

Appendiceal Mucoceles: Diagnostic Imaging in A 28-Case Series

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Abstract

Case Series

Appendiceal mucoceles are rare entities, accounting for approximately 0.2-0.3% of appendectomy specimens. They correspond to progressive distension of the appendix of obstructive origin, secondary to intraluminal accumulation of mucoid material. Their clinical presentation is often nonspecific or absent, and they are frequently discovered incidentally, particularly in patients older than 50 years. However, preoperative diagnosis is essential because it helps guide the surgical approach and prevents iatrogenic rupture, which may lead to intraperitoneal dissemination of mucin and pseudomyxoma peritonei. We report a retrospective imaging study of 28 cases of appendiceal mucocele managed at the University Hospital Center of Marrakech between January 2020 and December 2023. The aim of the study was to assess the contribution of imaging, particularly ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI), in the diagnosis, characterization, detection of complications, and radiological-histopathological correlation of appendiceal mucoceles. In our series, the mean age was 59.7 years, with a female predominance. Most cases were discovered incidentally. On CT, the typical appearance was a well-defined cystic lesion arising from or communicating with the cecum, usually located in the right iliac fossa, with fluid attenuation, smooth contours, and occasionally mural calcifications. MRI was useful in cases mimicking pelvic or adnexal masses. Imaging also identified signs suggestive of malignancy and complications, including pseudomyxoma peritonei and appendico-colic intussusception. These findings confirm the central role of imaging in the preoperative assessment of appendiceal mucoceles and in surgical planning.

Keywords: Appendiceal mucocele; appendix; computed tomography; ultrasound; MRI; pseudomyxoma peritonei; appendiceal mucinous neoplasm.

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INTRODUCTION

Appendiceal mucocele is a rare condition, reported in approximately 0.2-0.3% of appendectomies [1,3,4]. It is defined as progressive dilatation of the appendiceal lumen related to obstruction and intraluminal mucus accumulation. This distension may be secondary to a non-neoplastic obstructive process or to a mucinous appendiceal tumor, ranging from low-grade lesions to mucinous adenocarcinoma.

Clinically, appendiceal mucocele is often asymptomatic and discovered incidentally, especially after the age of 50 years. When symptomatic, it may present with right iliac fossa pain, bowel transit disorders, an abdominal mass, or a clinical picture mimicking acute appendicitis. In some cases, the diagnosis is made intraoperatively or only after histopathological examination.

Preoperative diagnosis is of major importance. It allows appropriate surgical planning, helps avoid iatrogenic rupture of the lesion, and prevents

intraperitoneal spread of mucinous material, a complication that may result in pseudomyxoma peritonei. Ultrasound may suggest the diagnosis, but CT is the key imaging modality for lesion characterization, evaluation of cecal relationships, detection of mural calcifications, assessment of suspicious features, and identification of complications. MRI may be useful in selected cases, particularly when the lesion mimics a pelvic or adnexal mass.

Objectives

The objectives of this study were to evaluate the contribution of imaging to the diagnosis of appendiceal mucoceles, describe the different radiological presentations observed in our series, and correlate radiological findings with operative and histopathological data.

MATERIALS AND METHODS

This was a retrospective study including 28 cases of appendiceal mucocele collected from the archives of the Radiology Department of the University

Hospital Center of Marrakech. All included patients underwent diagnostic and surgical management in the same hospital and had available cross-sectional imaging, either CT or MRI. Cases diagnosed by only ultrasound were excluded.

The study period extended over 4 years, from January 2020 to December 2023. The evaluation included clinical data, imaging findings, and comparison with operative and histopathological results.

RESULTS

Epidemiological and clinical data

The series included 28 cases. The mean age was 59.7 years, with a range from 39 to 78 years. A female

predominance was observed, with 18 women, representing 64% of cases, and 10 men, representing 36%. The lesion was discovered incidentally in 23 patients, whereas 5 patients were symptomatic. Clinical presentations included acute appendicitis-like symptoms with right iliac fossa pain in 3 patients, nonspecific abdominal pain in 1 patient, and pelvic heaviness with an adnexal pelvic mass in 2 female patients. Laboratory tests showed an inflammatory syndrome with leukocytosis in symptomatic patients presenting with abdominal pain.

Tumoral history or particular clinical contexts identified in the series are summarized in Table 1.

Table 1: Tumoral history and particular clinical contexts identified in the series.

Clinical background or context	Number of patients
Metastatic breast cancer under chemotherapy	4
Pulmonary adenocarcinoma under chemotherapy	1
Plasmablastic lymphoma under chemotherapy	1
Metastatic rectal adenocarcinoma under chemotherapy	1
Pleomorphic sarcoma of the thigh	1
Darier-Ferrand dermatofibrosarcoma	1

Ultrasound findings

Ultrasound showed variable appearances, but the key diagnostic feature was the intimate contact between the cystic mass and the cecum, with non-visualization of a normal appendix. The most frequent appearance was an appendiceal cystic mass with anechoic, finely echogenic, or echogenic content. Two cases showed appendiceal fluid retention upstream of an

obstructing stercolith associated with fat infiltration, and one case showed simple fluid retention. In two cases, the wall was focally thickened with a multilayered echogenic onion-skin appearance. Two patients had a swollen appendix with fat infiltration, and one case appeared as a heterogeneous hypoechoic mass in the right iliac fossa.

Table 2: Main ultrasound appearances of appendiceal mucocèles.

Ultrasound appearance	Number of patients
Appendiceal cystic mass with anechoic, finely echogenic, or echogenic content	14
Intimate contact with the cecum or continuity with the cecum, with absence of an identifiable normal appendix	All relevant cases
Appendiceal fluid retention upstream of a stercolith with fat infiltration	2
Simple fluid retention	1
Focally thickened wall with multilayered echogenic onion-skin content	2
Swollen appendix with fat infiltration	2
Heterogeneous hypoechoic mass of the right iliac fossa	1

CT findings

On CT, the lesion was located in the right iliac fossa in 26 patients and extended to the right flank in 2 patients. It showed close contact with the cecum, appearing attached, adherent, connected to, or continuous with it. The lesion was retrocecal in 2 cases, paracecal in 25 cases, and intussuscepted into the cecum in 1 case. Its shape was oval in 16 cases, oblong in 8 cases, and round in 4 cases. The mean long-axis diameter was 46.9 mm, with a range from 16 to 110 mm.

The wall was thin in most cases, focally mildly thickened in 2 patients, and associated with a mural nodule in 1 patient. The margins were regular and smooth in 25 patients, macrolobulated in 2 patients, and irregular in 1 patient. Fine mural calcifications were present in 4 patients. A stercolith was identified in 4 cases. The lesion had fluid attenuation in 26 cases and a heterogeneous appearance in 2 cases. Septations were seen in 4 patients. Enhancement was absent in 21 cases; septal enhancement was noted in 2 cases, mural enhancement in 4 cases, and enhancement of a mural nodule in 1 case. Fat stranding was absent in 23 patients,

mild in 1 patient, and marked in 4 patients. Locoregional lymph nodes were observed in 2 cases. Peritoneal effusion was absent except in the 2 cases of pseudomyxoma peritonei.

The typical CT appearance was a well-defined cystic lesion connected to the cecum, with smooth margins, fluid attenuation, occasional septations, usually without enhancement, and sometimes containing fine mural calcifications.

Table 3: CT features of appendiceal mucoceles

CT parameter	Findings
Location	Right iliac fossa: n = 26; right iliac fossa + right flank: n = 2
Relationship with the cecum	Close contact with the cecum; retrocecal: n = 2; paracecal: n = 25; intussuscepted into the cecum: n = 1
Shape	Oval: 16; oblong: 8; round: 4
Size	Mean long-axis diameter = 46.9 mm; range: 16-110 mm
Wall	Thin in most cases; focally mildly thickened: 2; mural nodule: 1
Margins	Regular and smooth: 25; macrolobulated: 2; irregular: 1
Fine mural calcifications	4
Stercolith	4
Attenuation	Fluid attenuation: 26; heterogeneous: 2
Septations	4
Enhancement	Absent: 21; septal: 2; mural: 4; mural nodule: 1
Fat stranding	Absent: 23; mild: 1; marked: 4
Locoregional lymph nodes	2
Peritoneal effusion	Absent, except in 2 cases of pseudomyxoma peritonei

MRI findings

Five patients in our series underwent MRI as the initial imaging modality, mainly because of suspected or characterization of a pelvic-adnexal mass. Signal intensity varied from case to case, probably depending

on mucin content. A thin wall with mural enhancement was found in 2 cases. One case showed an irregular wall with a few enhancing septations. No ADC restriction was identified.

Table 4: MRI signal of appendiceal mucoceles in documented cases.

Case	T1 signal	T2 signal
Case 1	Hyperintense	Intermediate
Case 2	Intermediate	Hyperintense
Case 3	Hypointense	Hyperintense
Case 4	Hyperintense	Hyperintense

Illustrative cases, associated findings, and histopathological results

A first case involved a 47-year-old woman referred for a right-sided cystic lesion suspected to be adnexal in origin. Unenhanced abdominal CT showed a cystic mass in the right iliac fossa adherent to the lower cecum, with focal mural thickening. Oblique coronal reconstruction demonstrated the connection of the appendiceal mucocele with the cecal wall. MRI showed an oblong cystic lesion in the right iliac fossa, with intermediate T1 signal and high T2 signal, in close contact with the cecal base. Histopathological examination at low magnification showed a pseudo-villous epithelial proliferation, consistent with a low-grade appendiceal mucinous neoplasm (LAMN), without evidence of invasion of the lamina propria. The specimen showed remodeling and low-grade peritoneal implants, with a free surgical margin located 10 mm from the lesion. (Fig 1,2)

Associated findings and complications included 2 cases of pseudomyxoma peritonei, 1 case of cecal

intussusception of the mucocele, and 2 simple ovarian cysts of variable size. No synchronous or metachronous malignant ovarian tumor was found. Available histopathological results included 4 cases of low-grade mucinous tumor, 2 cases of cystadenocarcinoma, 2 cases of cystadenocarcinoma with pseudomyxoma peritonei, and 5 cases of panappendicitis.

Among the complicated cases, a 65-year-old woman had pseudomyxoma peritonei secondary to appendiceal cystadenocarcinoma, documented at initial diagnosis and during follow-up at 6 months and 2 years (Fig3) Another case involved a 70-year-old woman who presented to the emergency department with right flank pain and a positive infectious work-up. Imaging showed an enlarged appendix with fluid retention upstream of a stercolith, associated with an obstructing common bile duct stone responsible for cholangitis. Histopathology was benign, showing suppurative panappendicitis after conventional appendectomy, with a favorable postoperative course.

One case in a 66-year-old patient with lung cancer under chemotherapy was discovered incidentally. Imaging showed an intracecal fluid-attenuation mass with peripheral calcifications, which could easily have

been overlooked. The mass was continuous with the appendix, leading to the diagnosis of appendico-colic intussusception of an appendiceal mucocele, corresponding to the ball-and-cup sign. **(Fig4)**

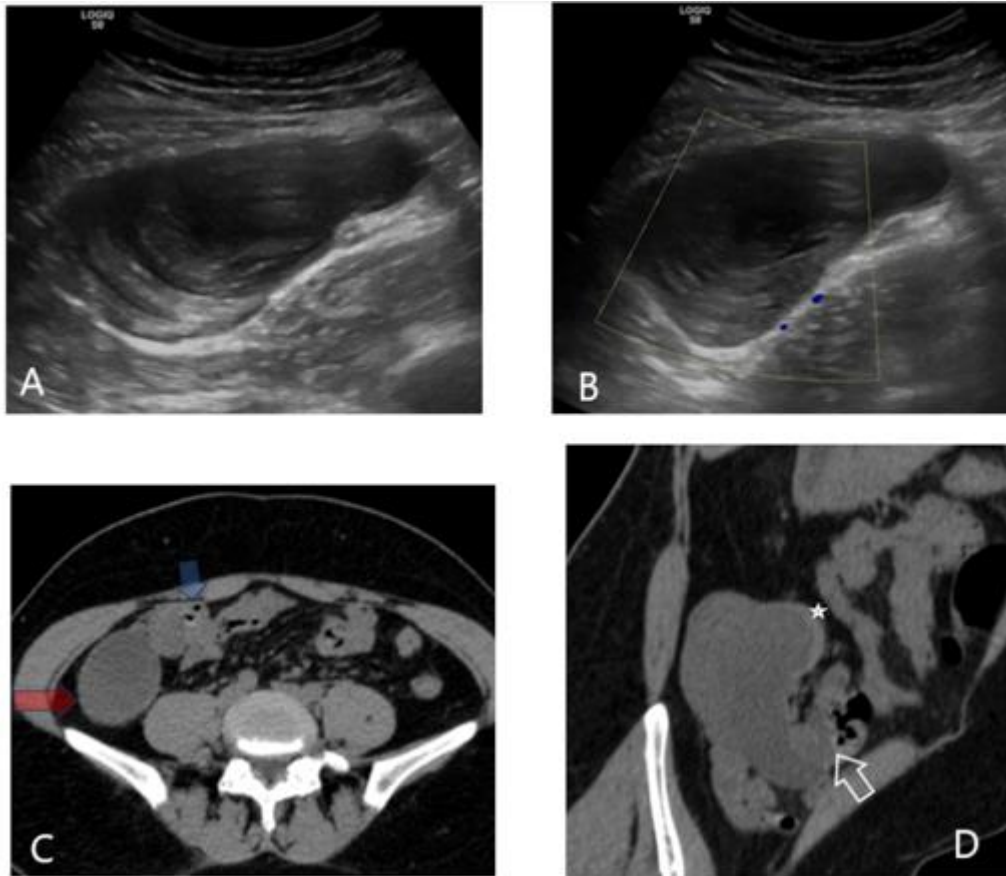


Figure 1: A 47-year-old female patient referred for a right-sided cystic lesion suggestive of an adnexal mass on ultrasound

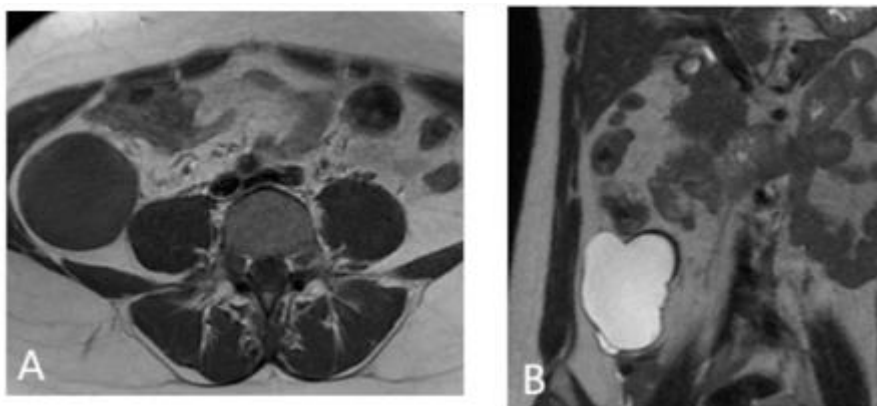
A: B-mode ultrasound showing a cystic lesion in the right iliac fossa, with lobulated margins and stratified echogenic content with an onion-skin appearance.

B: Absence of vascularity on color Doppler imaging.

C: Unenhanced axial CT image showing a cystic mass in the right iliac fossa (red arrow) communicating with the cecum (blue arrow).

D: Unenhanced coronal CT image showing a cystic mass in the right iliac fossa communicating with the cecum (arrow).
Note

The small focal mural thickening (star).



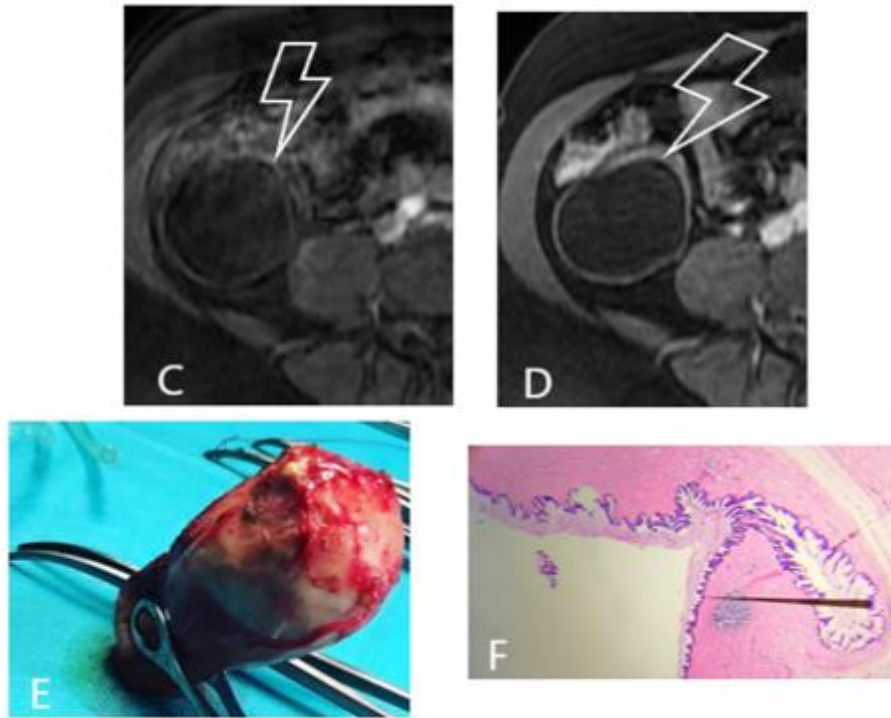


Figure 2: Same patient as in Figure 1

A: Axial T1-weighted MRI showing an oblong cystic lesion in the right iliac fossa with intermediate T1 signal intensity.

B: Coronal T2-weighted MRI showing an oblong cystic lesion with marked T2 hyperintensity, reaching close contact with the cecal base.

C and D: Pre- and post-contrast fat-suppressed T1-weighted MRI showing mural enhancement.

E: Surgical specimen.

F: Low-power histopathological appearance, H&E ×40, showing epithelial proliferation with pseudo-villous architecture.

Histological diagnosis: Morphological features consistent with a low-grade appendiceal mucinous neoplasm (LAMN), without evidence of lamina propria invasion. The lesion showed remodeling and was

associated with low-grade peritoneal implants. The surgical resection margin was free, located 10 mm from the lesion.

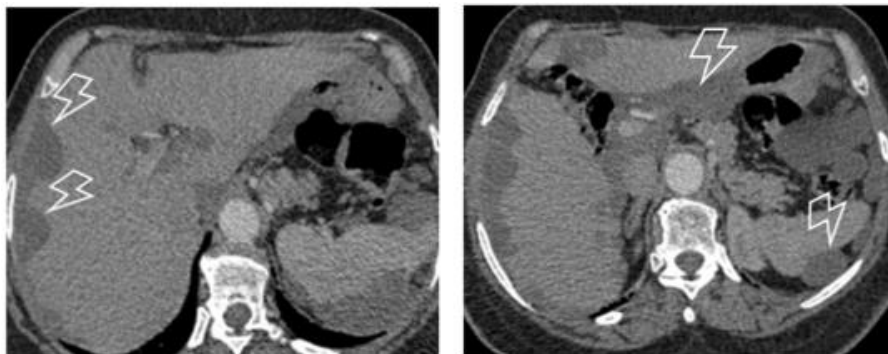


Figure 3: 65-year-old female patient: Peritoneal pseudomyxoma associated with appendiceal cystadenocarcinoma at CT: low-attenuating mass like lesions (Arrows) with loculated fluid throughout the peritoneum performing a scalloping of visceral surfaces, particularly the liver.

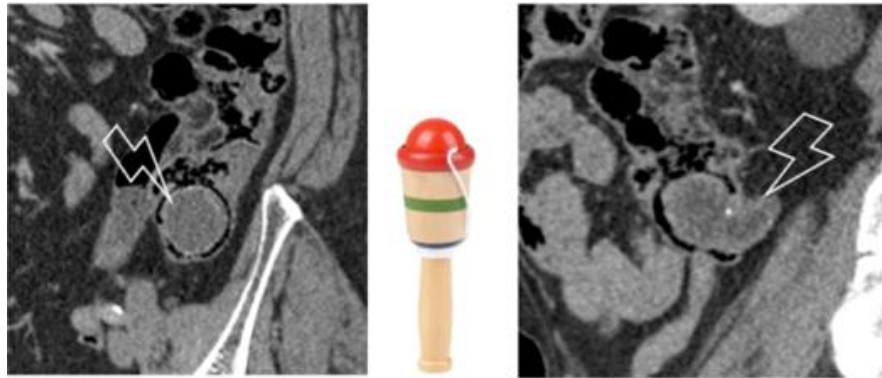


Figure 4: 66-year-old patient with lung cancer undergoing chemotherapy. Incidental finding

Intracaecal fluid-attenuation mass with peripheral calcifications, which may be easily overlooked. This appearance reproduces the “ball-in-

cup” sign, indicating invagination of an appendiceal mucocele into the caecal lumen (Arrows)

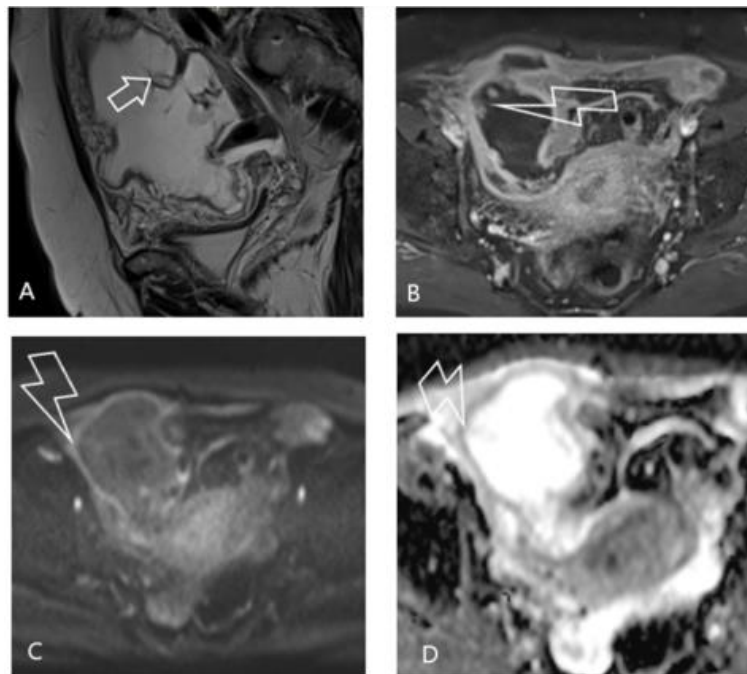


Figure 5: MRI of Aa73-year-old man presented for evaluation of a pelvic mass: An appendicular cystadenocarcinoma has been confirmed

- A: T2-weighted sequence showing lobulated contours with mural nodules (arrow).
- B: Contrast-enhanced T1-weighted sequence showing enhancement of the wall and mural nodules (arrow).
- C and D: Diffusion-weighted sequences showing areas of restricted diffusion (arrow).

DISCUSSION

Appendiceal mucocele remains a rare condition, representing approximately 0.25-0.3% of appendectomy specimens [1,3,4]. It corresponds to appendiceal dilatation due to mucus accumulation, often with a diameter greater than 15 mm. Its clinical presentation is nonspecific: it may be asymptomatic, discovered incidentally, or present with abdominal pain, bowel transit disorders, a palpable mass, or acute appendicitis-like symptoms.

Histologically, appendiceal mucoceles have classically been divided into four entities: retention cyst,

related to non-neoplastic obstruction by a stercolith or inflammatory stenosis; epithelial villous hyperplasia, corresponding to simple mucocele; mucinous cystadenoma, historically considered the most frequent benign mucinous tumor; and mucinous cystadenocarcinoma, characterized by malignant features and an increased risk of perforation. According to the 2019 WHO classification of appendiceal mucinous neoplasms, the main entities are low-grade appendiceal mucinous neoplasm (LAMN), high-grade appendiceal mucinous neoplasm (HAMN), and appendiceal mucinous adenocarcinoma [1].

The major complication of appendiceal mucoceles is pseudomyxoma peritonei, favored by spontaneous or intraoperative rupture of a mucocele, usually malignant. It corresponds to the intraperitoneal presence of mucus-secreting neoplastic cells. Three histological forms are described according to cellularity and atypia: disseminated peritoneal adenomucinosis, with sparse cells, limited atypia, and few mitoses; peritoneal mucinous carcinomatosis, characterized by numerous malignant cells; and an intermediate form. Other complications include intra-abdominal abscess, torsion, acute appendicitis, and endoceleal intussusception.

An associated ovarian mucinous tumor should be sought, with a reported frequency of approximately 18%. This association may reflect the independent development of two tumors or metastatic spread from one site to the other. In our series, no synchronous or metachronous malignant ovarian tumor was identified. An exceptional macroscopic form, occurring in less than 1% of cases, is myxoglobulosis, an appendiceal mucocele characterized by opaque globules of mucin.

Ultrasound can be highly contributive when it shows a well-defined paracecal cystic lesion with homogeneous fluid, hypoechoic, or anechoic content, sometimes with internal echoes, septations, or mural thickening. The key diagnostic feature is demonstration of the relationship between the mass and the cecum, with non-visualization of a normal appendix. A complex cystic mass with calcifications may also be observed. The onion-skin sign, corresponding to layered echogenic content, is considered characteristic.

On contrast-enhanced CT, appendiceal mucocele typically appears as an appendiceal fluid-attenuation lesion, sometimes septated, with a thin or finely calcified wall that may enhance. Imaging features suggestive of malignancy include large size, irregular margins, thickened or nodular wall, heterogeneous appearance, a solid component, calcifications within solid areas, and relatively thick or dense peritoneal fluid. In pseudomyxoma peritonei, scalloping of the liver and splenic surfaces may be present.

MRI may be helpful when the diagnosis is difficult, particularly in pelvic forms mimicking an adnexal mass. The signal varies according to mucin content, ranging from low to intermediate signal on T1-weighted images and high signal on T2-weighted images. MRI also allows better assessment of the soft-tissue component and contributes to differential diagnosis with pelvic masses.

The differential diagnosis may be difficult with an appendiceal abscess or appendiceal phlegmon, especially in the presence of fat stranding and an inflammatory clinical context. Other differential diagnoses include other appendiceal tumors, particularly

appendiceal carcinoma, ovarian or pelvic cystic masses, and peritoneal mucinous carcinomatosis. The latter may show solid tissue components within ascites or nodules suggestive of peritoneal carcinomatosis, and should be differentiated from carcinomatosis secondary to ovarian mucinous tumors.

Management is surgical. In the absence of rupture, treatment is based on appendectomy performed without rupturing the appendix, complete excision of the mesoappendix, and sampling of peritoneal fluid for cytology. In cases of pseudomyxoma peritonei, management may be more extensive and includes radical cytoreductive surgery with removal of as much mucus and tumor deposits as possible, including omentectomy, localized peritonectomy, right hemicolectomy, and bilateral oophorectomy in women, usually combined with intraoperative hyperthermic intraperitoneal chemotherapy.

In our series, patients were mostly older adults, with a mean age of 59.7 years, and there was a female predominance. Discovery was most often incidental, which may be partly related to the study selection criteria. A background of malignant neoplastic disease, often associated with chemotherapy, was found in several patients, probably reflecting a selection bias related to oncologic imaging follow-up. Ultrasound was diagnostically relevant when it demonstrated a cystic lesion in continuity with the cecum, but CT and MRI were more sensitive and more specific for diagnosis, lesion characterization, assessment of suspicious features, and detection of complications. The imaging criteria suggestive of malignancy in our series were consistent with those reported in the literature, particularly by Wang *et al.*, and Yakan *et al.*, Two cases of pseudomyxoma peritonei were observed, and one relatively rare complication, the ball-and-cup sign related to appendico-colic intussusception, was identified. No association with an ovarian mucinous tumor was found.

CONCLUSION

Appendiceal mucocele should never be considered a trivial finding. Although rare and often discovered incidentally, it may be associated with severe complications, particularly pseudomyxoma peritonei, whose therapeutic management can be complex and aggressive. Preoperative diagnosis is therefore essential in order to adapt the surgical strategy, avoid iatrogenic rupture, and improve prognosis.

Imaging plays a central role in this diagnostic approach. Ultrasound can suggest the diagnosis, whereas CT remains the reference modality for characterizing the lesion, analyzing its relationship with the cecum, detecting mural calcifications, identifying suspicious features, and assessing complications. MRI is particularly useful in pelvic forms or when

differentiation from adnexal masses is required. Early diagnosis of a limited lesion is associated with a more favorable prognosis.

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