

## Image Quiz for Surgeons

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# Preoperatively Diagnosed Mucocele of the Appendix

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### Rezumat

#### *Diagnosticul preoperator într-un caz de mucocele apendicular*

Mucocelele apendiculare sunt o nozologie rară, caracterizată prin distinderea lumenului apendicelui datorită acumulării conținutului mucinos și este rareori diagnosticat preoperator. Mucocelele netratate pot erupe producând pseudomixom peritoneal, o complicație potențial fatală. Modalitatea tratamentului chirurgical depinde de dimensiunile și tipul histologic al mucoceleului. Pentru mucocelele simple sau chistadenom se aplică apendicectomia. Pentru chistadenocarcinom se recomandă hemicolonectomia dreaptă. Prezentăm cazul unei paciente asimptomatice de 37 ani cu mucocele, care a fost primar depistat la examenul ecografic de rutină și tomografia computerizată preoperatorie. La intervenție a fost depistat un apendice vermicular mărit în dimensiuni 84 x 40 mm. Histologic a fost stabilit diagnosticul de mucocele simplu.

**Cuvinte cheie:** mucocele, apendice vermicular, diagnostic

### Abstract

Mucocele of the appendix is an infrequent entity, characterized by distension of the lumen due to accumulation of mucoid substance and is rarely diagnosed preoperatively. If untreated,

mucocele may rupture producing a potentially fatal entity known as pseudomyxoma peritonei. The type of surgical treatment is related to the dimensions and the histology of the mucocele. Appendectomy is used for simple mucocele or for cystadenoma. Right hemi-colectomy is recommended for cystadenocarcinoma. In this paper, we report a case of an asymptomatic 37-year-old woman in whom mucocele was found on a routine ultrasound examination and preoperative computed tomography scan. Surgery revealed a big appendix measuring 84 mm in length and 40 mm in diameter. The final pathologic diagnosis was simple mucocele.

**Key words:** mucocele, appendix, diagnosis

### Introduction

Appendiceal mucocele (AM) is a rare entity that can present with a variety of clinical symptoms or occur as an incidental surgical finding. The incidence is 0.2%-0.4% of all appendectomized specimens and 8% of appendiceal tumors (1-5). AM is a progressive dilatation of the appendix as a result of intra-luminal accumulation of the mucoid substance (5,6). It may be a benign or malignant process. Preoperative diagnosis is difficult, but is essential for the best choice of surgical approach to prevent peritoneal dissemination and perform the appropriate surgery (5,7). We describe a case of a young woman who was diagnosed with AM preoperatively.

### Case report

*A 37-year-old female patient was referred to our department*

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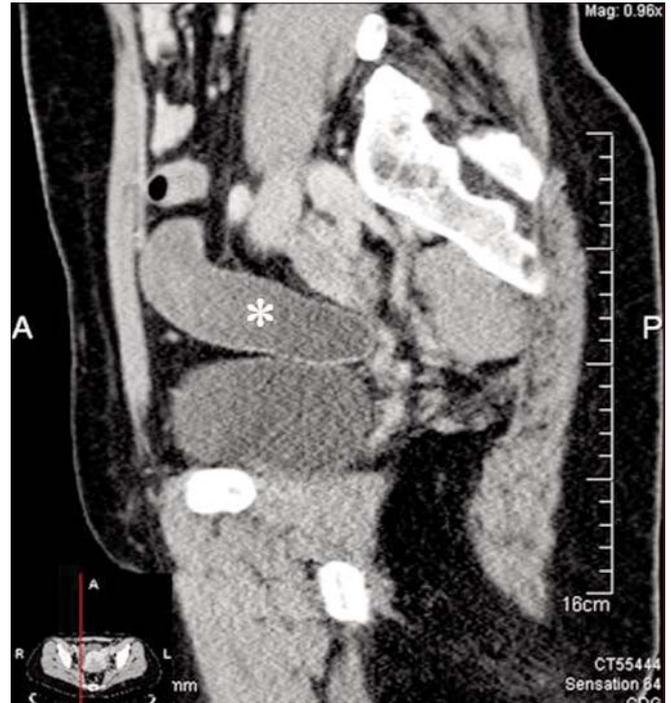
after a cystic mass was found incidentally during a routine ultrasonography examination performed according to a gynecologist's indication. Patient's past medical history was unremarkable. At admission she presented no complaints, physical examination didn't reveal any pathological changes. All the routine tests were within normal range. CEA concentration was 20.9 ng/ml (normal range 0.20-4.90), CA 19-9 – 6.21 u/ml (normal range 2.50-18.7). USG revealed a well-defined cystic lesion with heterogenous content 78 x 33 mm with thick walls up to 1.7 mm in the right iliac fossa (Fig. 1). Abdominal CT scan revealed a tubular mass with heterogenous content (0-50 HU) and mural calcifications 80 x 35 mm extending from the cecum to the right ovary (Fig. 2). A preoperative diagnosis of appendiceal mucocele was established and the patient was scheduled for surgery. A significantly enlarged vermiform appendix 84 x 40 mm, with narrow base was found on laparotomy (Fig. 3). The wall of the appendix was intact, without signs of content leakage. Peritoneal cavity was inspected thoroughly and no signs of peritoneal dissemination were found. Appendectomy was performed. After surgery gross examination of the resected specimen showed a distended appendix containing dense mucin (Fig. 4). Histological examination revealed a simple mucocele with inclusions of calcinate crystals in the appendiceal wall (Fig. 5). The postoperative course was uneventful. At 3 month follow up no signs of peritoneal involvement were found and the levels of CEA and CA 19-9 were normal.

## Discussion

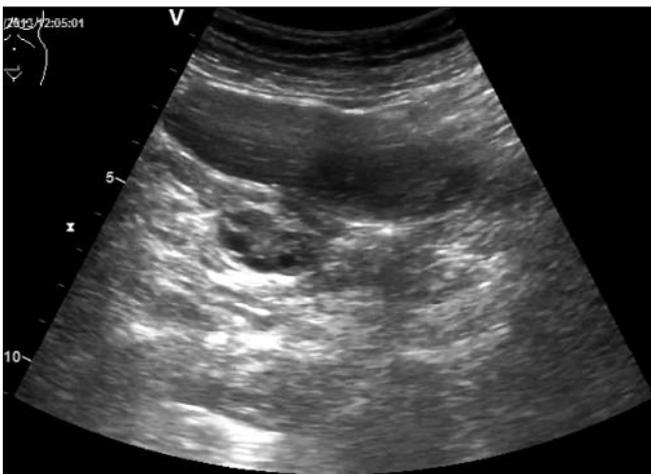
The mucocele of the appendix was first described in 1842 by Rokitsansky (8). The term "mucocele of the appendix" includes the histological diagnosis of simple mucocele or retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma, excluding all cases that were initially discovered as pseudomyxoma peritonei (9). Notwithstanding, some authors have recently questioned this classification and

terminology due to uncertain behavior and classified appendiceal mucinous neoplasms as either low-grade mucinous neoplasms or mucinous adenocarcinoma based on architectural and cytologic features (10).

Referring to gender distribution, there are discrepancies between different reports. Some studies describe female predominance (2,11), others show a similar incidence in men and women (12,13) and still others, show a higher frequency in men (1). In age distribution the incidence is predominating in the 5th and 6th decades of life, although



**Figure 2.** CT showing the enlarged appendix (asterisk) filled with nonhomogenous content (0-50HU)



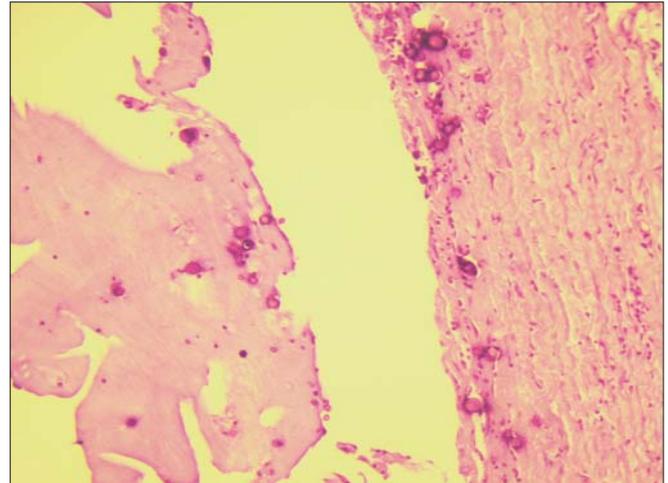
**Figure 1.** USG showing a cystic mass with hypoechoic content



**Figure 3.** Intraoperative view: significantly enlarged vermiform appendix with a narrow base



**Figure 4.** Resected specimen filled with dense mucin



**Figure 5.** Histologic image showing inclusions of calcinate crystals (HE $\times$ 40)

mucocele may be diagnosed at any age (12). Our patient's age was well below this range.

Mucocele of the appendix can be discovered incidentally in radiological or endoscopic tests or at laparotomy or laparoscopy performed for other reason. Some authors report that up to 50% of the cases are incidental findings, but most of the published series, revealed that acute abdominal pain is the main clinical manifestation of mucocele (14). Acute or chronic pain in right iliac fossa is the most frequent symptom, appearing sometimes as a mass at physical examination. Unusual manifestations are low gastrointestinal bleeding associated with intussusception of mucocele, intestinal obstruction, sepsis, fistula or genitourinary symptoms (11,12,15-19). In our case the patient was asymptomatic and AM was an incidental finding during routine USG.

Classically, preoperative diagnosis of mucocele has been considered as exceptional, but with the use of available diagnostic techniques that are more sensitive and specific than in the past, the number of cases diagnosed preoperatively has increased. Ultrasound is the firstline diagnostic modality for patients with acute abdominal pain or mass. Ultrasound shows cysts with variable echogenicity, depending of the composition of the mucus. Multiple echogenic layers along a dilated appendix produce the appearance of "onion-skin" circles and may be pathognomonic for mucocele (20). Appendix diameter 15 mm or more in USG examination has been determined as the threshold for AM diagnosis with a sensitivity of 83% and a specificity of 92% (5). In our case USG was very suggestive for AM. CT is also an effective diagnostic tool for AM. As in our case typical features of CT scan are cystic masses well circumscribed with low attenuation. Curvilinear mural calcifications are seen about 50% of the time and are very suggestive of mucocele (11,21,22). Barium enema may demonstrate a cecal filling defect or an ulceration (23). At colonoscopy, the appearance of the appendiceal orifice at the center of the mound has been labelled as the "volcano sign" moving in and out with respiration (4,24).

Though CEA and CA19-9 concentrations have not

been measured in many cases, these serum markers have occasionally been found to be elevated (25,26). High serum concentrations of CEA suggest a poor prognosis and possible recurrence of the disease. In the present case, the level of CEA was elevated, which returned to normal range after 3 months.

The worst complication is pseudomyxoma peritonei, characterized by peritoneal dissemination caused by iatrogenic or spontaneous rupture of the mucocele (7). An intact mucocele is considered to present no future risk for the patient, but once perforation occurs and epithelial cells escape into the peritoneal cavity, it becomes a potentially lethal entity (27).

Appendectomy is the treatment of choice. The tissues should be handled carefully during surgery in order to avoid rupture of the mucocele. Thus, conventional surgery is preferred rather than laparoscopic approaches for the treatment (4,7,26,28). This also allows the surgeon to explore the abdominal cavity, looking for the presence of mucoid fluid accumulations and mucin nodules in the omentum and peritoneum. After leakage, the accumulations of mucoid material are most commonly found in the right retrohepatic space, deep in the pelvis and in the cul-de-sac created in the left paracolic space above the junction of the sigmoid and descending colon, all anatomic sites difficult to explore at laparoscopy.

A simple and thorough evaluation of these patients with a new algorithm has been suggested by Dhage-Ivatury and Sugarbaker (27). Simple appendectomy is the choice of surgical treatment for patients with benign mucocele that has negative margins of resection without perforation. If the appendix shows histology of mucinous cystadenocarcinoma, perforated mucocele, with positive margins of resection, positive cytology and positive appendiceal lymph nodes, right colectomy/cytoreductive surgery (CRS)/heated intraperitoneal chemotherapy (HIIC) and early postoperative intraperitoneal chemotherapy (EPIC) should be performed. Long term follow-up is obligatory for these patients (27).

The outcome of simple mucocele, mucosal hyperplasia,

and mucinous cystadenoma after appendectomy is excellent, reaching 91% 10-year survival. Cystadenocarcinomas without peritoneal or adjacent organ involvement also show good outcome after surgical resection, but when they are at risk of progressing to pseudomyxoma peritonei, 5-year survival is 25%, with most deaths attributed to intestinal obstruction or renal failure (11).

In spite of an immediate good outcome of operation for mucocele, follow-up is recommended, because there are cases of recurrences as pseudomyxoma peritonei and instances of metachronic colonic neoplasms (12,29). Follow-up is recommended in all cases, even those with benign histology (simple mucocele, mucosal hyperplasia, and mucinous cystadenoma), because there are cases reported of development of pseudomyxoma peritonei with these histological types, although, obviously, less frequent (30).

In conclusion mucocele of the appendix is an infrequent pathology, manifested usually by nonspecific clinical signs. Ultrasound and CT are helpful in the preoperative diagnosis. The treatment of choice is appendectomy, although in mucinous cystoadenocarcinoma right hemicolectomy is needed. The pathologist must do a careful study, looking for inadvertent perforations by the surgeon that may radically change the outcome for the patient. Follow-up of all patients is justified, because of the risk of recurrence in the form of pseudomyxoma peritonei or colorectal neoplasms.

### Conflict of interests

Authors declare no conflicts of interest

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