Case Report

Three Cases of Appendiceal Mucocele: From Diagnosis to Management

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Abstract

Appendiceal mucocele is an appendicular dilatation secondary to the intraluminal accumulation of mucous material. Adequate pre-operative diagnosis and surgical resection remains the standard management. Here, we present three cases of appendiceal mucocele. In the first case, a 60-year-old female presented with signs and symptoms of acute appendicitis and was admitted and operated. An inflamed distended globular cystic mass of appendix measuring 10 × 6 × 4 cm with a wide base was found and the patient underwent right hemicolectomy. In the second case, a 30-year-old male with symptoms and signs of acute appendicitis was admitted to the emergency department. An open surgery was performed and a distended, tense, and inflamed appendix without perforation of size 6 × 1 × 1 cm was discovered and removed. The diagnosis of mucocele appendix was suspected and confirmed by postoperative dissection of the specimen and histopathology. In the third case, a 25-year-old female patient was subjected to diagnostic laparoscopy in view of non-specific pain abdomen. A diagnosis of mucocele of appendix was made intraoperatively and removed using a specimen bag. Appendiceal mucocele with acute presentation is a rare pathology that clinically resembles acute appendicitis. Preoperative detailed investigations to reach a definitive diagnosis are critical for adequate surgical resection and overall outcome.

Keywords: Appendectomy, appendix, mucinous cystadenoma, mucocele, rare disease

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INTRODUCTION

Appendiceal mucocele is an uncommon surgical emergency secondary to intraluminal accumulation of mucoid material due to blockage induced by a variety of neoplastic and non-neoplastic causes. Mucocele is found in approximately 0.2% to 0.7% of appendectomy specimens.[1,2] Because of the non-specific symptoms, this condition can be difficult to differentiate from appendicitis and may occasionally coexist.[3,4] Based on histologic examination, mucinous neoplasms of the appendix are classified on a spectrum from benign mucinous cystadenoma with no risk of recurrence to malignant mucinous adenocarcinoma with poor prognosis and high rate of metastasis to lymph nodes and liver.[5] In 2012, the Peritoneal Surface Oncology Group International[6] classified appendiceal mucinous lesions into two categories: non-neoplastic appendiceal mucinous lesions (simple mucoceles or retention cysts) and neoplastic appendiceal mucinous lesions (serrated lesions with or without dysplasia, mucinous appendiceal neoplasms, and mucinous adenocarcinomas of the appendix). Although the primary neoplasms of appendix are common,

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only few cases of mucocele appendix have been reported in the medical literature. Here, we present three cases of appendicular mucocele, with the diagnosis and management and a brief review of the literature.

CASE REPORTS

Case 1

A 60-year-old female was referred to our accident and emergency unit by the community health center after presenting with a 1-day history of pain in the right lower quadrant of the abdomen, nausea, and vomiting. The pain was acute in onset and progressive in nature. She had a strong appetite. Her surgical, medical, obstetric, personal, and family histories were not significant. The patient did neither had history of prior hospitalization nor any constipation/diarrhea, fever, rigors, or rectal bleeding. The baseline investigations are presented in Table 1. Leukocytosis (15.3 × 10^9/L) was notable from laboratory test results with 82% neutrophils. She was well, afebrile, and had pulse of 92 bpm, blood pressure 120/70 mmHg, respiratory rate 18 breaths/min, and oxygen saturation of 96%. She had tenderness to palpation in the right lower quadrant of the abdomen and had no peritoneal signs.

The digital rectal examination revealed normal anal tone and an empty rectum with no signs of bleeding. Ultrasonography of the abdomen was performed and revealed a cystic mass measuring 6.5 × 5.5 cm^2 arising from the appendix. A contrast-enhanced computed tomography scan of the abdomen and pelvis revealed a cystic mass arising from the caecum and adherent to adjacent small gut loops. There was no regional mesenteric lymphadenopathy. The liver's size and texture were normal, with no evidence of metastatic deposits. The patient was taken for emergency surgical exploration.

An inflamed globular cystic mass of appendix measuring 10 × 6 × 4 cm that arose from the appendix with dilated appendicular base without any perforation and abscess formation was seen [Figure 1]. The patient was subjected to right hemicolectomy and primary anastomosis. The postoperative period was smooth, and the patient was discharged on the fourth postoperative day in a good condition. Histopathological examination of the specimen confirmed the diagnosis of mucinous cystadenoma carcinoma with free resection margins. At the time of reporting this case, the patient was doing well after 3 years of regular follow-up.

Table 1: Biochemical and hematological parameters

Parameter	Case 1	Case 2	Case 3
Hb (g/dl)	11	13	11.4
TLC (cm/mm^3)	15.3	13.1	6.5
Neutrophil/lymphocyte (%)	82/41	87/33	68/42
PLT (/mm^3)	197	102	150
Urea (mg/dl)	23	20	28
Creatinine (mg/dl)	0.79	0.13	0.16
Sodium/potassium (mmol/l)	142/3.1	138/3.4	142/3.7
Blood glucose (mg/dl)	102	98	92
Total bilirubin	1.2	1	0.9
AST/ALT/ALP (units/l)	30/40/48	20/40/38	20/28/38
Total protein/albumin (g/dl)	7.2/3.8	7.7/4.5	7/4.2

Hb – Hemoglobin; TLC – Total leukocyte count; PLT – Platelets; AST – Aspartate aminotransferase; ALT – Alanine transaminase; ALP – Alkaline phosphatase

Case 2

A 30-year-old male who was normotensive, nondiabetic, and euthyroid presented with a 1-day history of diffuse abdomen pain associated with nausea, vomiting and a decreased appetite. Pain was insidious onset, migratory, and progressive in nature. Laboratory parameters indicated acute appendicitis [Table 1]. His blood glucose, liver and renal profiles were normal. The patient was hemodynamically stable. His respiratory and cardiovascular examinations were unremarkable. On palpation, the patient's abdomen was distended and there was moderate rebound tenderness in the right iliac fossa, which indicated localized peritonitis. McBurney's sign, Rovsing sign, and Dunphy's sign were all present, indicating acute appendicitis.

Digital rectal examination was unremarkable. Ultrasonography of the abdomen showed elongated cystic, dilated, aperistaltic and non-compressible gut loop in the right lower quadrant with internal echoes. No interloop fluid or mesenteric lymph nodes were recorded. Appendectomy was performed, and the intraoperative findings were suggestive of mucocele of appendix 6 × 1 × 1 cm and was removed [Figure 2]. There was no complication in the postoperative period. Histopathology revealed crowded, tubular structures, without epithelial atypia together with acellular mucin pooling (35-ml mucin). No evidence of any malignancy was seen. The patient was doing well as of 18 months of follow up following surgery.

Case 3

A 35-year-old female patient with no underlying comorbidity presented with a history of multiple emergency room admissions for pain in the right lower quadrant of the abdomen in the past 12 months. She reported no history of bleeding per rectum/malena, altered bowel habits, or weight loss. The patient did not have history of significant surgical or medical condition. The patient was thoroughly evaluated and subjected to the necessary investigation. Physical and clinical examination, digital rectal, pelvic and proctoscopy examination and biochemical parameters were unremarkable except for mild tenderness over the McBurney's point [Table 1]. Abdominal ultrasonography revealed a cystic mass measuring 2 × 2 × 1 cm arising from

right adnexa. An abdominal contrast-enhanced computed tomography scan demonstrated low-attenuation mass arising from the appendix in the right lower quadrant without any peri-appendiceal fluid, inflammation, or abscess.

The patient was listed for diagnostic laparoscopy. Mucocele of the appendix was diagnosed without perforation or collection. The caecum, terminal ileum loops, and bilateral adnexa were grossly normal. No discharge or peritoneal fluid was noted on laparoscopy. Then, the patient was subjected to appendectomy and the specimen was retrieved using a specimen bag [Figure 3]. Post-operative recovery was uneventful. Histological findings confirmed the diagnosis of a mucinous cystadenoma (mucocele) of the appendix. The patient was being followed-up regularly without any complaints.

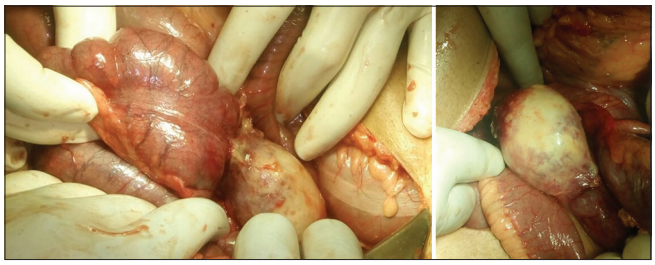

Figure 1: Mucocele of the appendix with a wide base

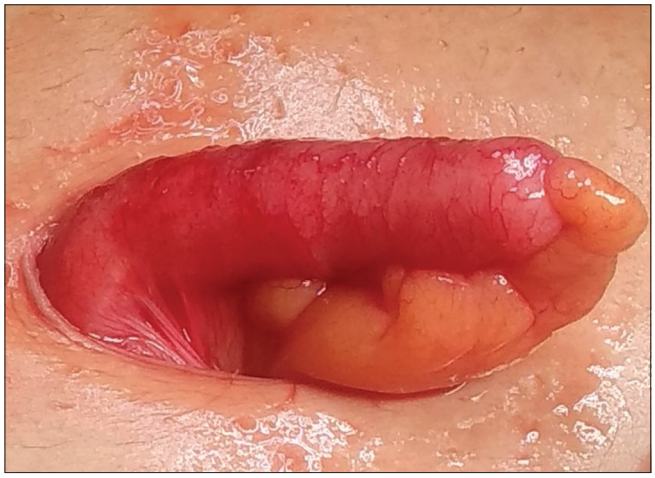

Figure 2: Intraoperative image of the mucocele of the appendix

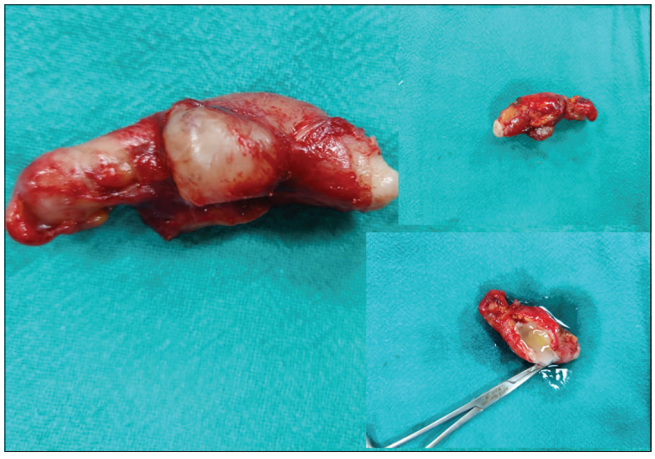

Figure 3: A mucocele of the appendix with lumen filled with mucus

DISCUSSION

A mucocele of the appendix is the result of obstruction of the appendiceal orifice with distention of the appendix caused by intraluminal accumulation of mucoid material.[7] It is rare disease, with a reported incidence of 0.2% to 0.7%,[1,2] four times more common in females, and typically detected in patients aged <50 years.[8] The typical clinical manifestations are non-specific and for some it may resemble acute appendicitis, while others may experience a palpable right lower quadrant mass, intermittent colicky pain, diarrhea, and/or rectal bleeding. The urinary symptoms are rare and palpable per abdomen mass is the most common presentation and is reported in about half the cases.[9-11] Some patients remain asymptomatic, and approximately half are accidentally diagnosed during an evaluation for another disease or during surgery.[9]

Four histopathological subtypes of mucocele of appendix reported are as follows: a simple mucocele, focal or diffuse mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.[12,13] A proper pre-operative workup to reach a definitive diagnosis is required for adequate surgical management to avoid the associated intraoperative and postoperative grave complications as well as repeated surgery. Pre-operative diagnosis aids in decision-making for the appropriate surgical procedure and alerts the operating surgeon to meticulous surgical dissection with extreme caution to avoid mucus spillage into the peritoneal cavity.

Pre-operative diagnostic imaging studies such as ultrasonography, contrast-enhanced computed tomography, barium enema, and colonoscopy are notable. Ultrasonography of the abdomen can distinguish between benign and malignant mucoceles,[4] and the "onion-skin sign," as described by Caspi *et al*., is specific for mucinous appendiceal lesions.[14,15] A multidetector CT scan is required to confirm the diagnosis and is the preferred radiological imaging. Appendiceal mucocele is confirmed by a low-to-mixed attenuated, well encapsulated, round or tubular cystic mass adjacent to the caecum.[16] Colonoscopy can be used to assess other colonic lesions and to diagnose synchronous or metachronous colonic cancers. Mucocele appendix is indicated by a mount-like elevation of the appendix orifice (volcano sign) and yellowish mucus discharge.[17,18] Due to the mass effect of a large mucocele,

barium contrast examination may reveal indentation or lateral displacement of the caecum.[19]

Because surgical resection is the preferred treatment for mucocele appendix, selecting an appropriate surgical method is critical. If the lesion is neoplastic, surgical removal of the mucocele appendix prevents future rupture and the development of pseudomyxoma peritonei. Both open surgery[20,21] and laparoscopic surgery[22,23] have yielded positive results in the literature. Survival after standard appendectomy for retention cysts, mucosal hyperplasia, or cystadenoma without perforation or spillage is excellent.[24] Despite the distinctive and specific colonoscopy and radiological features, mucocele appendix is frequently discovered incidentally during surgery.

We presented three cases of appendicular mucocele, of which one had a clear preoperative diagnosis, while the other two were not differentiated before surgery and were diagnosed intraoperatively. Our Case 1 had inflamed distended globular cystic mass of the appendix with a wide base and was subjected to right hemicolectomy. Histopathology of the specimen confirmed the diagnosis of mucinous cystadenoma carcinoma with negative resection margins and the patient was advised long-term follow-up. The other two cases were subjected to appendectomy, as the mucocele was not perforated and the base of the appendix was healthy.[9]

CONCLUSION

Appendiceal mucocele with acute presentation is a rare pathology that clinically resembles acute appendicitis. Preoperative detailed investigations to reach a definitive diagnosis are critical for adequate surgical resection and overall outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for their images and other clinical information to be reported in the Journal. The patients understand that their name and initials will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Peer review

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Conflicts of interest

There are no conflicts of interest.

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