

Appendiceal Mucocele in a Patient With Ulcerative Colitis – A Case Report

Mukokela crvuljka u pacijentice s ulceroznim kolitisom – prikaz slučaja

Kristina Samaržija^{1*}, Petar Milošević², Ivan Frketic²

¹ General Hospital Karlovac, Department of Radiology, Karlovac, Croatia

² General Hospital Karlovac, Department of Surgery, Karlovac, Croatia

Abstract. Aim: We present the rare case of a patient who was incidentally diagnosed with mucocele of the vermiform appendix by radiologic examination due to symptoms of ulcerative colitis (UC) and point out the importance of making this diagnosis before surgery. **Case report:** A 66-year-old woman was admitted for bloody diarrhea, nausea, diffuse abdominal pain and fever. Ultrasonography revealed an anechoic tubular lesion with dorsal enhancement in the right lower quadrant of the abdomen. Computed tomography showed a cystically dilated appendix with low-density intraluminal contents, without wall thickening or inflammatory changes in the surrounding area. Colonoscopy with biopsy revealed acute UC. After the symptoms had subsided, an appendectomy was performed with very careful manipulation to avoid leakage of fluid into the abdominal cavity. Histopathology revealed a dilated appendix filled with abundant mucus and lined with mucinous epithelium without mitosis or atypia, suggesting a simple mucocele. **Conclusion:** Appendiceal mucocele is a morphologic term for an enlarged, mucus-filled appendix, regardless of the underlying pathology, which may be non-neoplastic or neoplastic. It is a very rare disease and often goes unrecognized. The concomitant occurrence of appendiceal mucocele and UC is particularly rare and there is still no reliable evidence of a causal connection between the two. Resection is recommended for all mucoceles as there is a possibility of malignancy. Preoperative diagnosis is important to prevent spontaneous or iatrogenic rupture. If diagnostic imaging techniques reveal a cystic lesion in the right lower quadrant of the abdomen, an appendicocele should be considered in the differential diagnosis.

Keywords: appendix; mucinous neoplasms; mucocele; ulcerative colitis

Sažetak. Cilj: Prikazati rijedak slučaj pacijentice kojoj je dijagnosticirana mukokela crvuljka slučajno, radiološkim pregledima učinjenim zbog simptoma ulceroznog kolitisa, te ukazati na važnost postavljanja ove dijagnoze prije operacije. **Prikaz slučaja:** Pacijentica u dobi od 66 godina zaprimljena je zbog proljevastih stolica s nešto krvi i sluzi, mučnine, difuzne boli u truhu i povišene tjelesne temperature. Ultrazvučnim pregledom nađena je anehogena tubularna lezija s dorzalnim pojačanjem u donjem desnom kvadrantu abdomena. Kompjuterizirana tomografija pokazala je cistično dilatiran crvuljak s intraluminalnim sadržajem niskog denziteta, bez zadebljanja stijenke ili okolnih upalnih promjena. Kolonoskopijom s biopsijom dokazan je akutni ulcerozni kolitis. Nakon smirivanja simptoma ulceroznog kolitisa učinjena je apendektomija uz vrlo pažljivu manipulaciju i korištenje nepropusne vrećice da se izbjegne izlivanje tekućeg sadržaja u peritonealnu šupljinu. Histopatološki radilo se o dilatiranom crvuljku ispunjenom obilnom sluzi, obloženom mucinoznim epitelom bez mitoze ili atipije, te je postavljena dijagnoza jednostavne mukokele. **Zaključak:** Mukokela crvuljka morfološki je termin za dilatirani crvuljak ispunjen sluzi, neovisno o patologiji u pozadini koja može biti neneoplastične ili neoplastične prirode. To je vrlo rijetka bolest i često ostaje neprepoznata. Osobito je rijetka istodobna pojava mukokele crvuljka i ulceroznog kolitisa i još uvijek nema pouzdanih dokaza o uzročnoj povezanosti između njih. Kod svih mukokela preporučuje se resekcija zbog mogućnosti maligniteta. Predoperacijska dijagnoza važna je zbog sprječavanja spontane ili jatrogene rupture. Kad se slikovnim dijagnostičkim metodama otkrije cistična lezija u desnom donjem kvadrantu abdomena, u diferencijalnoj dijagnozi treba razmotriti mukokelu crvuljka.

Ključne riječi: crvuljak; mucinozne neoplazme; mukokela; ulcerozni kolitis

***Corresponding author:**

Kristina Samaržija, MD
General Hospital Karlovac, Department of Radiology
Andrije Štampara 3, 47000 Karlovac, Croatia
E-mail: kristinabaskot@yahoo.com

<http://hrcak.srce.hr/medicina>

INTRODUCTION

Appendiceal mucocele (AM) is a very rare condition that occurs in less than 1 % of surgically removed appendices^{1,2}. It was first described by Rokitansky in 1842 and later defined by Feren in 1976^{3,4}. AM is a broad morphologic term for a dilated appendix with abnormal intraluminal mucus accumulation, regardless of the underlying pathology^{1,3-6}. As patients are largely asymptomatic or present with a non-specific clinical picture, it is rarely diagnosed prior to surgery^{1,3,4,7}. In reviewing the literature, we found mainly case reports and only small case series with this entity. We present a case of AM diagnosed incidentally by radiologic examination due to symptoms of ulcerative colitis (UC) with the aim of raising awareness of this rare entity.

CASE REPORT

A 66-year-old woman came to our emergency department with a 10-day history of diarrhea with some mucus and blood (up to ten stools per day) along with nausea without vomiting. For the last two days she had diffuse abdominal pain with fever up to 39 °C. She was known to have UC for the past eight months and was in clinical remission with mesalazine therapy of 3 g/day in oral form and 1 g/day in rectal form. Ileocolonoscopy performed at the time of the initial diagnosis of UC showed a left-sided UC with inflammatory lesions located distally to the splenic flexure. Physical examination revealed diffuse tenderness to palpation of the abdomen, more on the right side, with normal peristalsis, without signs of peritonitis. Laboratory data indicated a slightly elevated white blood cell count ($11.2 \times 10^9/L$; reference values: $3.4 - 9.7 \times 10^9/L$), elevated C-reactive protein (288.8 mg/L; reference value <5 mg/L), elevated procalcitonin ($6.13 \mu\text{g/L}$; reference value $<0.50 \mu\text{g/L}$) and elevated fecal calprotectin ($>850 \mu\text{g/g}$; reference value $<80 \mu\text{g/g}$). The other laboratory values were normal. The abdominal X-ray showed no significant changes. Ultrasonography (US) revealed an anechoic, encapsulated tubular lesion in the right lower quadrant of the abdomen measuring 40 mm in length and 13 mm in diameter in continuity with the ap-

pendix (Figure 1). The acoustic enhancement typical of a cystic mass was clearly visible, and the wall of the lesion, and the proximal part of the appendix were not thickened. The mass was suspicious for AM. A computed tomography (CT) scan of the abdomen and pelvis was performed, which revealed a cystically dilated distal portion of the appendix with intraluminal contents of low attenuation coefficients, without wall thickening or surrounding inflammatory changes (Figure 2). No intraperitoneal free fluid or extraluminal gas and no nodal enlargement were detected. CT confirmed the ultrasound findings and provided sufficient evidence for the diagnosis of AM. In addition, CT showed an absent haustral pattern in the descending and sigmoid colon with circular wall thickening and hyperemia, suggestive of inflammatory bowel disease. The patient was admitted to the gastroenterology department.

Appendiceal mucocele is a very rare disease that occurs in less than 1% of surgically removed appendices. Patients are often asymptomatic or have a nonspecific clinical presentation, so the diagnosis is often made incidentally by radiologic examination due to other unrelated disease or on specimens after appendectomy.



Figure 1. Ultrasonographic image – anechoic tubular lesion in continuity with the appendix

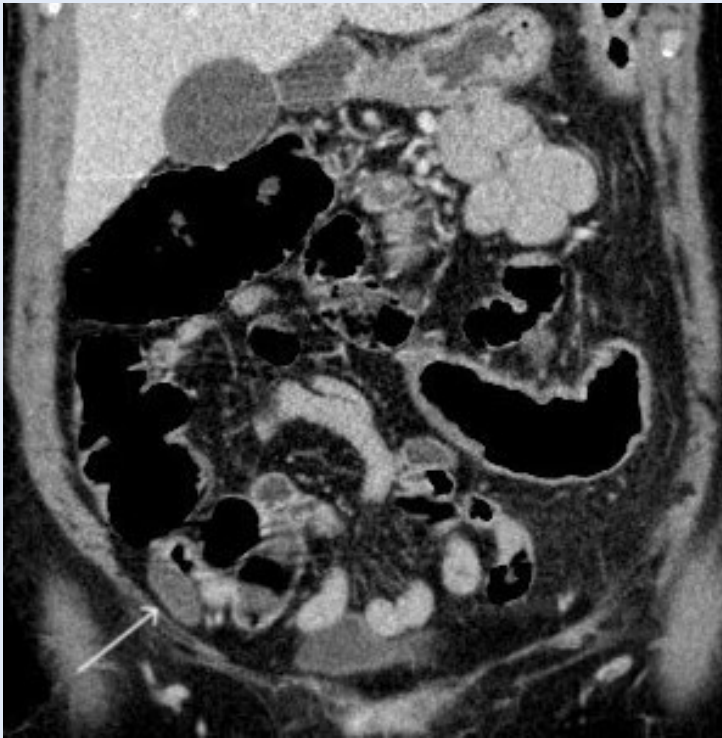


Figure 2. CT image – dilated appendix with low-attenuation intraluminal content, without wall thickening or surrounding inflammatory changes



Figure 3. Macroscopic finding of mucocele at operation.

Rectosigmoidoscopy without preparation revealed inflammatory changes with contact bleeding consistent with acute rectosigmoiditis, and histopathologic findings after biopsies were compatible with the changes suggestive of UC. The results of the microbiological examination of the stool and blood culture were negative. According to clinical presentation and basic laboratory tests, the patient met the Truelove and Witts criteria for acute severe ulcerative colitis^{8,9}. Treatment was started with systemic steroids administered intravenously (methylprednisolone sodium succinate 80 mg/day) for seven days and then switched to oral prednisone 60 mg/day. Due to fever with a temperature of up to 39 °C, a broad-spectrum antibiotic (ciprofloxacin 800 mg/day and metronidazole 1500 mg/day) were administered intravenously to reduce septic complications. The patient was also given oral mesalazine 3 g daily as well as fluid and electrolyte replacement. Since she tolerated the oral intake well, the regular colitic diet was continued. The patient responded well to the therapy by reducing the number of stools without visible rectal bleeding and gradual normalization of systemic inflammatory parameters. She was discharged from the hospital twenty days after admission with only elevated fecal calprotectin (>850 µg/g). On discharge, it was recommended to continue oral prednisone therapy at a dose of 40 mg per day with a gradual reduction of the dose by 5 mg per week until it is discontinued. Maintenance therapy with oral mesalazine 3 g daily and local application of mesalazine in the form of an enema 4 g/60 ml in the evening was also recommended. A follow-up plan was made using symptom-based and biomarker-based monitoring strategy, including the measurement of fecal calprotectin levels, which well reflect the severity of inflammation in the colon¹⁰. Colonoscopy was scheduled within 6 months to assess the healing of the mucosal lesions. The diagnosis of AM was explained to the patient along with the treatment recommendation.

One month later, when she was symptom-free, she was admitted to the Department of Abdominal Surgery for surgery. At surgery, a cystic lesion of the distal part of the appendix was found, which macroscopically looked like a simple mu-

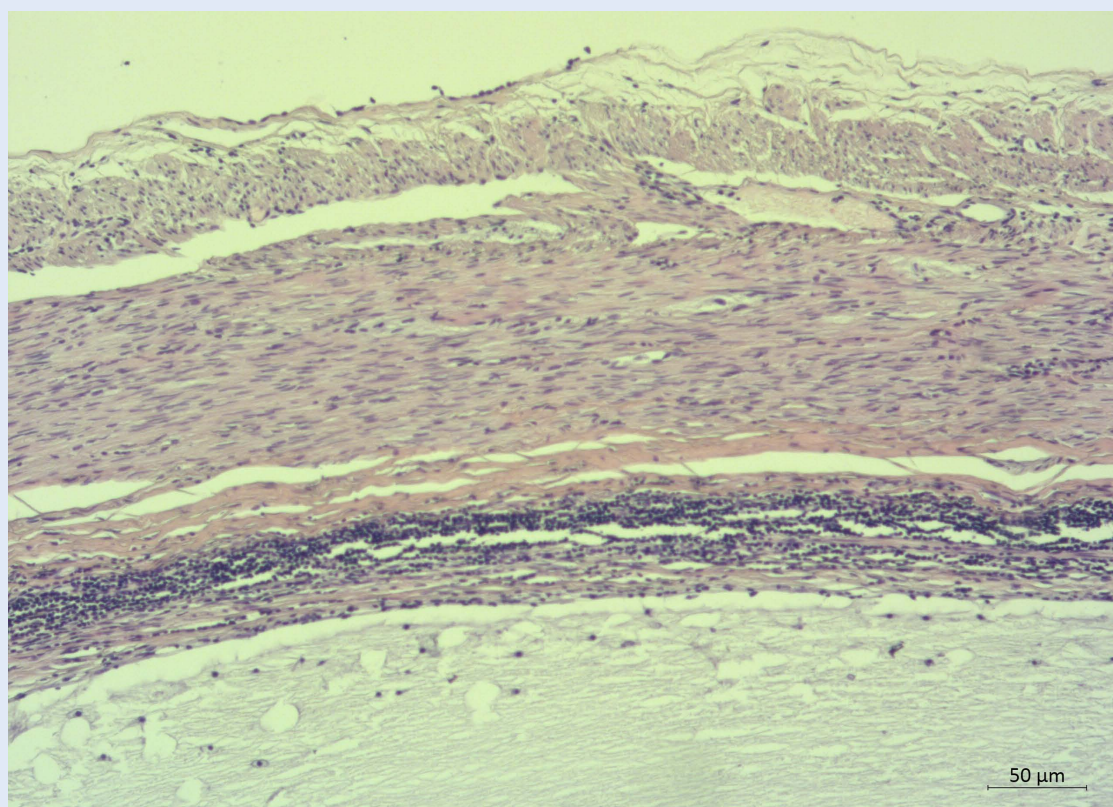


Figure 4. Histological image of the appendiceal specimen – lumen filled with mucus lined by epithelium with benign histomorphological characteristics.

cocele, with no pathologic process at the base of the appendix or pathologic content in the peritoneal cavity (Figure 3). Therefore, only an appendectomy was performed, handling the appendix very carefully and using an impermeable bag to collect the specimen to avoid fluid leakage. The histopathology report described a dilated appendix with a diameter of 13 mm, lined with single and multi-row, cylindrical, mucinous epithelium without mitosis or atypia (Figure 4). The lumen of the appendix was filled with abundant mucus, the walls were thinned in places but without signs of rupture, and AM was diagnosed. There were no postoperative complications, and the patient was discharged according to the algorithm without a recommendation for long-term follow-up.

On maintenance therapy with oral and rectal mesalazine, the patient was in clinical remission, but the fecal calprotectin level was high ($> 850 \mu\text{g/g}$), indicating active inflammation. Colonoscopy performed 3.5 months after discharge from gastroenterology department revealed inflammatory

lesions that extended to the hepatic flexure of the colon, in terms of progression from initial left-sided to extensive colitis. Six months after discharge, the patient experienced a relapse with diarrhea (up to five stools per day), with minimal rectal bleeding, without fever, which was resolved in outpatient treatment by adding oral corticosteroids to the therapy. After remission was achieved, azathioprine was introduced in maintenance therapy along with mesalazine. One year later, the patient is in clinical remission, but fecal calprotectin is elevated ($342 \mu\text{g/g}$) and colonoscopic reevaluation did not show healing of the mucosal lesions. Due to the progression from initial left-sided to extensive colitis and the severe and complicated course of the disease with frequent exacerbations, and continuously high levels of fecal calprotectin without signs of endoscopic remission, the patient was stratified into a high-risk category^{9,11}. She was directed to a tertiary referral center for further follow-up, where patients with the most complicated disease are treated.

DISCUSSION

AM is a common term for a heterogeneous group of lesions characterized by an enlarged, mucus-filled appendix but with varying underlying pathology, ranging from non-neoplastic to neoplastic. Different classifications and terminology were used in the older literature, and most commonly AM was categorized into four types: simple, hyperplastic, cystadenoma, and mucinous cystadenocarcinoma. In 2012, the Peritoneal Surface Oncology Group International (PSOGI) developed a consensus classification, published in the *American Journal of Surgical Pathology* in 2016, which has helped to eliminate confusion in diagnostic terminology^{12,13}. According to this consensus, mucinous lesions of the appendix are basically categorized into non-neoplastic and neoplastic lesions. The category of non-neoplastic mucinous lesions of the appendix includes simple or obstructive mucoceles characterized by degenerative epithelial changes due to obstruction and stretching, without signs of mucosal hyperplasia or neoplasia. Thus, this variant results from chronic obstruction of the appendiceal lumen by a process other than neoplasia leading to retention of mucus behind the obstruction. The category of neoplastic mucinous lesions of the appendix results from excessive mucus production by mucinous tumors of the appendix, which include serrated polyps with or without dysplasia, mucinous neoplasms, and mucinous adenocarcinomas. Mucinous appendiceal neoplasms are dysplastic, mucinous tumors that are confined to the mucosa and have no infiltrative or destructive pattern. These tumors may be low-grade or high-grade depending on their cytologic features. Mucinous adenocarcinomas of the appendix are mucinous tumors with infiltrative invasion that can be classified as well, moderately or poorly differentiated. Mucinous appendiceal neoplasms and mucinous adenocarcinomas can perforate and spread into the peritoneal cavity, resulting in a pseudomyxoma peritonei characterized by the accumulation of mucinous contents and peritoneal implants. In this study, we present a patient with a cystic lesion of the appendix without a macroscopically visible neoplastic process. Histopathology revealed a dilated, epithelium-lined appendix with-

out mitosis or atypia, the lumen of which was filled with mucus, allowing the lesion to be categorized as a non-neoplastic lesion, i.e. simple mucocele.

The majority of AM is asymptomatic and is discovered incidentally during surgery or on imaging due to other unrelated conditions^{1,4,7,14}. When symptoms are present, they are non-specific, most commonly pain or a palpable mass in the right lower quadrant that mimics appendicitis or adnexal disease in women^{3,4}. Other clinical signs include weight loss, nausea, vomiting, changes in bowel habits and unexplained anemia^{3,4,15}. Due to its rarity and atypical clinical features, preoperative diagnosis of AM is difficult even with imaging studies⁴.

In a patient with abdominal symptoms, the US examination is usually the diagnostic method of first choice. On US examination, the AM presents as a cystic mass in the expected area of the appendix. It is usually a clear tubular cystic structure with dorsal enhancement, but other sonographic patterns have also been published^{5,16}. Some authors describe a mass composed of concentric layers with echogenic contents that looks like an onion in cross-section^{6,16-18}. This sonographic "onion skin" sign was first described by Degani in 2002 and is considered a specific sonographic marker for AM^{16,18}. Despite its solid consistency, the acoustic enhancement typical of a cyst is visible behind this structure¹⁶. The exact etiology of layering within an AM is not clear. There are various theories to explain this phenomenon, such as repeated sedimentation of the mucinous substance, fluctuation in the secretion of mucin and temporal variations in the intraluminal pressure leading to different layer densities^{6,16}. Although the sonographic "onion skin" sign was originally described for mucinous lesions of the appendix, it can also be found in other mucinous cysts, such as ovarian cysts, further complicating the diagnosis¹⁹.

CT is considered the most accurate method for diagnosing AM. The typical CT finding is a well-encapsulated cystic mass in the right lower quadrant of the abdomen with a thin wall and intraluminal contents of low attenuation coefficients^{1,20}. The degree of attenuation depends on the amount of mucin in the fluid contents⁵. There may also be curved or punctate calcifications in

the appendiceal wall^{15, 20}. The difference from appendicitis is the thin wall of the appendix, the absence of periappendiceal inflammatory changes and a larger diameter¹. When it spreads to the pelvis in women, it must be differentiated from ovarian cystic neoplasms, tubo-ovarian abscesses and hydrosalpinx²⁰.

In our case, ultrasound showed a thin-walled cystic lesion with dorsal enhancement that was in continuity with the appendix and suspicious for a mucocele, which was confirmed by CT scan. Although the reported incidence of correct pre-operative diagnosis of AM is very low (15%–29% in chronic disease and only 7.5% in acute disease)⁴, we believe that US and CT scan are usually sufficient diagnostic tools to resolve this diagnostic challenge.

Not many cases of AM in combination with UC have been described in the literature, raising the question of whether there is a causal relationship between the two. According to some authors, inflammation of the appendix or appendiceal orifice in UC may block excretion from the cavity, which leads to mucus retention with the development of a mucocele^{15, 21–23}. Authors reported a high prevalence of mucosal inflammation in the appendixes of patients with UC^{24–26}. Heuthorst et al. assessed histological features of 140 appendix specimens from patients with UC and found active appendiceal inflammation in almost 60% of all patients. Inflammation of the appendix was not related to disease activity in the colon or the extent of UC (more than 50% of patients in remission showed active histological disease in the appendix)²⁶. A systematic review by Park et al highlighted a wide range of prevalence of appendiceal orifice inflammation in patients with UC in endoscopic studies, varying from 8% to 75%²⁷. Some authors have reported discontinuous appendiceal involvement in patients with left-sided UC, with histologically normal mucosa of other parts of the colon and cecum, representing a “skip” lesion^{15, 21, 27–29}.

In addition to the inflammatory process, obstruction of the appendiceal orifice may also be caused by a neoplasm in a patient with UC. As there are more mucus-producing goblet cells in the epithelium of the appendix than in the epithelium of the colon, most epithelial tumors of

the appendix are mucinous and start as mucoceles³⁰. Patients with inflammatory bowel disease (IBD) are at increased risk of colorectal neoplasia, but it is unknown whether IBD is associated with appendiceal mucinous neoplasms (AMN). Orta et al. performed a retrospective case-control study of incidental appendiceal neoplasms in colectomy specimens of adults with and without IBD. Although there was no significant difference in prevalence of appendiceal cystadenomas between IBD patients and non-IBD controls, appendiceal cystadenomas were 15-fold more prevalent among IBD patients with synchronous colorectal neoplasia compared with controls.

If imaging studies show a cystic lesion in the right lower quadrant of the abdomen, AM should be considered in the differential diagnosis. Resection is recommended for all mucoceles because of the possibility of malignancy.

They concluded that IBD with synchronous colorectal dysplasia or cancer is a risk factor for development of appendiceal cystadenomas, implicating this tumor as a neoplastic complication of IBD³¹. Also, Matsushita et al reported higher prevalence of appendiceal cystadenoma in patients with UC, compared with patients with Crohn's disease²³. In 2023, Bonomi et al reviewed all published studies on adult patients with IBD, subjected to surgical resection, who were histologically confirmed to have AMN. They hypothesized that long-standing and/or aggressive appendiceal inflammation related to IBD should predispose patients to AMN. After considering the limitations arising from the limited number and quality of evidence (22 case reports and only one case-control study to that date), they conclude that it is reasonable to consider the possibility of a direct connection between IBD and AMN. They believe that patients with long-standing IBD should be routinely screened not only for colorectal cancer but also for AMN during gastro-enterologic follow-up³². The diagnosis of AM must be considered when a distended or bulged appendiceal orifice is observed during colonoscopy^{15, 21}.

Chronic inflammation is a well-recognized risk factor for the development of cancer in humans,

and long-standing chronic IBD is one of those risk factors³³. We believe it is reasonable to hypothesize that patients with IBD could be at increased risk for appendiceal mucinous neoplasms in the same way as for colorectal neoplasia.

In our patient, ileocolonoscopy at the stage of the initial diagnosis of UC did not show inflammation of the cecal mucosa or appendiceal orifice, and during this relapse no endoscopic assessment of the cecum was performed due to the risk of perforation. Therefore, we cannot suggest the mechanism of AM formation, and it is possible that the concomitant occurrence of AM and UC in this case is coincidental. Our report adds a new case to the rarely reported cases of simultaneous occurrence of UC and AM emphasizing the need for a thorough evaluation of the appendiceal orifice in patients with UC. Additional studies are needed to further clarify the hypothesized association between inflammatory bowel disease and AM.

If AM is detected on imaging studies, early surgical resection is recommended to prevent spontaneous rupture and rule out mucinous neoplasms³⁴. Despite extensive imaging studies, there are no reliable criteria for excluding an underlying malignant lesion, so histopathologic examination of the appendix specimen after surgery is required to make a definitive diagnosis^{35,36}.

In patients with a non-neoplastic mucosal lesion of the appendix, such as a simple mucocele, standard appendectomy is the method of choice without the need for further follow-up^{34,36}. Careful intraoperative management is extremely important to avoid iatrogenic rupture and leakage of mucus into the peritoneal cavity. For this reason, many surgeons prefer open surgery, as the risk of rupture is considered lower than with the laparoscopic approach^{34,36}. If a malignant mucocele is suspected and the base of the appendix is involved so that a clear margin cannot be achieved by stapling, a cecectomy, ileocectomy or right hemicolectomy should be performed^{37,38}. In case of ruptured neoplastic mucocele with disseminated peritoneal disease (clinical syndrome known as pseudomyxoma peritonei), additional surgical treatments such as cytoreductive surgery and heated intraperitoneal chemotherapy are required^{37,38}. After receiving the final pathologic re-

port, a multidisciplinary team should be involved in the further management of patients with malignant mucocele^{7,38}.

CONCLUSION

When imaging studies reveal a cystic lesion in the right lower quadrant of the abdomen, AM should be considered in the differential diagnosis. When monitoring inflammatory bowel disease, special attention should be paid to patients with unusual symptoms. Resection is recommended for all mucoceles due to the possibility of malignancy.

Conflicts of Interest: Authors declare no conflicts of interest.

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