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(CASE REPORT)

SPONTANEOUSLY RUPTURED APPENDICEAL MUCOCELE DIAGNOSED ON CROSS-SECTIONAL IMAGING: A CASE REPORT

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ABSTRACT

Appendiceal mucocele is a rare condition characterized by mucinous distension of the appendix. Spontaneous rupture is a serious complication that can lead to pseudomyxoma peritonei, a gelatinous disease of the peritoneum requiring aggressive management. We present the case of a 54-year-old man without prior surgical history who presented with acute right lower quadrant pain. Initial ultrasonography revealed localized fluid in the right iliac fossa. CT and MRI confirmed the diagnosis of spontaneously ruptured appendiceal mucocele. The patient underwent complete cytoreduction with appendicectomy and hyperthermic intra-peritoneal chemotherapy (CHIP). Histological findings were consistent with a low-grade mucinous neoplasm with pseudomyxoma peritonei. The patient had no postoperative complications. This case demonstrates that ultrasonography can detect early signs of rupture, while CT and MRI enable definitive diagnosis. Systematic evaluation for extraluminal mucin is essential when mucocele is suspected. CHIP represents an innovative therapeutic approach for pseudomyxoma peritonei.

Keywords: Appendiceal mucocele; Low-grade appendiceal mucinous neoplasm; Pseudomyxoma peritonei; Computed tomography; Magnetic resonance imaging; Cytoreductive surgery

1 INTRODUCTION

Appendiceal mucocele (AM), first described by Rokitansky in 1855, is a rare entity characterized by cystic dilatation of the appendix due to intraluminal accumulation of mucus [1,2]. It accounts for approximately 0.2-0.7% of appendectomy specimens [1,3]. Appendiceal mucoceles represent a heterogeneous group of lesions ranging from non-neoplastic conditions, such as retention cysts and mucosal hyperplasia, to mucinous neoplasms [2]. Although lesion size has been reported to show a general association with underlying pathology, with larger mucoceles more frequently corresponding to neoplastic lesions, considerable overlap exists and size alone is not a reliable discriminator [4].

According to the World Health Organization (WHO) Classification of Digestive System Tumours, mucinous neoplasms include low-grade appendiceal mucinous neoplasm (LAMN), high-grade appendiceal mucinous neoplasm (HAMN), and mucinous adenocarcinoma [5,6]. LAMN represents the most frequent neoplastic subtype and is typically characterized by an indolent behavior; however, it may be associated with appendiceal rupture and peritoneal dissemination when the wall is compromised [7]. Mucinous adenocarcinoma is a frankly malignant entity associated with a worse prognosis and a higher risk of peritoneal spread [6].

Although often asymptomatic and incidentally detected, appendiceal mucocele may rupture spontaneously, resulting in intraperitoneal mucin dissemination and potentially leading to pseudomyxoma peritonei (PMP), a rare but severe condition requiring aggressive surgical management [8]. Spontaneous rupture is uncommon, as most reported cases are related to iatrogenic injury during surgery [9].

Cross-sectional imaging, particularly computed tomography (CT) and magnetic resonance imaging (MRI), plays a central role in diagnosis, detection of complications, and preoperative assessment of disease extent [5]. CT typically demonstrates a well-defined, low-attenuation cystic appendiceal lesion, sometimes associated with mural calcifications, while MRI provides superior soft-tissue contrast and improved detection of mucinous peritoneal deposits [10]. In cases with peritoneal dissemination, the standard of care consists of cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (HIPEC), which has significantly improved long-term outcomes [11].

We report a case of spontaneously ruptured appendiceal mucocele diagnosed on imaging and successfully treated with cytoreductive surgery and HIPEC, with favorable postoperative outcome.

2 CASE PRESENTATION

A 54-year-old man presented with the acute onset of right iliac fossa pain. The patient was apyretic. Physical examination revealed localized tenderness in the right lower quadrant without signs of generalized peritonitis. Laboratory tests were unremarkable, with no evidence of systemic inflammation.

Abdominal ultrasound demonstrated a localized fluid collection in the right iliac fossa, associated with marked acoustic shadowing and gaseous artifacts, limiting optimal assessment. Contrast-enhanced computed tomography (CT) of the abdomen and pelvis revealed a cystic appendiceal lesion with partially calcified walls. The lesion contained liquid material and demonstrated a small enhancing mural component. A focal discontinuity of the appendiceal wall was identified, consistent with rupture. Adjacent peri-appendiceal fluid collection was noted, in keeping with localized mucin spillage. Additional perihepatic fluid was also present (figure 1, 2, 3). CT demonstrated infiltration of the greater omentum and the peritoneum in the right iliac fossa.

Magnetic resonance imaging (MRI) confirmed the cystic nature of the lesion, which appeared hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences (figure 2,3), consistent with mucinous content. MRI further confirmed the presence of perilesional and peritoneal fluid.

Based on imaging findings, a ruptured appendiceal mucocele with suspected mucinous peritoneal dissemination was suggested. Exploratory laparotomy revealed diffuse mucinous deposits involving the diaphragmatic domes and the right paracolic gutter, consistent with pseudomyxoma peritonei. A complete cytoreductive surgery was performed, including right hemicolectomy with mechanical ileocolic anastomosis, omentectomy, and peritoneal stripping of the diaphragmatic surfaces. A cholecystectomy was also performed to prevent potential gallbladder complications during hyperthermic intraperitoneal chemotherapy (HIPEC). HIPEC was subsequently administered following complete cytoreduction.

The resected specimens were consistent with a low-grade appendiceal mucinous neoplasm (LAMN) with associated peritoneal mucinous deposits, in keeping with pseudomyxoma peritonei (PMP). Histopathological images were not available for inclusion in this report.

The postoperative course was uneventful. Postoperative CT was unremarkable, showing no residual disease or postoperative complications (figure 4).

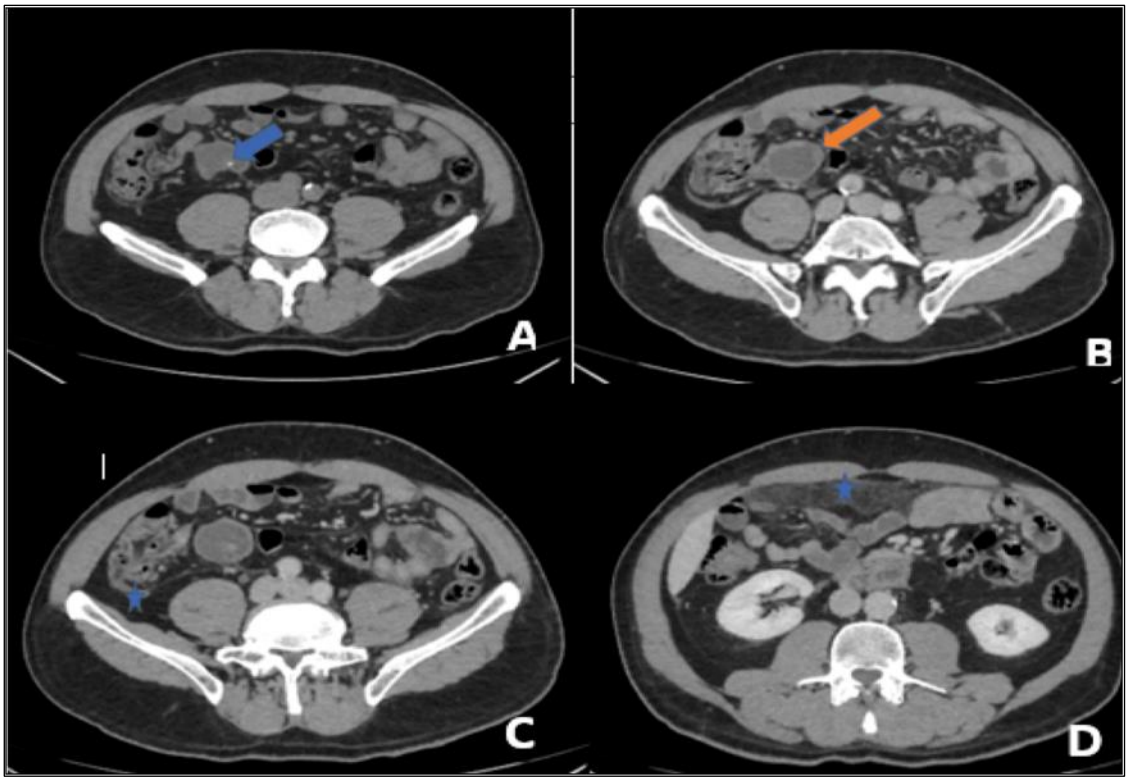

Figure 1: Axial pre-contrast (A) and post-contrast (B–D) CT images demonstrating a cystic appendiceal lesion containing a mural calcification (blue arrow), with an irregular thickened and enhancing wall (orange arrow), associated with infiltration of the peritoneum of the right paracolic gutter (asterisk).

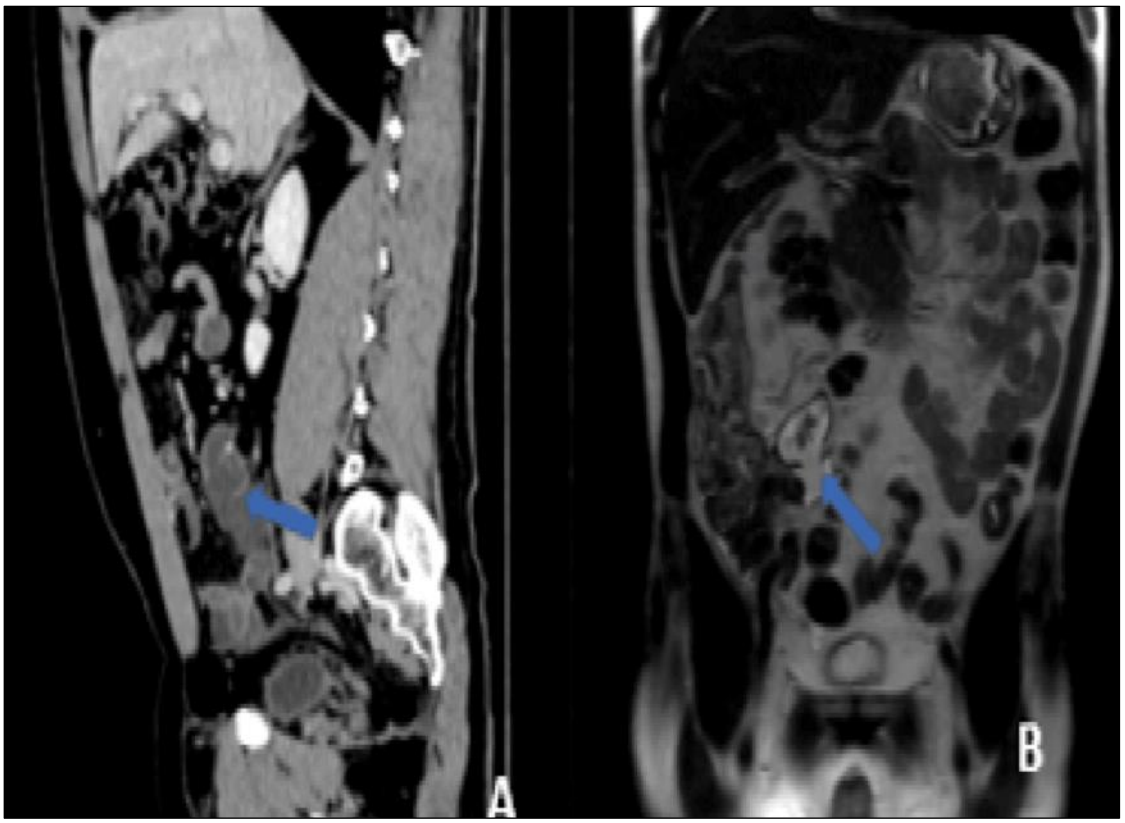

Figure 2: Contrast-enhanced sagittal CT (A) and coronal T2-weighted MRI showing focal discontinuity of the wall of the appendiceal lesion at its inferior pole(arrow), with prolapse of its mucinous content into the adjacent peritoneal space.

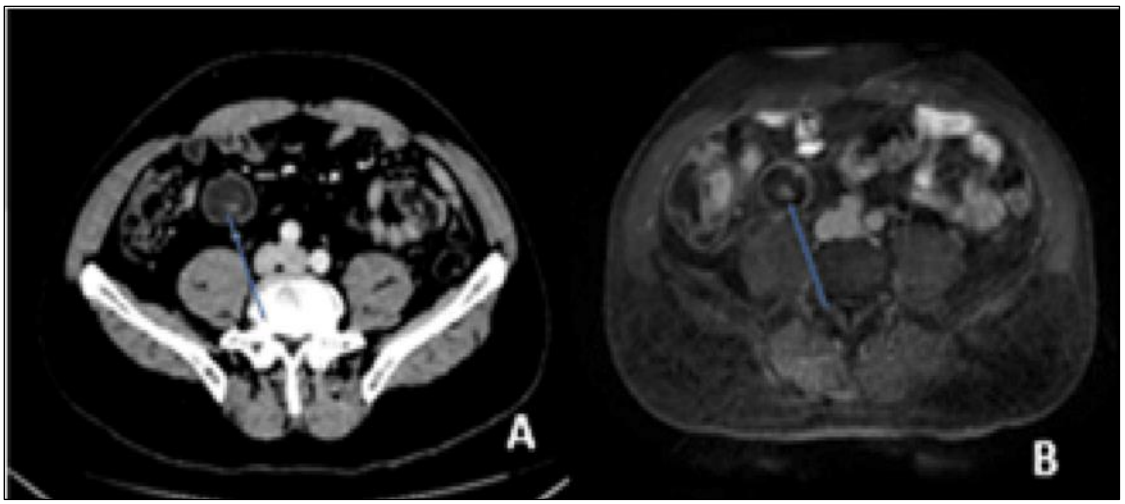

Figure 3: Axial contrast-enhanced CT (A) and axial fat-suppressed T1-weighted post-contrast MRI demonstrating a nodular enhancing component within the appendiceal lesion (arrow).

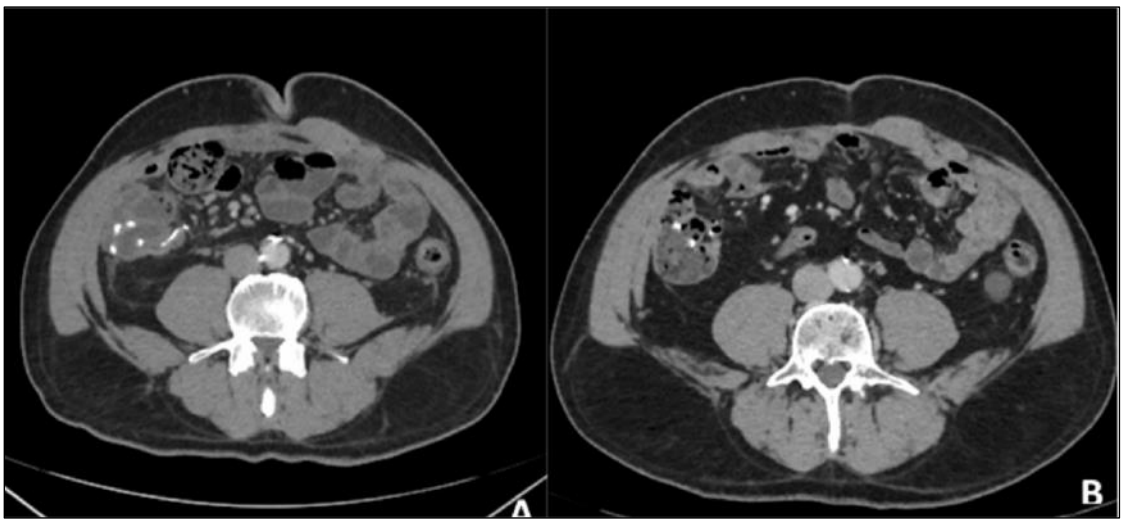

Figure 4: Axial CT images obtained at 3 months (A) and 6 months (B) postoperatively, showing no residual lesion or postoperative complications.

3 DISCUSSION

Appendiceal mucocele is a rare entity encompassing a heterogeneous spectrum of lesions ranging from non-neoplastic conditions, such as retention cysts and mucosal hyperplasia, to mucinous neoplasms. According to the World Health Organization (WHO) Classification of Digestive System Tumours, mucinous neoplasms include low-grade appendiceal mucinous neoplasm (LAMN), high-grade appendiceal mucinous neoplasm (HAMN), and mucinous adenocarcinoma [2].

LAMN is the most frequent subtype and typically follows an indolent clinical course; however, it carries a risk of appendiceal wall rupture with dissemination of mucin into the peritoneal cavity. Mucinous adenocarcinoma, in contrast, is a malignant entity associated with more aggressive behavior and a higher propensity for peritoneal spread [2,6].

Clinically, appendiceal mucocele is often asymptomatic or presents with non-specific abdominal pain, resulting in frequent diagnostic delay. Physical examination is often unremarkable, while laboratory inflammatory markers are frequently normal, even in complicated cases. This discrepancy between clinical findings and laboratory results contributes to delayed recognition and underscores the importance of imaging in atypical abdominal presentations [7].

Cross-sectional imaging plays a central role in diagnosis and preoperative assessment. Computed tomography (CT) typically demonstrates a well-defined cystic dilatation of the appendix filled with low-attenuation mucinous content, occasionally associated with mural calcifications [3,9]. Features suggestive of rupture include focal wall discontinuity, periappendiceal fluid collections, and mucinous deposits within the peritoneal cavity, particularly in dependent regions such as the pelvis, paracolic gutters, and subphrenic spaces [3,5].

Magnetic resonance imaging (MRI) provides complementary value due to superior soft-tissue characterization. Mucin typically appears hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences, with variability depending on protein concentration and viscosity [8,12]. MRI is particularly useful for detecting low-volume peritoneal disease, which may be underestimated on CT [5,12].

Despite advances in imaging, preoperative assessment may underestimate the true extent of peritoneal dissemination, especially in early pseudomyxoma peritonei (PMP). Therefore, surgical exploration remains essential for definitive staging [5].

Spontaneous rupture of an appendiceal mucocele is uncommon but clinically significant, as it may result in intraperitoneal dissemination of mucin and epithelial cells, leading to pseudomyxoma peritonei [3,5].

The standard of care for peritoneal dissemination consists of complete cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (HIPEC) [11,12]. In this patient, complete cytoreduction (CC-0) was achieved. Tumor markers, including CEA, CA 19-9, and CA-125, were within normal limits [5]. The resected specimens were consistent with LAMN with associated peritoneal mucinous deposits (PMP). Follow-up imaging at 3, 6, 12 and 24 months demonstrated no evidence of disease recurrence, supporting the effectiveness of complete cytoreduction combined with HIPEC in selected patients.

In the present case, CT and MRI were instrumental in suggesting the diagnosis of ruptured appendiceal mucocele and in assessing early peritoneal involvement. These findings were confirmed intraoperatively, revealing diffuse mucinous deposits involving the diaphragmatic domes and the right paracolic gutter.

Appendiceal mucocele should be considered in the differential diagnosis of a cystic right lower quadrant mass, particularly when mural calcifications are present on CT. Early recognition of imaging features suggestive of rupture is crucial, as timely diagnosis allows appropriate surgical planning and may prevent progression to extensive pseudomyxoma peritonei [3,5].

4 CONCLUSION

Appendiceal mucocele is an uncommon entity that often has a non-specific clinical presentation, frequently resulting in delayed or incidental diagnosis. Cross-sectional imaging, particularly CT and MRI, is essential for early detection, lesion characterization, and assessment of peritoneal extension, especially in cases of rupture with suspected pseudomyxoma peritonei.

Accurate radiological recognition is critical for guiding timely surgical management and preventing peritoneal dissemination. When complete cytoreduction is achieved, HIPEC offers an effective therapeutic strategy associated with favorable long-term outcomes in selected patients.

Early radiological recognition can fundamentally alter prognosis by preventing progression from a localized appendiceal lesion to diffuse peritoneal disease.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

Disclosure of conflict of interest

The authors declare that they have no conflict of interest.

Statement of informed consent

Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

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