commonly used method for the initial evaluation of mucinous appendiceal neoplasms, as it allows for the identification of findings such as appendiceal dilation, parietal thickening, and the presence of mucinous content; it is even more informative when combined with ultrasound, which can reveal additional abnormalities (Bai et al., 2023; Guo

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et al., 2024). However, these characteristics are not pathognomonic, which limits their definitive diagnostic value, since many findings can be observed in acute appendicitis or other abdominal lesions (Bai et al., 2023; Lu et al., 2024). In the present case, imaging studies revealed suggestive appendicular changes, but without unequivocal signs of advanced malignancy, corroborating previous reports and reinforcing that computed tomography is a relevant tool for diagnostic suspicion but insufficient for the isolated confirmation of the disease (Bai et al., 2023; Lathiya et al., 2023).

Diagnostic confirmation through histopathological analysis, considered the gold standard, proved decisive for the correct classification of low-grade mucinous neoplasia of the vermiform appendix, in accordance with the criteria established by the World Health Organization (2019). As described in the literature, histopathological evaluation allows for the differentiation of this entity from more aggressive appendiceal neoplasms, such as mucinous adenocarcinomas, based on histological characteristics such as cellular atypia, invasion patterns, and the extent of extracellular mucin, which directly impacts prognostic assessment and the determination of therapeutic management (Chang et al., 2024; Copur et al., 2021; Gundogar et al., 2018). Furthermore, recent studies indicate that preoperative diagnosis of these neoplasms is uncommon, with confirmation often occurring only after histopathological examination, a finding also observed in the present study, which highlights the limitations of clinical and radiological criteria alone for the accurate identification of this pathology (Guo et al., 2024; Maloku, 2023; Qian et al., 2024).

With regard to treatment, the surgical approach is widely recognized in the literature as the primary therapeutic strategy for low-grade appendiceal mucinous neoplasms, and complete and adequate resection of the appendix is considered sufficient in the absence of peritoneal spread, as observed in several clinical series (Aldakhil et al., 2025; Guner & Aydın, 2023; Lu et al., 2024). However, the described discrepancies regarding the extent of the surgical procedure—such as isolated appendectomy versus right hemicolectomy—underscore the need for individualized management based on intraoperative findings and the specific characteristics of each patient (Guner & Aydın, 2023; Istl et al., 2021).

From a clinical perspective, this case report underscores the importance of careful surgical management in the treatment of mucinous appendiceal neoplasms, particularly with regard to preventing rupture of the appendix, given the risk of developing peritoneal pseudomyxoma, a slowly progressing but potentially serious condition (Pareja et al., 2025; Sueda et al., 2020). The literature highlights that the prevention of this complication is directly related to appropriate surgical management, as well as to individualized treatment, taking into account factors such as the integrity of the appendix, surgical margins, and the presence of extravasated mucin (Guner & Aydın, 2023; Sipok et al., 2022). In this context, the present case demonstrates that careful intraoperative assessment is essential for appropriate decision-

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-making, in addition to reinforcing the need for long-term clinical and radiological follow-up, even in cases classified as low-grade.

Long-term clinical and radiological follow-up is recommended even for low-grade neoplasms, due to the possibility of late recurrence (Hannan et al., 2025; Lohani et al., 2024). In this regard, the present report reinforces the importance of continuous follow-up, as described in studies that demonstrate recurrences even after treatment considered adequate.

In terms of similarities, the case presented shares clinical, diagnostic, and therapeutic characteristics that have been widely described in the literature. However, differences related to the initial clinical presentation and the absence of advanced signs of the disease highlight the heterogeneity of this condition and reinforce the need for individualized assessment.

Regarding limitations, this study has constraints inherent to the design of a case report, such as the inability to generalize the results and the lack of comparative analysis with a larger patient sample. Furthermore, the limited follow-up period may restrict the assessment of long-term outcomes.

Finally, a critical interpretation of the case, combined with a comparison to the scientific literature, demonstrates that, although Low-Grade Mucinous Neoplasia of the Vermiform Appendix exhibits relatively indolent behavior, its diagnosis and management require specialized attention. The publication of additional case reports is essential to expand scientific knowledge, contribute to the standardization of management protocols, and improve clinical practice.

CONCLUDING REMARKS

Low-grade mucinous neoplasia of the vermiform appendix is a rare condition, often diagnosed incidentally, which can hinder early recognition and the determination of the best therapeutic approach. This case report contributes to the literature by describing the clinical, diagnostic, and therapeutic findings of this neoplasm, emphasizing the importance of clinical suspicion and appropriate diagnostic investigation.

The findings reinforce the fundamental role of histopathological analysis for diagnostic confirmation, as well as the relevance of imaging studies in assessing the extent of the disease and planning surgical treatment. The choice of individualized therapeutic management proved essential for the patient's favorable clinical outcome, highlighting the need for careful management to prevent complications, such as peritoneal dissemination.

Thus, this report emphasizes the importance of accurate diagnosis and appropriate follow-up of patients with low-grade appendiceal mucinous neoplasms, contributing to the advancement of scientific knowledge and evidence-based clinical practice. It is hoped that

this study will assist healthcare professionals in the recognition and management of this rare condition, as well as encourage further studies to expand understanding of its clinical behavior and prognosis.

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