

Otrzymano: 2007.07.19  
Zaakceptowano: 2007.11.30

## Mucocele of the appendix – case report

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### Summary

<b>Background:</b>	Mucocele of the appendix is a term describing a cystic dilatation of the appendiceal lumen caused by abnormal mucous accumulation. Mucocele of the appendix is a rare condition, found in about 0.2% - 0.6% of all appendectomies. Mucocele can appear at every age, usually after 50 years of age and more frequently in women. The preoperative diagnosis is correct only in 20% cases. Precise diagnosis is very important because the therapy is surgical and biopsy is not advised.
<b>Case report:</b>	We present a 75-year-old patient who was diagnosed because of abdominal pains. Abdominal sonography and computed tomography revealed well-circumscribed cystic lesion in the right lower quadrant of the abdomen. The lesion was surgically removed. Histopathologic evaluation demonstrated a benign mucocele of the appendix.
<b>Conclusions:</b>	In case of presence of a cystic lesion in the right lower quadrant of the abdomen in a patient without a history of appendectomy, appendiceal mucocele is one of the differential diagnoses.
<b>Key words:</b>	mucocele • appendix • ultrasound • computed tomography
<b>PDF file:</b>	<a href="http://www.polradiol.com/fulltxt.php?ICID=677396">http://www.polradiol.com/fulltxt.php?ICID=677396</a>

### Background

Mucocele of the appendix is a term describing a cystic dilatation of the appendiceal lumen caused by abnormal mucous accumulation [1]. It was first described by Rokitansky in 1842 [2]. Mucocele of the appendix is a rare condition, found in about 0.2% - 0.6% of all appendectomies [1, 3]. The clinical presentation is usually nonspecific and 23%-50% of mucoceles are diagnosed incidentally at surgery [2, 3]. About 23% of mucoceles of the appendix are asymptomatic [3]. The condition affects predominantly females (male to female ratio 1:4-1:7) above 50 years of age [1, 4, 5]. The preoperative diagnosis of mucocele of the appendix is correct in only 20% of cases [6]. Early and accurate diagnosis of the disease is very important because mucocele of the appendix can be complicated by the rupture during surgery with a development of pseudomyxoma peritonei or by malignant transformation [2, 7]. Imaging modalities that can reveal

a presence of the mucocele of the appendix are especially sonography and computed tomography [1, 3, 6, 7, 8, 9].

### Case report

A 75-year-old female came to the emergency because of acute onset of abdominal pain of a few hours' duration. The pain was located in the mesogastrium, radiated to the left subcostal region, and was accompanied by nausea and vomiting. Past medical history revealed recurrent mild abdominal pains in the right lower quadrant for the last two years, with no abnormality on sonography of the abdomen. Six years previously, the patient underwent laparoscopic cholecystectomy for cholelithiasis and then after three years lithotripsy of the left urethral calculus.

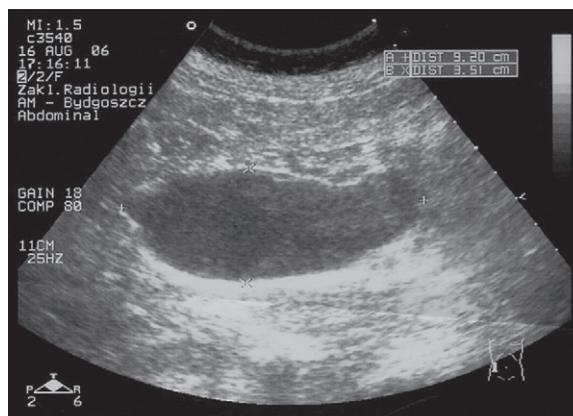
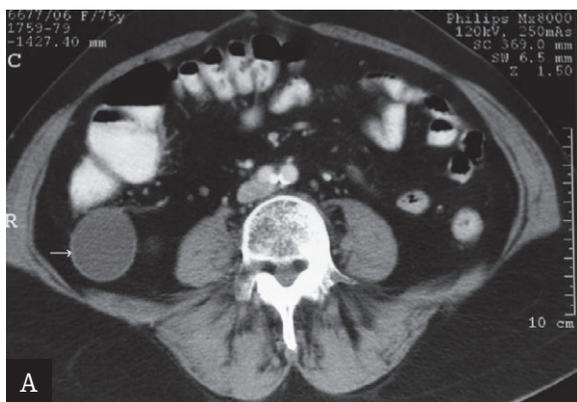
The body temperature was mildly elevated to 37.2°C. On physical examination, there was mild pain during

palpation of the periumbilical region. The blood pressure was 130/80 mmHg with regular pulse of 80/min. Laboratory blood tests revealed slight leukocytosis  $11.02 \times 10^3/\mu\text{l}$ . Plasma amylase level, C-reactive protein and carcino-embryonic antigen were normal. Urinalysis showed discrete proteinuria, leukocytes  $70/\mu\text{l}$ , erythrocytes  $25/\mu\text{l}$  and elevated activity of amylase 521 U/l (norm 5- 410 U/l). Abdominal plain film was normal except for few short fluid levels in the mesogastrium and left epigastrium, with no signs of ileus or perforation. Sonography of the abdomen showed an  $92 \times 35$  mm oval structure of low echogenicity beneath the lower pole of the right kidney, which was interpreted as possibly distended ileal loop. A small amount of free fluid was also noted in the hypogastrum. The pain subsided on the same day. The next sonographic examination after four days confirmed the presence of a lesion located between the lower pole of the right kidney and the iliac wing. It was a  $84 \times 35$  mm well-circumscribed, thin-walled, elongated hypoechoic mass, with slightly inhomogenous internal structure (fig. 1). Spiral computed tomography of the abdomen revealed a tubular low-attenuation, well-encapsulated, cystic lesion in the right hypogastrum, without any calcification or contrast enhancement (fig. 2 A, B). It coursed longitudinally and the lower part was adherent to an intestinal loop. The mass measured 97 mm in length, with a diameter of 36 mm. There were no signs of inflammation in the surrounding fat or thickening of the bowel wall. The definite diagnosis was not made, although a cystic lesion with high probability of association with gastrointestinal tract was suggested.

The only abnormality in laboratory tests was elevation of urine amylase (730 U/l).

The patient underwent gastroscopy that revealed no abnormalities. In colonoscopy, only a few diverticula were found in the sigmoid. Gynecological examination and sonography were also normal.

Abdominal sonography was repeated four months later. The lesion appeared similar except for local thickening of the wall with some echogenic irregular structures located peripherally in the lumen (fig. 3 A, B).



**Figure 1.** Initial ultrasonography of the right hypogastrum shows a hypoechoic oval structure with thin walls.

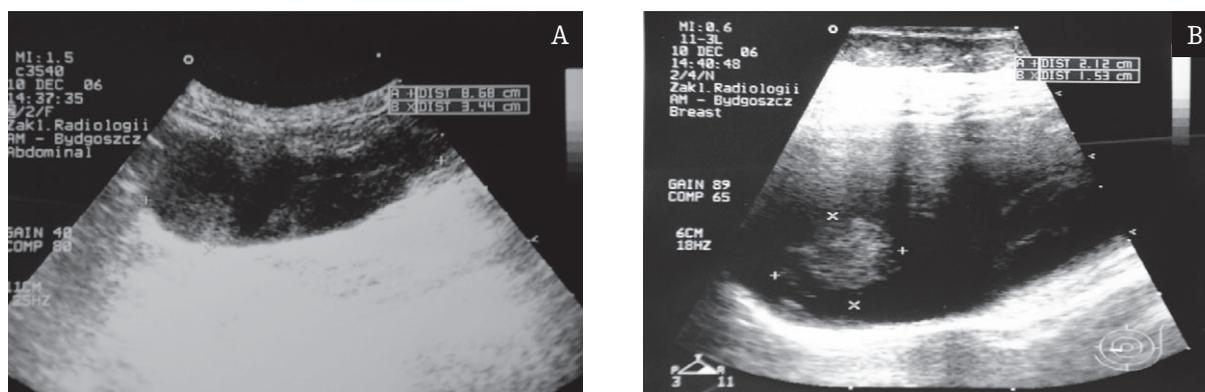
After colonic diverticulum, colonic tumor and ovarian cyst had been excluded, the surgery was performed. The cystic dilatation of the retrocecal appendix was found, without inflammatory periappendiceal reaction or adhesions. Appendectomy was performed. Macroscopic evaluation of the specimen showed a thin-walled 90 mm long appendix, with a lumen distended to 30 mm and filled with mucinous-gelatinous content. The mucosa was absent at an extensive part of the wall. Histopathologic evaluation revealed benign mucocele of the appendix (fig. 4 A, B). The postoperative course was unremarkable.

## Discussion

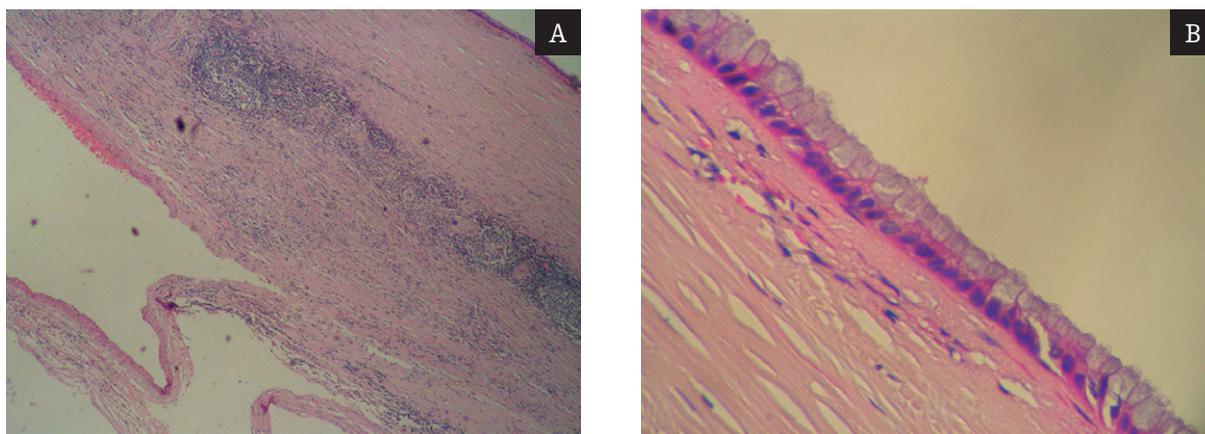
Mucocele of the appendix is caused by obstruction of the appendiceal lumen, which was confirmed by Berry, who produced mucoceles experimentally in rabbits by proximal ligation of the appendix [2]. The cause of the obstruction may be inflammatory stricture, carcinoid, carcinoma, villous adenoma, appendicolith, mucosal web, endometriosis or extrinsic compression. In most cases, as in our case, the cause of obstruction cannot be found [2]. As proposed in 1940 by Woodruff and McDonald, appendiceal mucocele can be classified into a benign and malignant type [2]. Higa and al. divided mucocele of the appendix into three groups: 1) Focal or diffuse mucosal hyperplasia, 2) Mucinous cystadenoma and 3) Mucinous cystadenocarcinoma [10].



**Figure 2.** Computed tomography with bowel opacification. **A.** Round cystic thin-walled mass coursing longitudinally from cranial to caudal parallel to the posterior wall of the ascending colon. **B.** Lower pole of the lesion adherent to and modeling the lateral wall of the cecum (arrow).



**Figure 3.** Follow-up sonography. **A.** Tubular cystic mass with focal internal irregularities of the wall (3,5 MHz transducer). **B.** Soft tissue mass in the lumen (7,5 MHz transducer).



**Figure 4.** Mucocele of the appendix. **A.** Atrophy of the muscularis membrane with proliferation of the connective tissue and inflammatory lymphatic infiltration. **B.** Mucocele lined with simple columnar epithelium. H&E stain, magnification x 400.

Cystadenoma was found in 50%-70%, cystadenocarcinoma in 10%-20%, mucocele with mucosal hyperplasia in 5%-25% and simple mucocele caused by obstruction in 5%-10% of all cases of appendiceal mucocele [1, 11, 12]. The mucocele can be complicated by a spontaneous or iatrogenic rupture which may result in pseudomyxoma peritonei – a spread of benign or malignant cells to the peritoneal cavity as multiple mucinous deposits [2, 3, 8, 13]. It can develop even up to 11 years after the removal of a benign appendiceal mucocele [2, 3]. Benign mucoceles are mostly reported in patients of mean age of 54 years and malignant types in older people (mean age 64 years) [13]. Dimensions of the mucoceles are different, but in cases of simple mucocele caused by obstruction, its diameter rarely exceeds 2 cm. Mucoceles larger than 2 cm are more likely of tumorous origin [8, 9, 14], although in our case the diameter of the simple mucocele was 35 mm measured at CT and 30 mm measured on macroscopic examination of the specimen. The length of the mucocele in most cases does not exceed 10-15 cm. Large mucoceles reaching 20-25 cm were also described, and a case report of a huge mucocele of 40 cm is cited [1, 6]. They can be unusually located e.g. on the left side of the abdomen [2].

Clinical presentation of the appendiceal mucocele may be nonspecific, and 25% of patients with even large mucoceles are asymptomatic [8]. The most common presenta-

tion is right lower quadrant pain. It can be recurrent, chronic or have a form of vague distress in the abdomen. In 20%-50% of cases, the pain can suggest acute appendicitis [8]. Rarely the pain is intermittent, colicky, caused by intussusception of the mucocele into the cecum, with sometimes gastrointestinal bleeding [2]. The patients can complain of fever, weakness, irregular stool, nausea, vomiting. Laboratory tests rarely reveal anaemia, leukocytosis or hematuria. In about 50% of patients, a painless and mobile mass is palpable in the right lower quadrant [1, 2, 4, 8, 11, 13, 15].

Akerlund was the first to establish preoperative radiographic diagnosis of mucocele in 1936. In 1947, Euphrat defined its radiographic criteria. These are: a) a sharply circumscribed globular or reniform soft tissue mass, with considerable mobility, but firmly attached to the cecum, b) medial displacement of the cecum by the mass, c) calcium deposits in the wall or substance of the mass, d) failure of the appendix to fill with contrast agent, and a concentric ring of the folds of cecal mucosa [2]. Dachman, in contrast to Euphrat, points to the lateral and/or cephalad displacement of the cecum. He also observed a „star” pattern of the mucosa at the base of the appendix on the barium enema, but this finding is not specific [2]. Barium enema may show also the usual features of intussusception or an intraluminal mass, which can mimic a colonic carcinoma;

an impression on the medial aspect of the cecum suggesting an extramucosal or extrinsic process can be visible.

Sonography in typical cases demonstrates the presence of a cystic tubular mass in the right lower quadrant. The wall may contain calcifications and the hypoechogenic lumen may be nonhomogenous. The cases were also presented with internal echoes, septations or polypoidal structures extending from the wall toward the lumen. The latter was seen in our patient. The content of the mucocele sometimes shifts after changing the position of the body during examination [1, 2, 7, 8]. Caspi et al. noticed echogenic layers inside the lumen of the mucocele, resembling onion skin and they found it to be a specific sonographic marker of appendiceal mucocele [16], although too few such cases have been described to regard it as a pathognomonic sign. In Polish literature, a case of an epidermoid cyst of the testis with sonographic sign of onion skin has been described lately [17]. Large mucoceles may have pear-shaped or drumstick appearance due to extrinsic compression or lesser dilatation of a portion of the appendix [14]. On either color or power Doppler imaging, no signs of vascularity are detected in mucoceles [6].

Computed tomography is very important in the diagnosis and in evaluation of the extent of the disease, because it has advantages over other imaging modalities. In CT examinations, mucocele of the appendix in typical cases is visible as a thin-walled cystic mass, well-encapsulated, with close relationship to the cecum in the expected location of the appendix. The degree of attenuation of the mucocele content depends on the amount of mucin in the mass, the density in HU can range from near water density to soft tissue density [1,2,3,7,14]. In most cases, no enhancement was seen, but exceptionally areas of faint enhancement were described inside the mass [6]. Sometimes there are calcifications in the wall of the mucocele - Zissin found them in 7 out of 10 cases [3]. The calcium deposits can be curvilinear or punctate [2, 3, 9]. Intramural calcifications seem to be highly suggestive of a neoplastic process, not necessarily malignant [9]. Soft-tissue thickening and irregularity of the mucocele wall and surrounding fat are nonspecific findings that suggest malignancy or secondary inflammation [14]. We found no calcification or contrast enhancement in our patient. No periappendiceal inflammation was seen, differentiating the image from appendicitis, in which also the wall is thicker with a characteristic „target sign” [7]. It is to be mentioned that adequate opacification with oral contrast material of the terminal ileum and cecum is essential to diagnose the mucocele of the appendix, because e.g. a fluid-filled terminal ileum loop may mimic mucocele. It can cause false-positive or false-negative findings [7,14].

Myxoglobulosis is a rare variant of mucocele, found in 0.35%-8% of cases, in which the appendiceal lumen is filled

with pearly, translucent spherules of 1-10 mm in diameter or a cluster of „frog eggs”, that can calcify and then they may be visible at radiography or CT [2, 7, 14]. Some studies suggest that the granulation tissue from the wall of the mucocele break off, undergo necrosis and calcify [2].

MR imaging confirms a finding of a cystic mass, but calcification will be less apparent [3,14].

At colonoscopy pathognomonic image is the „volcano sign” – an erythematous, soft mass with a central crater, from which mucus is discharged [8].

The differential diagnosis of the appendiceal mucocele might include other cystic lesions such as cystic ovarian neoplasm, tubo-ovarian abscess, hydrosalpinx, enteric duplication cyst, mesenteric cyst, urachal cyst, echinococcal cyst, abscess, organized hematoma or other entities as Meckel`s diverticulitis, appendicitis, neoplasm of the ileo-cecal region, extrauterine pregnancy [3, 7, 11, 14]. The diagnosis may be difficult especially when the mass is huge, when it is hard to precise the relationship with the cecal region [6], or when it is not clearly cystic.

In our case, the definite preoperative diagnosis was not established, although a cystic lesion with high probability of association with the gastrointestinal tract was suggested.

Mucocele of the appendix should be treated only by surgery - in cases of benign mucocele the appendectomy is performed and in malignant lesions the treatment of choice is right hemicolectomy [1, 8, 11, 13]. Laparoscopic approach is not advised because of the risk of rupture [8].

The prognosis depends on histology with 5-year survival rates of 91%-100% in cases of simple or benign mucocele and of 25% in malignant mucocele complicated with pseudomyxoma peritonei [8].

It is important that about 30% of cases of appendiceal mucocele are associated with other neoplastic tumors, especially colorectal malignancies [6, 9]. Therefore, the colon should be carefully examined before and as follow-up after an appendectomy for mucocele [8, 9, 12, 13]. In women it is essential to look for coexisting ovarian cystic tumors and in cases of cystic lesion in the ovary, especially with peritoneal pseudomyxoma, the appendiceal mucocele is also possible to coexist [3].

## Conclusions

In case of presence of a cystic lesion in the right lower quadrant of the abdomen in a patient without a history of appendectomy appendiceal mucocele is one of the differential diagnoses.

## References:

1. Souei-Mhiri M., Tlili-Graies K., Ben Cherifa L. et al.: Les mucoceles appendiculaires. Etude retrospective a propos de 10 cas. J Radiol, 2001; 82 (4): 463-468.
2. Dachman A.H., Lichtenstein J.E., Friedman A.C.: Mucocele of the appendix and pseudomyxoma peritonei. AJR, 1985; 144: 923-929.
3. Zissin R., Gayer G., Kots E. et al.: Imaging of mucocele of the appendix with emphasis on the CT findings: a report of the 10 cases. Clin Radiol, 1999; 54: 826-832.
4. Pospisil I., Jaros E., Hadzi D.N. et al.: Appendiceal mucocele. Rozhl Chir, 2004; 83 (9): 451-455.

5. Landen S., Bertrand C., Maddern G.J. et al.: Appendiceal mucoceles and pseudomyxoma peritonei. *Surg Gynecol Obstet*, 1992; 175 (5): 401–404.
6. Francica G., Lapicciarella G., Giardiello C. et al.: Giant mucocele of the appendix. Clinical and imaging findings in 3 cases. *J Ultrasound Med*, 2006; 25: 643–648.
7. Madwed D., Mindelzun R., Jeffrey R.B.: Mucocele of the appendix: imaging findings. *AJR*, 1992; 159: 69–72.
8. Rampone B., Roviello F., Marrelli D. et al.: Giant appendiceal mucocele: report of a case and brief review. *World J Gastroenterol*, 2005; 11 (30): 4761–4763.
9. Rakovich G., Larue N. : Mucocele of the appendix associated with a carcinoid of the terminal ileum. *J Can Chir*, 2007; 50 (1): 66–67.
10. Higa E., Rosai J., Pizzimbono C.A., et al.: Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendiceal “ mucocele”. *Cancer*, 1973; 32: 1525–1541.
11. Stang A., Braumann D., Teichmann W.: Mukozele der appendix. Sonographischer Zufallsbefund und laparoskopische Resektion. *Dtsch Med Wochenschr*, 2004; 129: 2295–2298.
12. Cordoliani Y.S., Simon P., Wannin G. et al.: Retrocecal mucus-secreting tumors of the appendix (mucoceles). Apropos of 3 radiologically diagnosed cases. *J Radiol*, 1988; 69 (12): 741–750.
13. Zullo A., Botto G., Pastormerlo M.: Un caso di mucocele appendicolare associato a cancro del colon. *Minerva Chir*, 1995; 50: 1095–1098.
14. Pickhardt P.J., Levy A., Rohrmann C.A. et al.: Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. *Radiographics*, 2003; 23: 645–662.
15. Kunin N., Musolino A., Skufca J. et al.: Appendiceal mucocele. *J Chir*, 2003; 140 (1): 46–47.
16. Caspi B., Cassif E., Auslender R. et al.: The onion skin sign. A specific sonographic marker of appendiceal mucocele. *J Ultrasound Med*, 2004; 23: 117–121.
17. Richter B., Walecka A., Gawrych E.: Epidermoid cyst of the testis in 12-year-old child: Case report. *Pol J Radiol*, 2007; 72 (1): 103–105.