

# Mucinous cystadenoma of the appendix – case report

## Authors' Contribution:

A – Study Design  
B – Data Collection  
C – Statistical Analysis  
D – Data Interpretation  
E – Manuscript Preparation  
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Dariusz Janczak<sup>1,2,ADEF</sup>, Mateusz Szponder<sup>2AEF</sup>, Dawid Janczak<sup>3EF</sup>, Michał Leśniak<sup>2ADE</sup>, Agnieszka Ziomek<sup>2EF</sup>, Mariusz Chabowski<sup>1,2 ADEF</sup>

<sup>1</sup>Department of Surgical Specialties, Faculty of Health Science, Wrocław Medical University; Head: Prof. Dariusz Janczak, MD PhD

<sup>2</sup>Department of Surgery, 4th Military Teaching Hospital in Wrocław; Head: Prof. Dariusz Janczak, MD PhD

<sup>3</sup>Department of Palliative Care Nursing, Faculty of Health Science, Wrocław Medical University

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## ABSTRACT:

Tumors of the appendix are extremely rare and constitute about 0.4% of all tumors of the gastrointestinal tract. The most common benign neoplasm is mucinous cystadenoma, which can be found in 0.6% of all excised appendices and it rarely produces any symptoms. We present the case of a female patient who underwent surgery in the Department of Surgery due to suspicion of a periappendicular abscess. On the postoperative pathology study, the diagnosis of a tumor of the appendix (mucinous cystadenoma) was made. Mucinous cystadenoma is rarely included in the differential diagnosis of a non-specific abdominal pain accompanied by non-characteristic laboratory test results and imaging studies. There are no unequivocal guidelines and algorithms of managing this disease. Long-term prognosis is good in the case of a benign tumor.

## KEYWORDS:

mucinous cystadenoma, appendicular tumors, pseudomyxoma peritonei

## INTRODUCTION

Tumors of the appendix are rare. They constitute approximately 0.4% of all gastrointestinal tract tumors [1]. The most common is the benign mucinous cystadenoma, which can be found in 0.6% of all excised appendices and does not produce any symptoms. However, it can often cause formation of so-called mucous cyst of the appendix [1,2]. A rare but potentially lethal complication of this condition is formation of pseudomyxoma peritonei, which is a collection of gelatinous matter in the abdominal cavity. The initial correct diagnosis preoperatively is extremely difficult in such cases due to the lack of clinical symptoms and non-specific clinical presentation.

## CASE REPORT

A 66-year-old female patient was admitted to the Department of Gastroenterology due to ascites and a 5-month history of persistent abdominal pain of varying intensity with flatulence. On physical examination, the only noticeable abnormality was abdominal tenderness on deep palpation, mainly in the right lower quadrant. Laboratory tests showed mildly increased level of CRP – 11.2mg/l (normal < 5.0 mg/l) and CEA – 6.86 ng/ml (normal < 4.0 ng/ml), other results being within the normal range. Abdominal ultrasound revealed a trace of free fluid in the peritoneal cavity, surrounding the liver and between bowel loops. Abdominal and pelvic CT scan showed large collection of free fluid, which was localized mainly between bowel loops on the right side, and in the vesico-uterine and recto-uterine pouches. At the border between the pelvis and the abdomen on the right side, an oval mass in the size of 5x5.5x3cm surrounded by fluid and contrast-enhancing wall was visible. Additionally, its connection to the thickened appendix was noted. Radiological image suggested periappendiceal abscess (Figure 1). Antibiotics were introduced in this patient. Then, having obtained surgical consultation, the patient was transferred to the Department of Surgery for further surgical management. During laparotomy, the presence of ca. 500ml of gelatinous matter in the peritoneal cavity and a distended and ruptured end of the appendix

were observed (Figure 2). Intraoperatively, a diagnosis of a ruptured mucous cyst of the appendix was made, the appendix itself was excised in a typical manner and sent with a sample of the intraperitoneal mucous for pathology study. Further intra- and postoperative course was uncomplicated. The patient was discharged home in good general and local condition. On pathology study, the mucinous cystadenoma of the appendix with low-grade dysplasia was diagnosed. Within the subserosal layer and on the serosa, the presence of mucous accompanied by chronic inflammation and calcium deposits were noted. Intraperitoneal mucous did not contain cancerous cells. One year after surgery, contact with the patient was restored and she did not report any long-term complications or symptoms associated with the disease or treatment.

## DISCUSSION

Benign tumors of the appendix are rarely observed. The most common clinical presentation is mucocele, i.e. a mucous cyst, which predominates in females, the mean age at the diagnosis being 50-55 [3]. It can be found in 0.2-0.3% of all removed appendices [1,4]. It is characterized by lumen dilatation and excessive collection of mucous, and it can develop in a number of ways; a simple cyst forms by obstruction of the lumen of the appendix with normal secretion of mucous, which can be caused by e.g. a fecolith, gallstone, endometriosis, postoperative adhesions, volvulus of the appendix or intestinal tumor [5,6]. Similar mechanism, although with more secretion, is observed in the case of mucosal overgrowth without atypia, however, even in such instance, the lesions do not exceed 2cm in diameter and rarely pose a threat of perforation. The situation is different in the case of a mucinous cystadenoma (benign neoplasm) and mucinous cystadenocarcinoma (cancer) with moderate or advanced dysplasia, which can reach 6cm in size (or even 25cm – gigantic myxoma) and it can be symptomatic and perforate [1,3,4,5].

In the majority of cases, those tumors are mildly symptomatic or asymptomatic. Although clinical symptoms are non-specific,

a palpable mass, tenderness or pain in the right lower quadrant are often found on physical examination. It can sometimes be accompanied by urinary tract infection or hematuria, very rarely by lower gastrointestinal bleeding caused by intussusception [1]. Laboratory test results do not indicate any specific answer, although – in addition to elevated CRP level – elevated levels of tumor markers (CEA and CA19-9) can be observed, which suggests neoplastic etiology or a coexisting disease [3,7]. The best diagnostic imaging method is the computed tomography, which allows to visualize an encapsulated thin-walled mass filled with hypoechogenic fluid with possible calcifications in 50% of patients. Abdominal ultrasound usually does not bring much information while it yields an image corresponding with many possible diagnoses. Some authors report a pathognomonic ‘volcano sign’ on colonoscopy, however, it is usually impossible to conduct this examination properly in such situations [3,7]. It all causes that mucinous cysts are diagnosed accidentally during surgery for other indications in half of all cases. The most common preoperative misdiagnoses include acute appendicitis, intraabdominal abscess, but also other conditions such as ovarian or kidney cysts, echinococcosis, intraabdominal hernia etc. [8]. Due to an asymptomatic course, the diagnosis of mucinous cystadenoma can be delayed, which may result in perforation with formation of so-called pseudomyxoma peritonei and spreading of cancerous cells within the peritoneal cavity as multiple mucinous deposits (gelatinous mass, as in this case). It usually manifests itself with gradual (throughout months or years) abdominal distention due to slow accumulation of mucous inside the abdominal cavity [7,9]. The chronic process can lead to fibrosis and adhesions in the peritoneum, an hence to ileus – and therefore it is a potentially lethal complication [8]. The prognosis is even worse when pseudomyxoma develops from a malignant lesion.

Mucinous tumors of the appendix can be accompanied by other gastrointestinal tumors (colorectal tumors, mucinous ovarian cysts, pancreatic and liver cancer), especially when features of advanced atypia were determined postoperatively [3]. For example, the risk of developing colorectal adenocarcinoma in such patients is even 6 times higher than in general population. Therefore, it is reasonable to conduct detailed oncologic follow-up (e.g. colonoscopy not long after surgery) [5,6]. As for the surgery itself, authors present varied approaches. Usually, the method of operation (laparoscopic vs open surgery) is chosen based on other indications and local technical capabilities, considering rarity of accurate preoperative diagnosis of mucocele. Generally, it is assumed that regardless of the surgical approach, classical appendectomy is performed when a distended appendix is noticed during surgery, pathological changes involving only the appendix. When the lesion widely involves the base of the appendix or protrudes into the intestinal lumen, partial resection of the caecum is recommended. However, when the intestinal wall is involved, or malignancy is suspected, right colectomy is usually performed. Particular caution during preparation and thorough inspection of the abdomen are always advised after diagnosis of mucocele, in order to assess mucinous inoculation or concurrent lesions [1]. In the case of pseudomyxoma peritonei, effort should be made to remove all visible inoculations, although it is difficult and usually unsuccessful [7]. The optimal surgical technique in the case of a preoperative diagnosis of mucocele is being discussed. Some authors recommend laparoscopic appendectomy in simple cases, suggesting that it allows for better

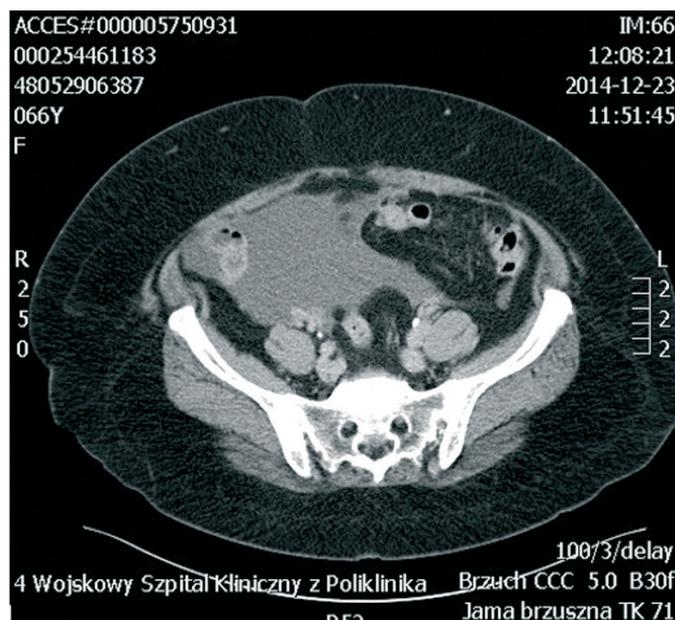


Fig. 1. Preoperative pelvic CT scan. Free fluid between intestinal loops and an oval structure connecting with the appendix were visualized.



Fig. 2. Intraoperative image. Distended and perforated appendix.

inspection of the abdominal cavity and to avoid mucinous inoculation by using endobags [9,10]. Most authors, however, prefer open surgery, preferably applying intraoperative quick pathology assessment. Conversion is unarguably necessary in the case of wall damage to the pathologically altered appendix or in the presence of pseudomyxoma peritonei. When the intraoperative pathology study suggests cancer, right colectomy is performed [11]. Further treatment should be conducted in the tertiary referral oncology center and it can include surgical cytoreduction of peritoneal inoculations with concurrent application of chemotherapeutic agents into the abdominal cavity. It is performed in selected advanced cases at very few facilities and is, unfortunately, associated with high mortality rate [6,7]. 5-year survival in patients with a malignant mucinous tumor of the appendix and pseudomyxoma peritonei is only 53% [8]. In the case of benign tumors, 5-year survival reaches 100% [3,12].

## SUMMARY

Mucinous cystadenoma is extremely rarely included in the differential diagnosis of a non-specific abdominal pain accompanied by non-characteristic or even misleading laboratory test results and abdominal imaging studies. Thus far, there are no unequivocal

guidelines and algorithms for managing this disease. The prognosis is, in the case of a benign tumor, good, however, the diagnosis

requires special attention and caution during surgery and oncologic surveillance during follow-up observation.

## REFERENCES

1. Jha N.K., Sinha D.K., Anand A. et al.: Mucinous cystadenoma of the appendix with enterocutaneous fistula: a therapeutic dilemma. *Gastroenterol. Rep. (Oxf)*. 2015; 3 (1): 86–89.
2. Dellaportas D., Vlahos N.F., Polymeneas G., Gkiokas G. et al.: Collision Tumor of the Appendix: Mucinous Cystadenoma and Carcinoid. *A Case Report. Chirurgia*. 2014; 109: 843–845.
3. Djuranovic S.P., Spuran M.M., Kovacevic N.V. et al.: Mucinous cystadenoma of the appendix associated with adenocarcinoma of the sigmoid colon and hepatocellular carcinoma of the liver: Report of a case. *World J. Gastroenterol*. 2006; 12 (12): 1975–1977.
4. Alese O.B., Irabor D.O.: Mucinous cystadenoma of the appendix: a case report. *African Health Sciences*. 2010; 10 (1): 99–100.
5. Honnef I., Moschopoulos M., Roeren T.: Appendiceal Mucinous Cystadenoma. *Radio Graphics*. 2008; 28: 1524–1527.
6. Rabie M.E., Al Shraim M., Al Skaini M.S. et al.: Mucus containing cystic lesions "mucocele" of the appendix: the unresolved issues. *Int. J. Surg. Oncol*. 2015; 2015:139461.
7. Rymer B., Forsythe R.O., Husada G.: Mucocele and mucinous tumours of the appendix: A review of the literature. *International Journal of Surgery*. 2015; 18: 132–135.
8. Krieg A., am Esch J.S. 2nd, Poll L.W. et al.: Mucinous cystadenoma of the appendix misdiagnosed as cystic hydatid disease of the liver: a case report. *Journal of Medical Case Reports* 2008, 2: 218.
9. Rouchaud A., Glas L., Gayet M., Bellin M.F.: Appendiceal mucinous cystadenoma. *Diagnostic and Interventional Imaging*. 2014; 95: 113–116.
10. Ng S.S.M., Lee J.F.Y.: Mucinous cystadenoma of the appendix. *Can. J. Surg*. 2009; 52 (2): 158–159.
11. Park K.J., Choi H.J., Kim S.H.: Laparoscopic approach to mucocele of appendiceal mucinous cystadenoma: feasibility and short-term outcomes in 24 consecutive cases. *Surg. Endosc*. 2015; 29 (11): 3179–3183.
12. Hibi K., Mizutani M., Imazawa M. et al.: Mucinous cystadenoma of the appendix associated with muscular and neuromatous hyperplasia: Report of a case. *Nagoya J. Med. Sci*. 2008; 70: 35–40.

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Corresponding author: Mariusz Chabowski; Postal address: Dept of Surgery, 4th Military Teaching Hospital, 5 Weigla street, 50-981 Wrocław, Poland, Phone: (+48) 261 660 247, Fax: (+48) 261 660 245; e-mail: [mariusz.chabowski@gmail.com](mailto:mariusz.chabowski@gmail.com)

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