Original Article

Mucocele of Appendix: A Rare Cause of Surgical Abdomen

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Abstract

Background: Appendiceal mucocele is a rare surgical emergency caused by intraluminal accumulation of mucoid material due to obstruction. Proper diagnosis and adequate surgical management are paramount to prevent the complications. Objectives: The aim of this study is to report on our management of appendiceal mucocele and highlighting the importance of proper pre-operative workup to reach a definitive diagnosis as a possible means of avoiding the associated grave complications. **Materials and Methods:** The patients of mucocele of appendix encountered over the period of five years in our department were managed and reported. **Results:** We present a series of three cases of appendiceal mucocele and included two females and one male patient with a median age of 41.67 years. Clinical examination, biochemical parameters, and imaging studies showed features of acute appendicitis. A 65-year-old female patient, presented with signs and symptoms of acute appendicitis was admitted and on exploratory laparotomy, showed inflamed globular cystic mass of appendix 10 cm × 6 cm × 4 cm, with dilated base and subjected to right hemicolectomy. A 35-year-old male patient was operated for appendicitis and diagnosed with mucocele appendix of 6 cm × 1 cm × 1 cm. Another patient, 25-year-old female was subjected to diagnostic laparoscopy in view of recurrent pain in right iliac fossa. A diagnosis of mucocele of appendix was made and removed via endobag. **Conclusion:** Appendiceal mucocele with acute presentation remains a rare diagnosis. Proper pre-operative workup to reach a definitive diagnosis is imperative for adequate surgical management to prevent the associated post-operative grave complications. Pre-operative diagnosis helps in decision making for the selection of the appropriate surgical procedure and alarms the operating surgeon for meticulous surgical dissection, adequate removal of mesoappendix with utmost cautious to prevent the mucus spillage into the peritoneal cavity.

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

Introduction

Appendiceal mucocele is a rare surgical emergency caused by the intraluminal accumulation of mucoid material due to obstruction by various neoplastic and nonneoplastic causes. The reported incidence of mucocele in appendectomy specimens is approximately 0.2%–0.7%.[1,2] The diagnosis of appendicitis and appendiceal mucocele may be difficult to differentiate and may sometimes coexist.[3,4] In 2012, the Peritoneal Surface Oncology Group International (PSOGI) classified the mucinous lesions of the appendix into nonneoplastic 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 the appendix are common, only a limited number of cases of mucocele appendix have appeared in the medical literature. We hereby report our experience of three cases of mucocele appendix and their management.

Materials and Methods

The patients of mucocele of appendix encountered over the period of five years in our department were managed and reported. The data of patients with diagnosis of mucocele appendix was collected from medical record department (MRD), operation theatre record files, and histopathological unit. The patients' medical record included demographic

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characteristics, clinical presentation, biochemical parameters, radiological, and histopathological data was collected. Further, the pre-operative workup, operative methods, post-operative status, and the conditions on follow-up was recorded, analyzed, and reported..

Case Details

Case 1

Mrs. X, a 65-year-old hypertensive woman, was referred from the community health center to our accident and emergency unit with the presentation of a 1-day history of pain right iliac fossa and a few bouts of vomiting. Her appetite was good with no appreciable changes in her weight. Detailed surgical, medical, obstetric, personal, and family history were recorded, which were not significant. The patient was thoroughly examined, resuscitated, and subjected to standard baseline investigation [Table 1]. Leukocytosis (16.3×10^9 /L) was notable from laboratory test results. On physical and systemic examination, her pulse rate was 92 bpm, tongue was dry, blood pressure (BP) was 120/70 mm Hg, respiratory rate was18 breaths/min, temperature was 99.8°F, and saturation was 96% at room air. On palpation, the abdomen was nondistended; minor tenderness was noted in the right iliac fossa. The digital rectal examination was normal and finger-stained with normal color stools. As a first radiological investigation, ultrasonography abdomen/pelvis was performed and showed a cystic mass measuring 6.5 cm × 5.5 cm arising from the appendix with features suggestive of mucocele appendix. Computed tomography (CT) scan of abdomen and pelvis with intravenous contrast revealed a cystic mass with tubular fluid density arising from the caecum and adherent to adjacent small gut loops suggestive of mucocele appendix. No enlarged mesenteric lymph nodes were seen. The liver was normal in size and texture with no evidence of metastatic deposits. Following informed written consent, the patient was decided and taken to emergency surgical exploration.

An inflamed globular cystic mass of the appendix measuring 10 cm × 6 cm × 4 cm arising from the mid appendix with the 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 uneventful and the patient got discharged on the fourth postoperative day in a good condition and attached to our outpatient department for follow-up. Histopathological examination of the specimen confirmed the mucinous cystadenoma carcinoma with free margins of resection. The patient is doing well on regular follow-up of 3-year period.

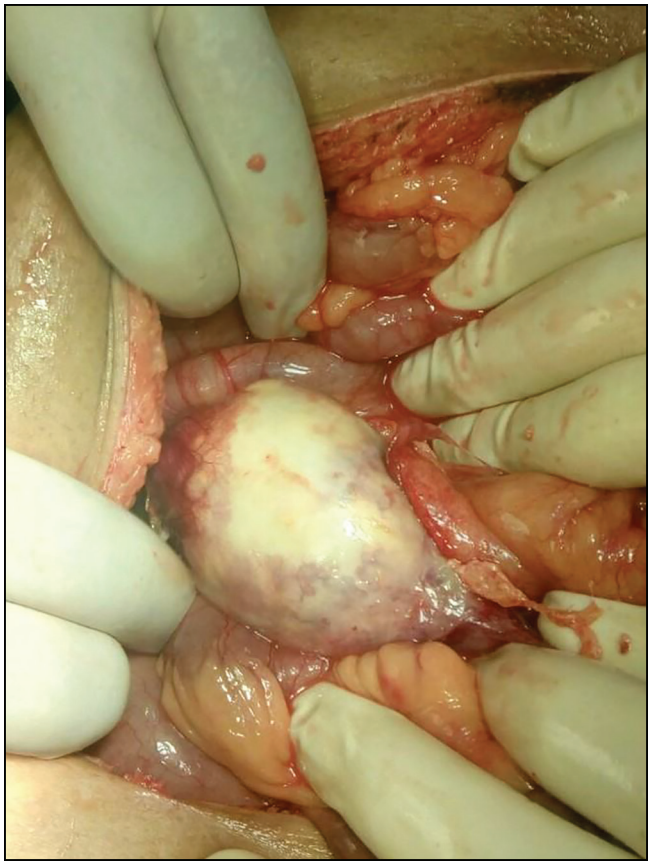

Figure 1: Operative photograph of case 1

Table 1: Biochemical–hematological parameters

Parameter	Case 1 results	Case 2 results	Case 3 results
Hb (g/dL)	11	13	11.4
TLC (cm/mm^3)	16.3	13.1	6.5
Neutrophiles/lymphocytes (%)	62/41	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)	101	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

AST = aspartate transaminase, ALT = alanine transaminase, ALP = alkaline phosphatase, PLT = platelet count, TLC = total leucocyte count

Case 2

Mr. Y, a 32-year-old man with no underlying medical comorbidity, presented with a 1-day history of pain right lower quadrant. The pain was insidious onset, migratory, and progressive in nature associated with nausea, vomiting, low-grade fever, and decreased appetite. The patient reported history of typical Murphy's triad of central abdominal pain followed by nausea and vomiting with the migration of the pain to the right iliac fossa. His investigations showed normal blood glucose, liver and renal functions, and blood picture showed neutrophilic leukocytosis [Table 1]. Clinically, patient was hemodynamically stable. Abdominal examination revealed localized tenderness in the right iliac fossa, percussion tenderness, guarding, and rebound tenderness. The findings of digital rectal examination were unremarkable. Ultrasonography abdomen showed elongated cystic, dilated, aperistaltic, and noncompressible gut loop in right lower quadrant with internal echoes. No interloop fluid or mesenteric lymph nodes were recorded.

Due to high clinical and radiological suspicion of acute appendicitis, the patient was taken for emergency appendectomy after proper informed consent. The intra-operative findings were suggestive of mucocele of appendix 6 cm × 1 cm × 1 cm and appendectomy was performed [Figure 2]. 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 is doing well on regular follow-up for 1 year of surgery.

Case 3

A 35-year-old female patient, P1L1, no underlying medical comorbidity, had a history of multiple emergency room admissions of pain right lower abdomen in the past 1 year. She reported no changes in her eating, bowel habits, and no history of bleeding per rectum or melena. The patient had no history of any significant past surgical or medical disease. The patient was thoroughly evaluated and subjected to the required investigation. Physical and clinical examination, digital rectal, pelvic and proctoscopy examination, and biochemical parameters were unremarkable except for mild tenderness over McBurney's point [Table 1]. Ultrasonography showed enhancing echogenic mass of 3 cm × 2 cm × 1 cm arising from the right adnexa. Contrast-enhanced CT scan showed low-attenuation mass arising from the appendix in right lower quadrant without any periappendiceal fluid, inflammation, and abscess. Following the informed written consent, the patient was listed for diagnostic laparoscopy. On diagnostic laparoscopy, cystic mass arising from mid appendix with inflamed wall and without perforation was discovered in right iliac fossa. Caecum, terminal ileum loops, liver, and bilateral adnexa were grossly normal. No discharge or peritoneal fluid was noted on laparoscopy. The patient was subjected to appendectomy and specimen retrieved via endobag [Figure 3]. The patient did well in postoperative period and was discharged on the third postoperative day. Histopathology showed mucinous cystadenoma and resection margins were free. The patient is doing well on regular follow-up and is satisfied.

Discussion

Mucocele appendix is the macroscopic appearance of an appendix distended with mucus due to obstruction by various neoplastic and nonneoplastic causes. It is uncommon with a reported incidence of 0.2%–0.7%,[1,2] four times more common in women usually detected in patients over 50 years of age.[5] The typical clinical manifestations are nonspecific and may resemble acute

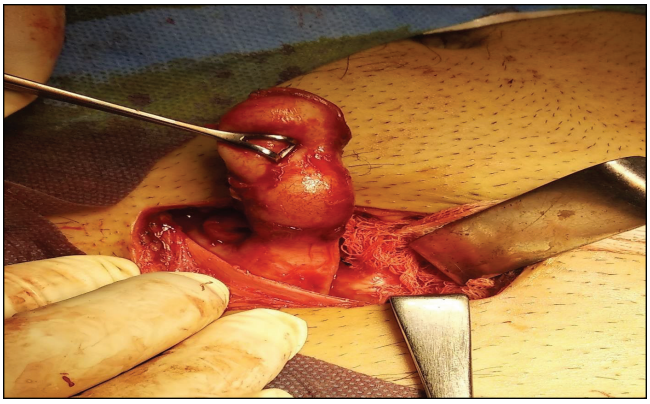

Figure 2: Operative photograph of case 2

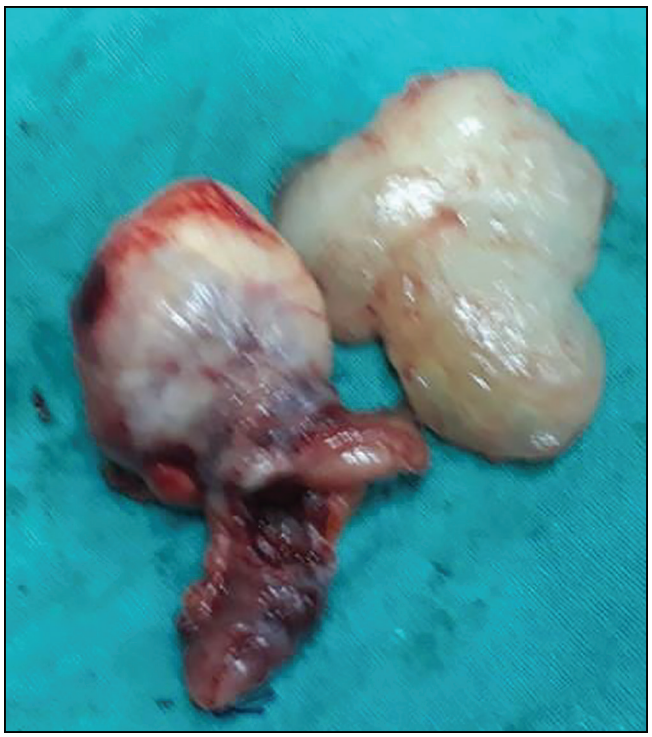

Figure 3: Operative photograph of case 3

appendicitis. Others may have palpable right lower quadrant mass, intermittent colicky pain, diarrhea, and bleeding per rectum. The urinary symptoms are rare, and palpable per abdomen mass is the most common presentation and has been reported in 50% of cases.[6-8] Some patient remains asymptomatic and approximately 50% may be diagnosed accidentally during evaluation for some other disease or at the surgery.[6] Some patients may present with complications such as intestinal obstruction, bleeding, malena, pyonephrosis, and pseudomyxoma peritonei due to spontaneous rupture.[8]

Four histopathological subtypes of mucocele of the appendix reported are a simple mucocele, focal or diffuse mucosal hyperplasia, mucinous cyst-adenoma, and mucinous cystadenocarcinoma.[9,10] Proper preoperative workup to reach a definitive diagnosis is imperative for adequate surgical management to prevent the associated intraoperative and postoperative grave complications and repeated surgery. Preoperative diagnosis helps in decision-making for the selection of the appropriate surgical procedure and alarms the operating surgeon for meticulous surgical dissection with utmost caution to prevent the mucus spillage into the peritoneal cavity. Imaging studies including ultrasonography, contrast-enhanced CT, barium enema, and colonoscopy are notable preoperative diagnostic modalities. Ultrasonographic examination of abdomen can differentiate benign and malignant mucoceles,[4] and "onion-skin sign" as described by Caspi *et al.* is specific for mucinous appendiceal lesion.[11,12] Multidetector CT scan is important to confirm the diagnosis and is the radiological imaging of choice. A low to mixed attenuated, well-encapsulated round or tubular cystic mass adjacent to the caecum provides strong evidence of appendiceal mucocele.[13] Colonoscopy can be used to evaluate other colonic lesions and for the diagnosis of synchronous or metachronous colonic cancers. The mount-like elevation of appendix orifice (valcano sign) and yellowish mucus discharge[14,15] on colonoscopy is suggestive of mucocele appendix. Barium contrast examination may show indentation or lateral displacement of the caecum due to the mass effect of large mucocele.[16] Despite the characteristic and specific colonoscopy and radiological features, the mucocele appendix is frequently an incidental finding during surgery.

The surgery is the only curative and the treatment of choice for the mucocele appendix; therefore, the selection of an adequate surgical method is very important. Both the open surgery[17,18] and laparoscopic method[19,20] have been reported in the literature with good results. Dhage-Ivatury and Sugarbaker designed an algorithm for deciding the surgery to be undertaken ranging from simple appendectomy to right hemicolectomy, cytoreductive surgery, and intraperitoneal chemotherapy based on the various intra-operative factors.[17] Simple appendectomy remains the procedure of choice in patients with benign mucocele with normal caecum and appendicular base. Patients with ruptured mucocele, involvement of caecum, ileum, and surrounded mesenteric lymph nodes, and malignant cytology should be subjected to the right hemicolectomy.

Survival is excellent after standard appendectomy of retention cysts, mucosal hyperplasia, or cystadenoma without perforation or spillage during the surgery.[21] Patients with neoplastic lesions, particularly high-grade appendiceal mucinous neoplasms, and mucinous adenocarcinomas are associated with poor prognosis.

We present the three cases of appendicular mucocele: only one patient had a clear-cut preoperative diagnosis and the rest of the two cases were not differentiated before surgery and were diagnosed intraoperatively. Preoperative diagnosis helps in decision-making for the selection of the appropriate surgical procedure and alarms the operating surgeon with utmost caution to prevent the mucus spillage into the peritoneal cavity. Proper preoperative radiological diagnostic tools and postoperative histopathological examination of specimen are needed for the definitive diagnosis.

Conclusion

Appendiceal mucocele with acute presentation remains a rare diagnosis. Proper preoperative workup to reach a definitive diagnosis is imperative for adequate surgical management to prevent the associated intraoperative and postoperative grave complications and repeated surgery. Preoperative diagnosis helps in decision-making for the selection of the appropriate surgical procedure and alarms the operating surgeon for meticulous surgical dissection and adequate removal of mesoappendix with utmost caution to prevent the mucus spillage into the peritoneal cavity.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Ethical approval

The study was conducted in accordance with the ethical principles that have their origin in the Declaration of Helsinki. It was carried out with patients verbal and analytical approval before sample was taken. The study protocol and the subject information and consent form were reviewed and approved by a local ethics committee according to the document number 1118 (including the number and the date in 28/4/2020) to get this approval.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have

given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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