

Appendiceal Mucocele: Clinical and Imaging Features of 14 Cases

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Rezumat

Mucocel apendicular: aspecte clinice și imagistice în cadrul unei serii de 14 cazuri

Scop: Mucocelul apendicular, dilatație chistică conținând material mucinos, este o patologie rară a apendicelui vermiform. Posibilitățile de diagnostic preoperator sunt în continuare limitate de mijloacele imagistice de bază disponibile.

Metode: Analiză retrospectivă a pacienților cu diagnostic anatomopatologic de mucocel apendicular cu accent asupra aspectelor clinice și imagistice.

Rezultate: Grupul de studiu a inclus 14 pacienți cu vârsta medie de 51 de ani (limite 17-82 de ani). Simptomele predominante au fost durere și senzație de plenitudine în fosa iliacă dreaptă la 9 (64%), respectiv 5 (36%) pacienți. În scopuri imagistice, utilizarea tomografiei computerizate a determinat diagnosticul preoperator al mucocelului apendicular la jumătate din pacienți (50%). 93% din cazuri au fost supuse apendicectomiei, iar hemicolectomie dreaptă a fost efectuată în cazul unui pacient (7%). Mucocel și chistadenom au fost diagnosticate la 11 (79%) și, respectiv, 3 (21%) pacienți. Prezența apendicitei acute și a cancerului de colon au fost confirmate ulterior histologic la 4 (29%) și, respectiv, 1 (7%) pacient.

Concluzii: În ciuda utilizării mijloacelor imagistice, diagnosticul preoperator al mucocelului apendicular este în continuare dificil de stabilit în majoritatea cazurilor. Pe parcursul trata-

mentului chirurgical, adaptat descoperirilor imagistice și intraoperatorii, trebuie luate măsuri pentru a preveni ruptura intra-peritoneală și diseminarea.

Cuvinte cheie: mucocel, apendice, tomografie computerizată, apendicectomie

Abstract

Purpose: Appendiceal mucoceles as a cystic dilatation filled with mucinous material is a very rare disease of the appendix vermiformis. Its preoperative diagnosis is still lacking behind common use of imaging techniques.

Methods: Retrospective analysis of the patients with a pathological diagnosis of appendiceal mucoceles with regard to clinical and imaging features.

Results: The study group included 14 patients with a mean age of 51 years (range from 17 to 82 years). Predominant symptoms were pain and feeling of fullness in the right iliac fossa in 9 (64%) and 5 (36%) patients, respectively. For imaging purposes, use of computed tomography resulted in preoperative diagnosis of appendiceal mucoceles in half of the patients (50%). 93% of the cases underwent appendectomy, and right hemicolectomy was performed in one patient (7%). Mucoceles and cystadenoma were detected in 11 (79%) and 3 (21%) patients, respectively. Presence of acute appendicitis and colon carcinoma were confirmed afterwards histologically in 4 (29%) and one (7%) patients, respectively.

Conclusions: Despite the common use of imaging studies, preoperative diagnosis of appendiceal mucoceles is still not possible in most of the cases. During surgical treatment, which is tailored according to imaging and intraoperative

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findings, precautionary measures to avoid intraperitoneal rupture and dissemination should be taken.

Key words: mucocoele, appendix, computed tomography, appendectomy.

Introduction

Appendiceal mucocoele (AM) is defined as a cystic dilatation filled with mucinous material (1). Although it is a very rare disease of the appendix vermiformis which is seen in 0.2 to 0.3 % of all appendectomies, its preoperative diagnosis remains to be an important issue for differential diagnosis of several acute and elective abdominal conditions (2-4). In almost half of the cases, the diagnosis is shown to be incidental and misdiagnosed as acute appendicitis (AA), retroperitoneal tumor or adnexal mass during several radiologic or endoscopic procedures and surgical operations. Familiarity to specific imaging findings especially taken from computed tomography (CT) helps physicians to diagnose this entity with high accuracy.

Although surgical resection is accepted as the treatment modality, approach either by laparoscopic or open surgery and the extension either by only appendectomy or right hemicolectomy are not defined clearly (1-3). However, it is generally accepted that avoidance to spill the mucinous material due to inadvertent handling should be the most important surgical rule for prevention of future recurrences (1,5).

In this retrospective study, our aim was to show clinical and imaging features of AM and to focus on its unique

radiologic appearances, which can help determine the surgical treatment and to prevent unplanned surgical interventions and misdiagnoses.

Material and Methods

Patients with a pathological diagnosis of AM over a 4-year period were included. The term "appendiceal mucocoele" includes histologic diagnosis of simple mucocoele or retention cyst, mucosal hyperplasia, mucinous cystadenoma, mucinous cystadenocarcinoma, and excluding of all cases that were diagnosed and discovered as pseudomyxoma peritonei (PP).

Parameters analysed were age, gender, and clinical presentation, complementary imaging studies, preoperative diagnosis, surgical findings and techniques, pathological diagnosis, length of hospital stay, postoperative course and follow-up.

Results

The relevant data with regard to demographic features, type of admission, pre- and postoperative pathological diagnosis, diameter and type of surgery was given in *Table 1*. The study group included 14 patients with a mean age of 51 years (range from 17 to 82 years). Eight of them (57%) were women. The clinicopathologic features of all cases were summarized in *Table 2*. For imaging purposes, ultrasonography and CT were performed in 3 and 14 patients, respectively. A cystic mass in the right iliac fossa suggesting AM was diagnosed in all patients in whom ultrasonography was performed. Use of CT resulted in diagnosis of AM in seven patients (50%) with imaging features of a long tubular cystic mass originating from the cecum (*Fig. 1, 2*). Calcification in the wall of the appendix (*Fig. 3*) was detected in two patients (14%). There

Table 1. Demographic and clinical features of the patients

No	Age/sex	Admission	Preoperative diagnosis	Pathology	Associated pathology	Diameter (mm)	Surgery
1	65/F	Elective	Retroperitoneal mass	Cystadenoma	None	40	LA
2	58/F	Elective	Adnexal mass	Cystadenoma	None	70	OA
3	30/M	Emergent	AA	Mucocoele	AA	15	OA
4	73/F	Elective	Colon adenocarcinoma	Cystadenoma	CA	20	RH
5	56/M	Emergent	AA	Mucocoele	AA	20	OA
6	17/M	Emergent	AM	Mucocoele	None	12	LA
7	47/F	Emergent	AA	Mucocoele	AA	12	OA
8	39/M	Elective	AM	Mucocoele	None	25	OA
9	46/O	Elective	AM	Mucocoele	None	60	OA
10	39/M	Emergent	AA	Mucocoele	AA	20	OA
11	82/M	Emergent	AM	Mucocoele	None	19	OA
12	50/F	Emergent	AM	Mucocoele	None	30	OA
13	63/F	Emergent	AM	Mucocoele	None	35	OA
14	54/F	Elective	AM	Mucocoele	None	27	OA

AA: acute appendicitis; AM: appendiceal mucocoele; CA: colonic adenocarcinoma; LA: laparoscopic appendectomy; OA: open appendectomy; RH: right hemicolectomy

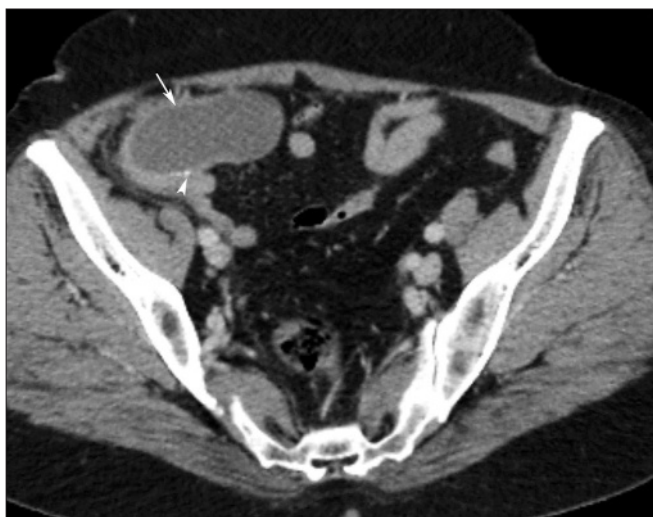


Figure 1. Contrast-enhanced (intravenous) axial CT scan of lower abdomen shows long tubular cystic mass with low-density content (arrow) with mural calcifications (arrowhead) on the right lower quadrant

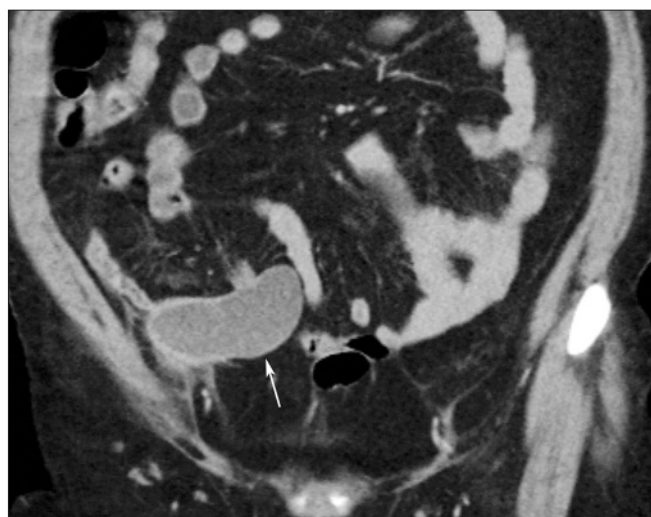


Figure 2. Contrast-enhanced (intravenous) coronal multiplanar reconstruction CT image shows a dilated cystic mass (arrow) in the expected region of the appendix



Figure 3. Contrast-enhanced (intravenous and positive oral) coronal multiplanar reconstruction CT image reveals mural calcifications (arrows) at the wall of AM

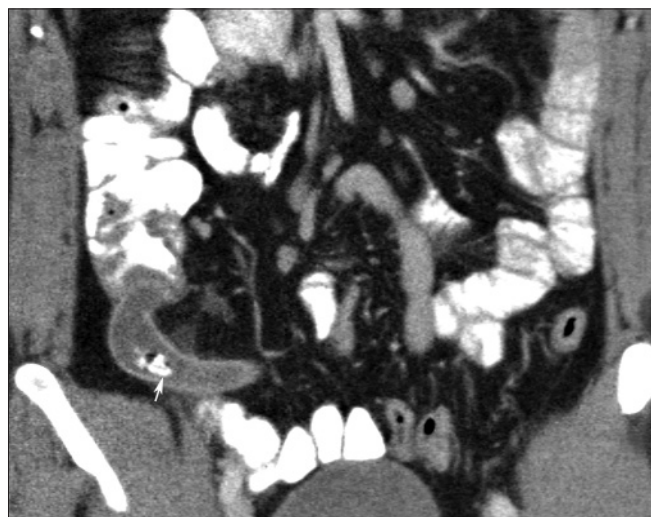


Figure 4. Contrast-enhanced (intravenous and positive oral) coronal multiplanar reconstruction CT image shows an inflamed AM containing appendicoliths (arrow) in an AA case

were four diagnoses (29%) of AA. Presence of appendicoliths (Fig. 4) was detected only in one patient. Retroperitoneal cystic mass, an ovarian mass in the right iliac fossa and wall thickening of the cecum were the other CT diagnoses. In all three patients (21%), incidental AMs were diagnosed during right hemicolectomy for right-sided colonic adenocarcinoma, diagnostic laparoscopy for retroperitoneal cystic mass and diagnostic laparotomy for a right-sided adnexal mass.

Appendectomy was the most common type of surgical treatment for AM (Table 2). During the operations, there was no PP, perforation of AM or synchronous ovarian mass in any patient.

Histopathological analysis of the appendix revealed only

benign pathologies as mucocoele or cystadenoma (Table 2). In all cases, mucoid material within the appendix was detected. Presence of AA and colon carcinoma were confirmed afterwards histologically in 4 (29%) and one (7%) patients, respectively. The mean diameter of AM was 29 mm. However, it was 43 and 25 mm for benign cystadenoma and mucocoele, respectively. The mean diameter of AM associated with AA was measured as 16.8 mm.

The mean length of stay was 2.86 ± 2.6 days with a range of 1 to 11 days. Wound infection treated by conservative approach developed only in one patient (7%). There was no perioperative mortality associated with AM. Follow-up data could be carried out in 7 out of 14 cases (50%), with a mean

Table 2. Clinicopathologic features of appendiceal mucocoele

Parameter		Number (n(%))
Clinical manifestation	pain in the right iliac fossa	9 (64)
	feeling of fullness in the right iliac fossa	5 (36)
Preoperative diagnosis	AM	7 (50)
	AA	4 (29)
	retroperitoneal cystic mass	1 (7)
	adnexal mass	1 (7)
Mode of admission	colon adenocarcinoma	1 (7)
	elective	6 (43)
	emergency	8 (57)
Intraoperative diagnosis	AM	9 (64)
	AM + AA	4 (29)
	AM + colon adenocarcinoma	1 (7)
Surgical treatment	open appendectomy	11 (79)
	laparoscopic appendectomy	2 (14)
	right hemicolectomy	1 (7)
Pathology	mucocoele	11 (79)
	cystadenoma	3 (21)

AM: appendiceal mucocoele; AA: acute appendicitis

of 10.14 ± 5.4 months (range 3-21 months). The patient with right-sided colonic adenocarcinoma associated with AM was free of the disease at six months postoperatively.

Discussion

Mucocoeles which were defined as cystic mucinous neoplasms of the appendix account for the majority of appendiceal tumors detected at imaging. The term mucocoele is simply defined as a macroscopic description of an appendix that is grossly distended by mucus. It lacks a specific pathologic diagnosis and can be caused by a variety of non-neoplastic, benign neoplastic and malignant conditions. Therefore, the term "appendiceal mucocoele" was used for all histological types of mucinous tumors of the appendix throughout the paper. It was described first by Rokitsansky in 1842 as an obstructive dilatation of the appendiceal lumen caused by intraluminal accumulation of mucoid material. It is a rare entity so that its incidence has been reported as up to 0.7% of all appendectomy specimens (1,6,7). Besides the case reports, there have been very limited numbers of case series with regard to AM (1,4,8-11). Mucocoele or mucinous tumors of the appendix are generally evaluated in four different histological groups as simple mucocoele or retention cysts, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma (8,11,12). However, there are some questions about its classification. The term "low-grade appendiceal mucinous neoplasm" has been offered to be used instead of all benign appendiceal mucinous tumors including simple mucocoele or retention cysts, mucosal hyperplasia, mucinous cystadenoma (13).

According to the sub-groups analysis of AM, it can be said that most of the cases are benign, and simple mucocoele and cystadenoma are the most commonly seen types among

the benign group (1,4,8,11). However, in Stocchi' study, which has the largest number of case series, the analysis was performed on the basis of simple mucocoele, cystadenoma and cystadenocarcinoma (1). In the absence of a clear appendiceal neoplasm as cystadenoma or cystadenocarcinoma, simple mucocoele is systematically diagnosed by pathologists (8). Therefore, all mucocoeles presented in this study were classified into three groups as simple and hyperplastic mucocoele, cystadenoma and cystadenocarcinoma revealing mucocoele in 79% and benign cystadenoma in 21%. There was no cystadenocarcinoma in our patients.

In the literature, there are some discrepancies with regard to gender and age distribution of these tumors. Although female or male predominance has been reported by some case series, others have showed similar incidences in men and women (4,8,9,11). There was a slight female predominance detected in the present study. Fifth and sixth decades of life have been regarded as the most predominating age group in accordance with ours (4,8,10). However, mucocoele may be diagnosed at any age ranging from 17 to 82 years as in this study. Therefore, AM should be considered in the differential diagnosis of an acute pain localized in the right lower quadrant in a patient, especially older than 30 years.

According to most of the published case series, acute abdominal pain is the most common presenting symptom (4, 7,8). In more than half of the patients (64%), abdominal pain, either acute (57%) or chronic, was detected as the main symptom. An association of AA or synchronous colorectal tumors with AM is another common finding which has been seen in up to 20% of the cases. Although presence of both AM and AA were seen in 29 % of patients, there was only one colonic adenocarcinoma, with an incidence of 7%. Nevertheless, it is recommended to perform colonoscopy and detailed systematic screening, either pre- or postoperatively, because of the possible associations with colorectal, breast, kidney, prostate, urinary bladder and gynecologic tumors which can be seen in one third of the patients (1,4,8,10).

It is thought that preoperative diagnosis can be an important factor for determination of surgical approach and its extension (14,15). With the use of preoperative imaging techniques, it is possible to diagnose AM preoperatively (8). According to the published series, the rate of a correct preoperative diagnosis has been achieved in up to 30% of cases (1,8). However, half of the cases in this study could be diagnosed as AM preoperatively. Nevertheless, it should be kept in mind that AM is usually discovered incidentally during radiologic evaluation of right lower quadrant pain or mass, or at laparotomy or laparoscopy performed for a pre-operative diagnosis of AA (8).

Cross-sectional imaging as CT, ultrasonography or magnetic resonance imaging is useful for evaluating the full extent of the lesion, considering the overall size and morphologic features. However, CT is accepted as the ideal imaging technique for evaluating AM because of its demonstrative features, including cystic well-circumscribed masses with low attenuation (7,12). However, differential diagnosis between sub-groups is usually not possible. Curvilinear mural calcifications

and enhancing nodules in the wall may be regarded as diagnostic clues for cystadenoma or cystadenocarcinoma (6,8). Stromal invasion besides the wall of the mucocoele, soft-tissue thickening and irregularity of the mucocoele wall and surrounding fat may predict the malignant or inflammatory potential of these tumors (6). Therefore, we preferred to use CT to evaluate acute abdominal pain or mass in all patients, although a cystic mass located in the right iliac fossa was diagnosed by ultrasonography in three of our patients. An important advantage of CT is that the anatomic relationship between the elongated cystic mass and the cecum is usually the most apparent at CT. Another advantage is detection of mural calcifications, which were detected in two patients (14%) (6). Cystic dilatation of the appendix, mural calcification of the wall, luminal diameter larger than 15 mm, and lack of appendicolith are regarded as the important imaging findings favoring AM (14).

The diameter of mucocoeles resulting from any type of non-neoplastic occlusions such as simple mucocoele, retention cyst and mucosal hyperplasia generally tends to be less than 2 cm, unless there is presence of cystadenoma or adenocarcinoma, in which the tumors are usually larger than 6 cm (6-8). It was shown that there was a statistical difference in diameter between simple mucocoele (mean diameter of 4.1 cm) and cystadenoma (mean and minimum diameters of 8.1 cm and 2 cm, respectively) in the study of Stocchi. However, any statistical difference in diameter between benign and malignant lesions was not demonstrated (7). Although the mean diameter of 25 mm for mucocoele and 43 mm for cystadenoma in this study were lower than the other reported values, cystadenomas also tended to be larger than mucocoele.

For differentiation of AM from AA or detection of AM and AA in the same patient, diameter of mucocoele can be regarded as an important landmark. After exclusion of the case with appendicolith, the mean diameter of the cases with AM and AA was calculated at 15.7 mm (not shown in the text). Therefore, in the absence of an appendicolith, the diameter should not be larger than 15 mm for a diagnosis of AA. As a practical proposal, it can be also recommended to operate all mucocoeles measuring at least 15 to 20 mm to eliminate indeterminate risk of progression to malignancy (7).

The treatment option of AM is surgery. Preoperative or intraoperative perforation of the mucocoele is a potentially important factor for future recurrences of PP (8). For that reason, attention should be paid to keep a mucocoele intact during operation and to avoid dissemination of mucoid material in the peritoneum (7). It was shown that future recurrences and progression of the disease could be prevented by avoiding rupture of the mucocoele during the operation (13). Although some surgeons prefer to use open surgery for these reasons, the optimum approach should be chosen according to the experience of the surgical team in laparoscopy (3,7). Careful use of laparoscopic graspers and absolute use of endobag during extraction are important maneuvers during laparoscopy (7-9). In considering the extension of the surgical treatment, the base of the appendix should be carefully assessed during the operation. Presence of positive margins and

difficulty to close the appendiceal stump necessitate more extensive surgery like cecectomy (7). The choice of right hemicolectomy should be selected in the presence of luminal diameter of the appendix larger than 2 cm, extension beyond the appendix, lymph node metastasis, PP and cystadenocarcinoma (4,8).

The survival for benign subgroups of AM has been shown as almost 90% for a 10-year period (5,8). Although good survival rates have also been reported for cystadenocarcinoma without peritoneal or adjacent organ involvement, presence of PP causes a significant decrease in overall survival rates. Follow-up is generally recommended for all cases with AM to detect the development of recurrences or PP, which may be seen even after benign pathology (8).

Although AM may be regarded as a rare disease with an incidence of up to 0.7% of all appendectomies, it should be kept in mind that the cumulative number of new cases of AM can reach high values because of the frequency of appendicitis as a general surgical problem (1,6,7). In addition, progression to PP in 10-15% of cases, the risk of malignancy and peritoneal spillage during surgery remind that AM is not a rare disease and has potentially dangerous complications (8).

In this study, there were some limitations including its retrospective design, lack of follow-up in half of the patients and the presence of only benign cases. However, it may be one of the largest series of AM considering the number of cases.

Conclusion

Despite the common use of imaging studies, preoperative diagnosis of AM is still not possible in most of the cases. CT can be regarded as the most accurate imaging test for diagnosis of AM and exclusion of other pathologies. AM should be sought in cases with a diagnosis of AA with a diameter of 2 cm taken during imaging studies, and considered in differential diagnosis of adnexal or retroperitoneal cystic masses. Detailed systematic screening is important because of the significant association with synchronous tumors. During surgical treatment which is tailored according to imaging and intraoperative findings, precautionary measures to avoid intraperitoneal rupture and dissemination should be taken.

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