

## Laparoscopic Right Colectomy For Appendiceal Mucinous Cystadenoma: A Case Report

Dion A. Putra\*

Adianto Nugroho<sup>†</sup>

Ibrahim Basir<sup>‡</sup>

\*University of Indonesia, dison21308@yahoo.com

<sup>†</sup>University of Indonesia, adiyusuf97@gmail.com

<sup>‡</sup>University of Indonesia, wimbaser@gmail.com

# Laparoscopic Right Colectomy For Appendiceal Mucinous Cystadenoma: A Case Report

Dion A. Putra, Adianto Nugroho, and Ibrahim Basir

## Abstract

**Background:** Appendiceal mucoceles are an uncommon pathology of the appendix characterized by an expansion of the lumen with retention of mucus. The symptoms of cystadenoma is non-specific or often mimics a chronic appendicitis.

**Case Report:** We present a case of a 63-year-old male with appendiceal mucinous cystadenoma, presenting with intermittent pain in the right lower quadrant of his abdomen.

**Results:** Laparoscopic assisted right colectomy was done with ileotransverse colonic anastomosis. The postoperative course was uneventful.

**Conclusions:** Prevention of rupture with minimal manipulation and negative surgical margin are two most important technical considerations in the resection of appendiceal mucinous cystadenoma. Patient has benefited from the use of laparoscopic assisted technique because of less pain and shorter hospital stay after the procedure.

**KEYWORDS:** appendiceal mucinous cystadenoma, laparoscopic assisted, colectomy

## Introduction

Appendiceal mucinous cystadenoma is a rare cystic neoplasm of the appendix characterized by villous adenomatous changes in the epithelium associated with a marked appendiceal intraluminal distention with mucin. Incidental finding during procedures for other diagnoses is a common presentation of appendiceal mucinous cystadenoma since there are no specific symptoms related to this pathology. A high index of suspicion should be applied for patients greater than 50 years old with symptoms mimicking chronic appendicitis. Early treatment before dissemination to form a pseudomyxoma peritonei is advisable.

## Case Report

We present a case of a 63-year-old male with intermittent pain in the right lower quadrant of the abdomen, with a history of hematochezia and altered bowel habits. The patient had minimal tenderness in the right lower quadrant. His physical examination was otherwise. Previous colonoscopy revealed an internal hemorrhoid and a small cystic mass at the cecum. No further treatment was done after the first colonoscopy. On the second visit, he underwent colonoscopy and CT scan which revealed a large mucocele of the appendix (4 x 5 x 13.8 cm).

The patient underwent laparoscopic assisted right colectomy with ileotransverse anastomosis. The histopathologic examination showed appendiceal mucinous cystadenoma

## Discussion

Mucinous cystadenoma is one subtype of cystic type/mucocele of appendix.<sup>1</sup> The mucocele of the appendix is a descriptive and non-specific term to define the cystic dilation of the appendix caused by the accumulation of mucus secretion.<sup>2,3</sup> This process is slow and gradual, with no signs of infection inside the organ. Cystadenoma and mucinous cystadenoma are frequently used terms for benign neoplastic mucoceles. The term mucocele is a general macroscopic description that implies a dilated appendiceal lumen caused by an abnormal accumulation of mucus. It does not constitute a pathologic diagnosis. It may be caused by a variety of non-neoplastic, benign neoplastic, and malignant underlying pathologic entities. Mucoceles may be discovered incidentally, at the time of physical examination or abdominal imaging, or as a secondary surgical finding. Most patients present with acute or chronic right lower abdominal pain secondary to cystic distention of the

appendix by mucus. A palpable mass at right lower abdomen is found in up to 50% of patients and may be associated with nausea and vomiting. Symptoms may be similar to chronic appendicitis. Rare manifestations include intussusception, torsion, gastrointestinal bleeding, altered bowel habits, weight loss, vomiting, and urologic symptoms such as right ureteral obstruction. Approximately 25% of patients with an appendiceal mucocele are asymptomatic. Mucoceles of the appendix are uncommon. They have been observed in only 0.2–0.3% of surgical appendectomy specimens. They typically occur in people who are middle aged or older, and more often in women than in men. Some sources may claim a higher frequency in women (4:1) and in patients >50 years of age,<sup>2,4</sup> while others demonstrate a higher incidence among men.<sup>5</sup>

Four varieties of mucoceles have been described, according to the predominant feature of the epithelium: one has a normal epithelium, and three show varying degrees of epithelial atypia. The first type, the simple retention cyst, is characterized by a normal appendiceal epithelium and is associated with mild luminal dilatation that rarely exceeds 2 cm in diameter. There is no evidence of hyperplasia or mucosal atypia. Any mucocele with a diameter of more than 2 cm is more likely to represent one of the other three types of mucocele. The second group, which accounts for 5–25% of mucoceles, is defined by a hyperplastic epithelium and mild dilatation. The appendix dilation occurs due to the hyperplastic growth of the appendix or cecal mucosa, just like hyperplastic polyps in the colon. Simple and hyperplastic mucocele mucus is usually acellular. In the third group, which accounts for 63–84% of mucoceles, the appearance of the epithelium is similar to that in villous adenomas and adenomatous polyps of the colon, with mild (low-grade) epithelial dysplasia. Often termed cystadenomas, as in the case presented here. These mucoceles produce marked distention of the lumen. Appendiceal perforation occurs in 20% of cases, with resultant mucinous spillage into the periappendicular area or onto the serosal surface. No neoplastic cells are found in the mucus at histologic examination, and surgical resection is usually curative. The fourth category, which accounts for 11–20% of cases, encompasses malignant mucinous cystadenocarcinomas. The neoplastic epithelium in these tumors is similar to that seen in adenocarcinomas of the colon, with glandular stromal invasion. Appendiceal distention is severe, and spontaneous rupture has occurred in 6% of cases. The rupture of the appendix may lead to the dissemination of the epithelium that produces mucins in the abdominal cavity, causing mucinous ascites or pseudomyxoma peritonei. The term pseudomyxoma peritonei refers to the accumulation of gelatinous material on intraperitoneal surfaces either because of the localized rupture of a benign mucocele or, more commonly, because of the diffuse proliferation of viable neoplastic cells throughout the peritoneum. Simultaneous disease is found in the appendix and the ovary in most women with

this type of peritoneal extension. The peritoneal cavity is distended by adhesive, semisolid mucin.<sup>2,3,6</sup>

Thus, both the benign and the malignant mucocele may cause pseudomyxoma peritonei. However, this complication is more frequent and has worse prognosis for malignant cases. This is probably because, in this situation, the appendix ruptures more easily, and the cellular seeding is more aggressive.<sup>3</sup>

The macroscopic appearance is gross enlargement of the appendix, with luminal distention by mucin. The mucin-rich epithelium often demonstrates a villous architecture. Mucinous cystadenomas tend to be low grade, with circumferential involvement. Progressive mucocele formation results in pressure atrophy and thinning of the wall with fibrosis. The presence of invasive neoplastic cells beyond the muscularis mucosa is indicative of adenocarcinoma.<sup>6</sup>

The elevation of preoperative carcinoembryonic antigen (CEA) level may suggest malignancy in the appendix or in the colon.<sup>3</sup> Imaging tests, such as ultrasound, computed tomography and contrast enema, besides colonoscopy, may suggest the presence of mucocele of the appendix, which helps to define the treatment.<sup>3</sup>

The ultrasound shows an encapsulated cystic lesion in the lower quadrant of the abdomen with a liquid content of variable echogenicity, according to the density of the mucus. Some tests present images of multiple echogenic layers in the dilated appendix, which may be considered as pathognomonic of appendix mucocele.<sup>3</sup>

CT is the modality of choice because of its ability to depict the anatomic location of a mass, as well as the tissue characteristics. Mural curvilinear calcification aids considerably in the diagnosis but is thought to occur in less than 50% of cases. The typical finding in cystadenomas of the appendix is a round, thin-walled, encapsulated cystic mass that communicates with the cecum. The attenuation of the contents of the cyst may range from that of water to that of soft tissue, depending on the amount of mucin or debris within the mucocele. Adjacent bowel segments may be displaced, but usually no periappendicular inflammation or abscess is seen. These findings help differentiate a mucocele from a periappendiceal abscess or pelvic inflammatory process. Other possible diagnoses, depending on the imaging findings, may be an enteric duplication cyst or mesenteric cyst. Soft tissue thickening and irregularity of the cystadenoma wall and surrounding fat are nonspecific, atypical findings that suggest malignancy, secondary inflammation, or an unusual pathologic variation. Intraluminal gas bubbles or an air-fluid level within a calcified right lower quadrant cystic mass are helpful indicators of superinfection, which may occur in both benign and malignant variants. In

suspected cases of the most severe form of this disease entity, pseudomyxoma peritonei, CT is widely used to establish the diagnosis and characterize the extent of disease.<sup>6</sup>

With contrast enema, the presence of mucocele can be characterized by a cecal filling defect, besides the lateral displacement of the cecum and terminal ileum.<sup>3</sup>

Colonoscopy may show a soft erythematous mass, with a central crater due to the protrusion of appendiceal ostium, which can increase or decrease according to the respiratory movement. This condition is known as “volcano sign”. This examination may still show the presence of synchronic neoplastic lesions in the colon, which occur in up to 20% of the cases.<sup>3</sup>

The cytology of the mucus inside the mucocele obtained by puncture with a thin needle may distinguish benign and malignant forms, but it should not be used due to the risk of cell dissemination and evolution to pseudomyxoma peritonei.<sup>3</sup>

The treatment of cystic appendiceal masses is primarily surgical excision to eliminate any malignant potential. A laparoscopic approach is not advised by some authors, because of the risk of rupture and associated peritoneal contamination.<sup>6</sup> The treatment of appendix mucocele is surgery and determined by some factors, such as the integrity of the wall of the organ, the dimensions of its base and histopathological examination of the cause of mucocele.<sup>3</sup>

However, some authors disagree as to the recommendation to avoid laparoscopic for mucocele, by saying that laparoscopic appendectomy to handle appendix tumors has more recent results, similar to the open technique. In this case, there is a careful manipulation of the organ and the use of protective envelopes. However, although it is technically possible to remove the intact appendix with free resection margins with laparoscopy, the advantage of open surgery is the release and exteriorization of the cecum, avoiding the contamination of the cavity in case of accidental rupture of the appendix.<sup>3</sup>

As to follow-up, a recent study suggests histological, clinical and genetic similarities in proliferative lesions of the appendix and colonic mucosa.<sup>6</sup>

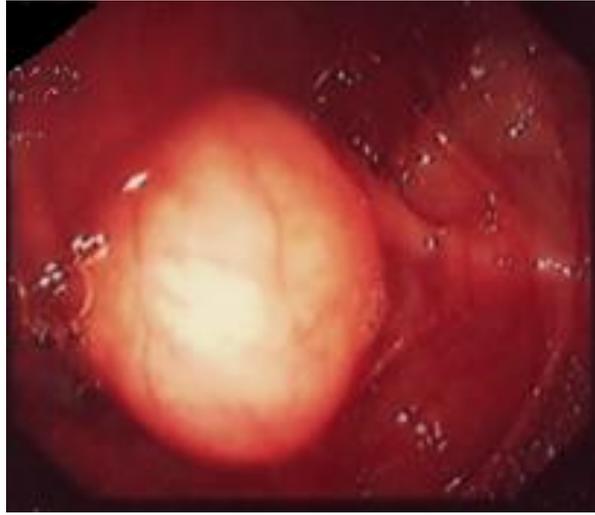
Therefore, simple and hyperplastic mucocèles would not require follow-up, benign mucocele would be followed up as an adenomatous polyp with colonoscopies, according to the follow-up of colonic adenomas, and the malignant one would be followed up as a colonic adenocarcinoma, with surveillance of CEA and serial colonoscopies.<sup>6</sup>

## References

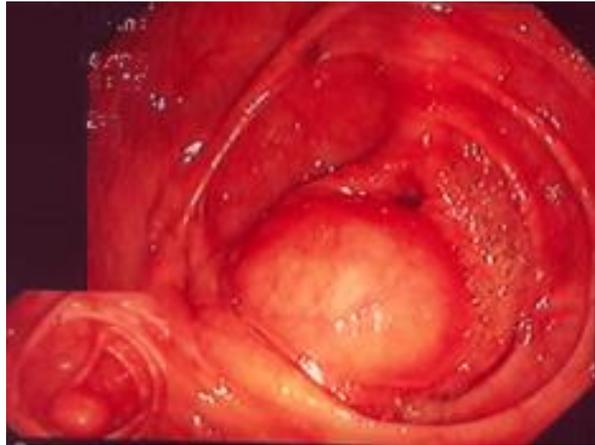
1. Uihlein A, McDonald JR. Primary carcinoma of the appendix resembling carcinoma of the colon. *Surg Gynec & Obst.* 1943;76:711-4.
2. Sierra-Montenegro E, Sierra-Luzuriaga G, Leone-Stay G, Quinonez-Auria C, Salazar-Menendez V. Mucinous cystadenoma of the appendix: a case report. *J Digest Surg.* 2010;255-8.
3. De Abreu Filho JG, De Lira EF. Mucocele of the appendix: appendectomy or colectomy? *J Coloproctol.* 2011;31:276-84.
4. Stocchi L, Wolff BG, Larson DR, Harrington JR. Surgical treatment of appendiceal mucocele. *Arch Surg.* 2003;138(6):585-9.
5. Ruiz-Tovar J, Teruel DG, Castineiras VM, Dehesa AS, Quindos OL, Molina EM. Mucocele of the appendix. *World J Surg.* 2007;31(3):542-8.
6. Persaud T, Swan N, Torreggiani WC. Giant mucinous cystadenoma of the appendix. *Radiographics.* 2007;27(2):553-7.

**Figure 1.** Colonoscopy images. **A.** Round mass, cystic at cecum sized 1.5 x 2 cm. **B.** Protrusion of the appendiceal ostium, “volcano sign”.

**A.**

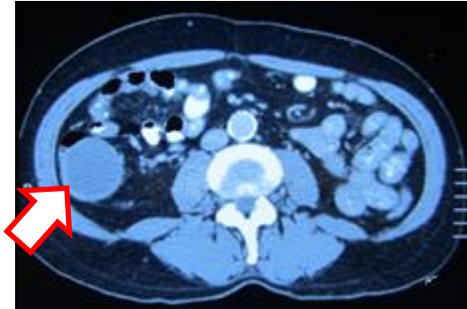


**B.**

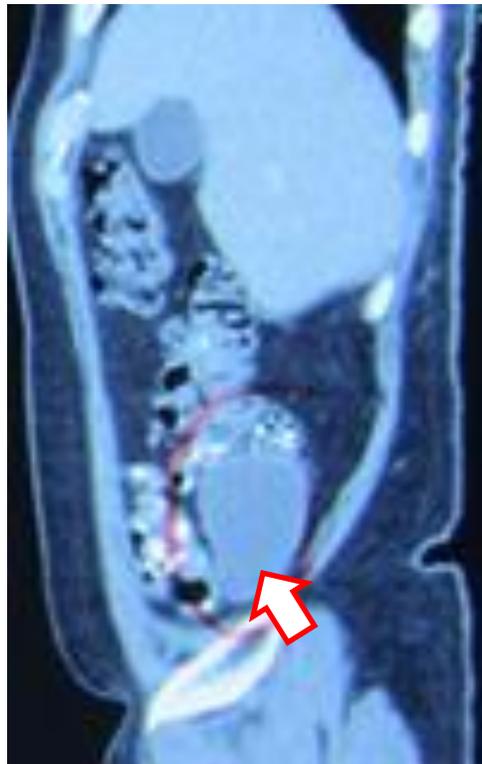


**Figure 2.** Abdominal computer tomography demonstrated a hypodense lesion, homogen density, cystic mass with a thin wall, which communicates directly with the cecum. The cecum was normal. There was no lymphadenopathy

**A.**



**B.**



C.

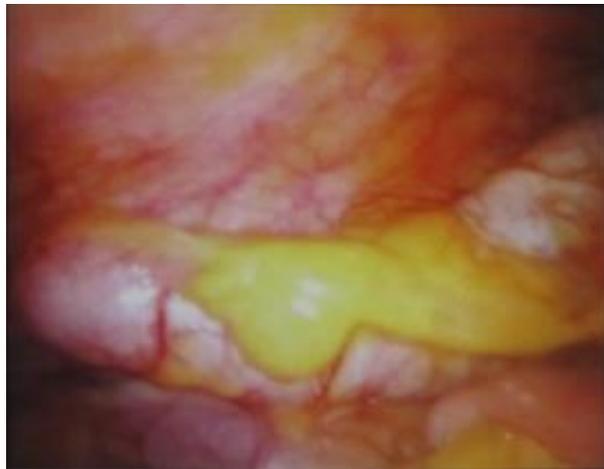


**Figure 3.** Colonoscopy demonstrated round cystic mass covering the appendiceal ostium.



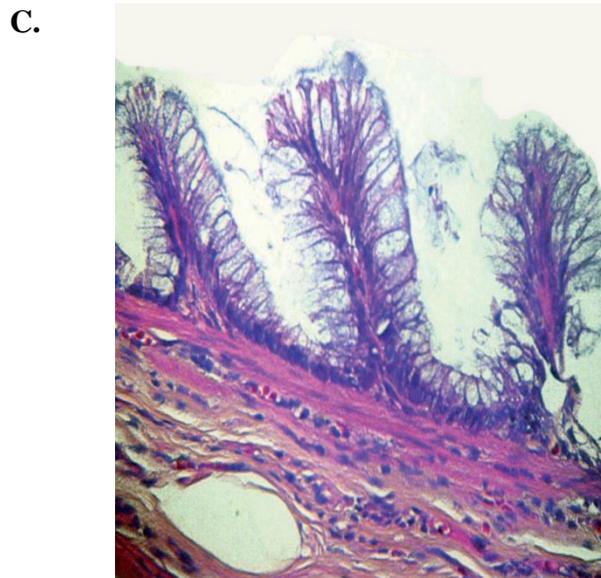
**Figure 4. A.** Intraoperative finding of inflamed and edematous mucocele of appendix. **B.** Appendiceal specimen with base. >2 cm wide and 10 cm long. Appendix contained mucin/gelatin like semisolid mucin. **C.** Histopathology result. Free margin, appendix contained gelatin like-mucin and lined by simple cylindrical mucous epithelium without atypia. 8 lymph nodes identified. No signs of malignancy or spillage of mucus from lumen. Histologically was compatible with cystadenoma mucinosum appendix.

**A.**



**B.**





**Figure 5. Treatment protocol of mucocoele<sup>3</sup>**

