Case Report

Giant mucocele of the appendix – laparoscopic management: A case report and review of the literature

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ABSTRACT

A mucocele is an uncommon lesion of the appendix characterized by cystic distension of the appendicular lumen due to the accumulation of a mucoid substance known as mucin. It is often asymptomatic and surgical treatment depends on the histology and the dimension of the mucocele. We, herein, report a laparoscopically managed case of a 55-year-old male who presented with a large mobile mass in the lower abdomen. Abdominal contrast-enhanced computed tomography revealed a large mucocele of the appendix.

Key words: Appendix, Laparoscopy, Mucinous cystadenoma, Mucocele, Pseudomyxoma peritonei

Mucocele of the appendix is a rare entity, as it accounts for only 0.2–0.3% of all surgical specimens of the appendix. It is more frequently observed in females (4:1) and those who are in their fourth-fifth decade of life [1]. It can have either inflammatory or neoplastic etiology. The latter poses a great threat of resulting in a clinical condition called pseudomyxoma peritonei (PMP), an entity caused by either spontaneous or iatrogenic rupture of the mucocele. It results in the spread of malignant cells throughout the peritoneal cavity and is associated with high mortality and morbidity [2,3]. Contrast-enhanced computed tomography (CECT) of the abdomen is the diagnostic tool of choice for mucocele of the appendix. Surgical resection of the mucocele is the treatment of this condition. Here, we report the case of a giant mucocele of the appendix and discuss the diagnostic aspects, surgical options, and prognosis.

CASE REPORT

A 55-year-old male presented with complaints of dull, non-radiating lower abdominal pain and a large lump in his lower abdomen for 15 days. He did not give any history of fever, vomiting, altered bowel habits, and loss of appetite or weight.

On examination, his pulse rate was 84/min, respiratory rate was 14/min, and body temperature was 97.8 degrees Fahrenheit with a blood pressure of 110/80 mmHg. A per abdominal examination revealed a large, non-tender, well-defined, mobile lump with a smooth surface and firm consistency extending horizontally from the right iliac fossa to beyond the midline. Hernial orifices were normal. There was no hepatosplenomegaly.

CECT abdomen showed a large, oval, well-defined mass measuring 15.2 cm×7.5 cm in the right iliac fossa, and medial to the cecum. It had a moderately thickened wall (9 mm) with minimal post-contrast-enhancement. CT attenuation value of its contents was 17HU, most likely representing a mucocele of the appendix (Fig. 1). The laboratory test results were as follows: Hemoglobin – 12.4 mg%, white blood cells – 10060, platelet counts – 1.5 lakhs, serum creatinine – 0.60, sodium – 139.00, potassium – 3.54, chloride – 107.00, total bilirubin – 0.6 mg/dl, direct bilirubin – 0.2 mg/dl, serum glutamic-oxaloacetic transaminase – 28 IU/dl, serum glutamic-pyruvic transaminase – 22IU/dl, alkaline phosphatase – 56 U/dl, total protein – 6.5 g/dl, albumin – 4.0gm/dl, carcinoembryonic antigen (CEA) – <0.50 ng/ml (normal range:0–5.0), and CA 19–9:6u/ml (normal range: 0–37.0). His chest X-ray was normal.

The patient was then planned for surgery. At laparoscopy, he was found to have a huge greyish-white appendiceal mass, extending from the right iliac fossa to just beyond the midline. The ileum was adherent to its base. Meticulous adhesiolysis was done. The base of the appendix could not be bared and identified distinctively and was invaginating into the cecum. There were no obvious enlarged lymph nodes, no free fluid/mucin, and the visualized surface of the liver appeared normal. Partial typhlectomy with excision of the mass was performed using 60 mm blue cartridge loaded on an Endo GIA stapler (Fig. 2). The resected specimen was extracted in a plastic bag through the widened hypogastric trocar site (Fig. 3). There was no breach

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in the wall of the specimen and spillage of its contents during dissection and retrieval.

The specimen was then sent for histopathological examination (HPE). The HPE report on gross examination revealed a specimen of appendiceal mass with a part of the cecum measuring 17 cm in length and 8 cm in maximum diameter. The appendix was markedly dilated, and the wall was thickened, fibrotic, and the lumen contained mucinous material. The mucosa was predominantly ulcerated and shaggy at places. The part of the cecum measured 3.5 cm × 1 cm, the cecal resection margin began 1 cm away from the mass and free grossly. Microscopic examination revealed a benign appendiceal mucocele. The appendiceal wall was thickened and fibrotic with chronic inflammatory infiltrate and was lined by a single layer of intact to ulcerated mucinous epithelium. Mucinous dissection into the appendiceal wall was not present. The resection margin was free of mucinous epithelium, and there was no evidence of dysplasia or malignancy (Fig. 4).

The patient had an uneventful post-operative recovery and was discharged on post-operative day 6. The patient was followed up on day 10 and 1 month after discharge for a wound check and any new complaints, respectively. He was not advised any additional investigations in view of the HPE report. For the purpose of this study, he was interviewed telephonically at the time of writing this paper. He remains asymptomatic, as of the present day, 9.5 months after his surgery.

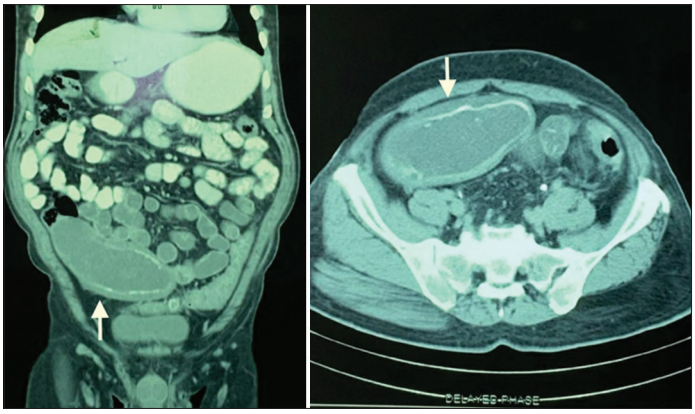

Figure 1: Abdominal contrast-enhanced computed tomography scan showing large mucocele of appendix (left: coronal section and right: axial section)

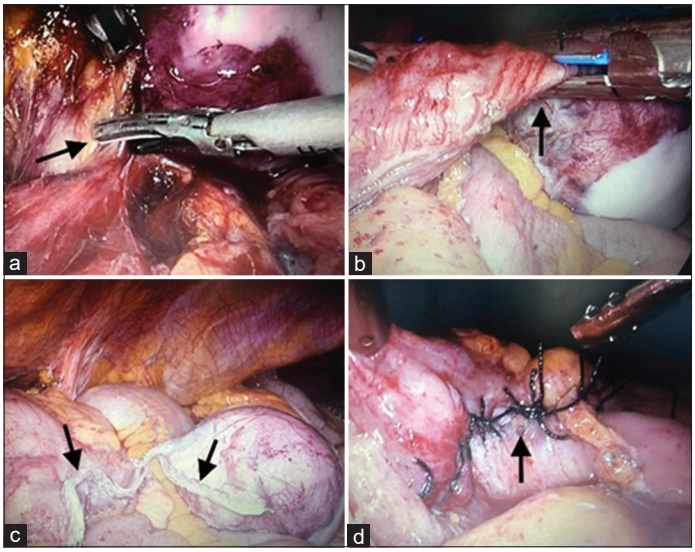

Figure 2: Intraoperative images showing (a) dissection at the base to free the adhesions; (b) stapler being used to resect the specimen; (c) staple lines after resection; and (d) over suturing of staple line

DISCUSSION

Mucocele of the appendix is described as a dilatation of the appendicular lumen due to the accumulation of mucinous secretions within it. The most common clinical manifestation of it is acute or chronic pain in the right iliac fossa, sometimes associated with a lump on physical examination, in about 50% of patients. However, the uncommon presentations can also be lower

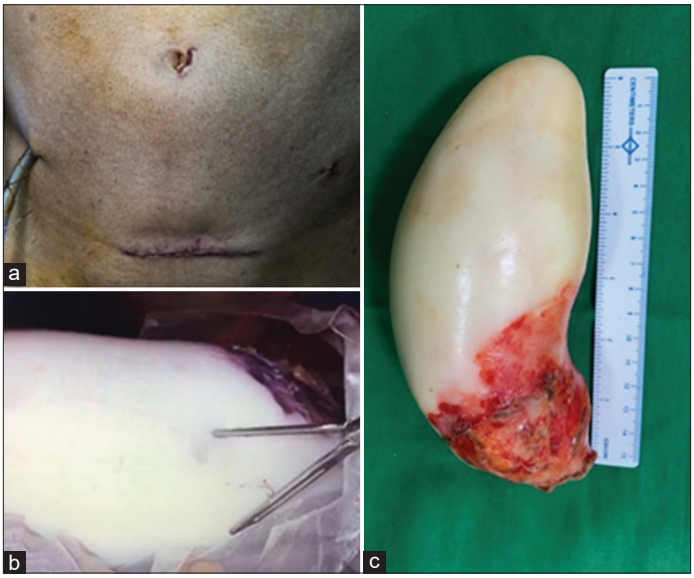

Figure 3: (a) Widened hypogastric trocar site for retrieval of the specimen; (b) Extraction of the specimen in a plastic bag; (c) Gross specimen of mucocele of appendix with partly resected cecum

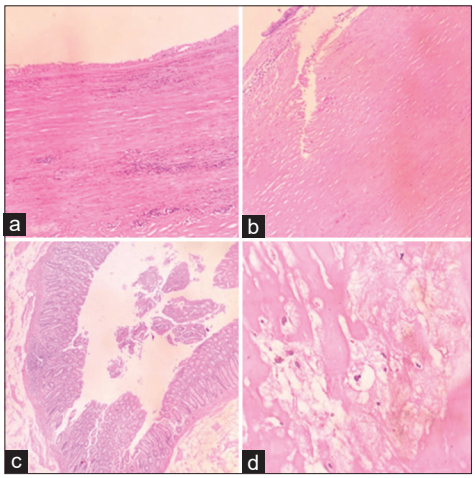

Figure 4: Photomicrographs showing (a) thickened, fibrotic appendiceal wall with denuded lining; (b) thickened, fibrotic appendiceal wall with focally preserved lining; (c) cecal resection margin showing unremarkable cecal mucosa; and (d) mucin within the appendicular lumen

gastrointestinal bleeding, intussusception, intestinal obstruction, genitourinary symptoms, sepsis, or fistula [4].

Based on histology, the World Health Organization (WHO) classifies it into four subgroups – (A) simple or retention mucocele has normal epithelium and mild dilation up to 1 cm due to appendicular outflow obstruction, more often due to fecolith; (B) mucocele with hyperplastic epithelium: This constitutes about 5–25% of all mucoceles; and (C) benign mucocele: The most common form of this is mucinous cystadenoma; also known as low-grade appendiceal mucinous neoplasm, which exhibits epithelial atypia with moderate distension up to 6 cm. Benign mucoceles constitute about 63-84% of cases. Histological examination of the mucus does not reveal any neoplastic cells; (D) malignant mucinous cystadenocarcinomas: These account for 11–20% of cases. Its feature is severe appendiceal distension, glandular stromal invasion and/or epithelial cell implants in the peritoneum [1-3,5]. Some tumor marker levels (CEA, CA 19–9, CA 125, CA 15–3 and CA 72–4) may also be elevated [6,7]. Our patient belonged to Group C – benign mucocele.

Mucinous cystadenoma presents with perforation of the appendix in 20% of cases, while mucinous cystadenocarcinoma may present with spontaneous rupture in 6% of cases [8]. Rupture of the mucocele can have a serious prognostic implication, regardless of whether it is benign or malignant and can result in PMP. In benign mucocele, it is confined to the periappendicular area, while in malignant cases, it is a metastatic entity. Retroperitoneal and pleural implants have also been reported [6,7]. Aggressive surgical interventions may be needed to manage PMP such as extirpation of mucinous material, debulking, peritonectomy, and heated intraperitoneal chemotherapy [9]. Five-year survival rates in the case of benign and malignant mucocele rupture are 91–100% and 25%, respectively [10].

Because of the non-specific nature of the disease, accurate pre-operative diagnosis is often difficult. An acutely inflamed and severely distended appendix can often mimic a mucocele on basic imaging modalities like an ultrasound scan of the abdomen. CECT scan of the abdomen typically shows a round, low density, thin-walled, encapsulated mass communicating with the cecum. This is diagnostic. It also helps in the evaluation of the extent of the mucocele [5].

Open surgical resection has been recommended traditionally for the treatment of appendiceal mucocele. However, laparoscopic resection is advocated by some due to its obvious benefits [11]. The mode of surgery should be decided based on the expertise and facilities available. In our case, great care was taken to avoid iatrogenic rupture of the mucocele and the operative specimen was retrieved, intact, in a plastic bag. Appendectomy alone is the definitive management for intact and benign mucocele. The frozen section may be kept on standby for such cases to opine on the resection margins of the specimen and the status of the lymph nodes. If the resection margins of the specimen are involved (as shown on the frozen section or the final HPE report), partial cecectomy, ileo-cecectomy, or right hemicolectomy may have to be performed, with the ultimate goal of achieving clear resection margins [12].

Lymph node metastasis secondary to mucinous appendiceal neoplasm is rare and accounts for 4.2% of patients with mucinous malignancy [13]. Mucocele of the appendix also has an association with other intra-abdominal neoplasms, especially carcinoma of the colon (13-42%) and the tumors of the ovary [14]. Patients with the WHO type D mucoceles should be enrolled in a systematic surveillance program and followed up with serial CECT scans and monitoring of tumor marker levels (CEA, CA 19-9, CA 125, CA 15-3, and CA 72-4) for 5–10 years, for early pick up of possible recurrent disease.

We were fortunate to be working with a pre-operative diagnosis of mucocele of the appendix, given the sheer size of our specimen and the fact that CECT abdomen was done pre-operatively for the same. A review of the literature was done for 13 studies [4,5,9,15-18, 20-24] and we found that ours is one of the largest reported size of mucocele of the appendix which was successfully managed laparoscopically (Table 1).

Table 1: Review of the literature on mucocele of the appendix

S. No	Authors (year)	Size of the mucocele	Mode of surgery
1	Rampone *et al* [5]	17 cm×4 cm	Open (Appendectomy)
2	Motlaleselelo *et al* [9]	17 cm×5 cm	Open (Right hemicolectomy)
3	Orcutt *et al*. [15]	Case 1:9.5 cm ×3.7 cm Case 2: 2.2 cm×2 cm	Laparoscopy (Partial typhlectomy) Laparoscopy (Partial typhlectomy)
4	Sertkaya *et al*. [16]	14 cm×5 cm×4 cm	Open (Partial typhlectomy)
5	Park *et al*. [17]	7.9 (range 3–20) cm × 3.2 (range 1-7.5) cm	Laparoscopy (Mix of Appendectomy, Partial typhlectomy and Right hemicolectomy)
6	Idris *et al*. [18]	14 cm×5 cm×3 cm	Open (Appendectomy)
7	Rojnoveanu *et al*. [4]	8.4 cm×4 cm	Open (Appendectomy)
8	Singh *et al*. [19]	14 cm×15 cm	Laparoscopy (Appendectomy)
9	Demetrashvili *et al*. [20]	7 cm×4 cm×3 cm	Open (Appendectomy)
10	Ju *et al*. [21]	Case 1: 14 cm Case 2: 15 cm	Laparoscopy (Partial typhlectomy) Laparoscopy (Partial typhlectomy)
11	El Ajmi *et al*., [22]	13 cm×5.5 cm	Open (Stumpectomy with excision of the mass)
12	Palanivelu *et al*. [23]	10 cm×6 cm	Laparoscopy (Rt hemicolectomy)
13	Korkolis *et al*. [24]	8 cm×5.5 cm	Open (Stumpectomy with excision of the mass)

CONCLUSION

Mucocele of the appendix closely mimics appendiceal distension caused by appendicitis, on radiological investigations. Accurate pre-operative diagnosis is therefore rare, especially when the mucocele is not very large, as appendicitis is a far commoner clinical condition. We believe that even in those majority situations, wherein, during a routine laparoscopic appendectomy for appendicitis, one is not pre-operatively aware of the presence of a mucocele, but is faced with a turgid and/or cystic distension of appendix intraoperatively, one should have a high index of suspicion for mucocele and then take utmost care while handling and retrieving the specimen. We believe that, in such situations, the specimen should be compulsorily retrieved in a retrieval bag after adequately widening the concerned trocar site. When pre-operatively aware of the presence of mucocele, operating surgeons would naturally take more precautions not only during the intraoperative handling of the appendix but also during specimen retrieval so as to prevent spillage of contents and possible PMP. This case report underscores the fact that laparoscopy is a feasible option even while dealing with giant mucoceles of the appendix provided, there is no compromise with the basic principles of it's surgical removal.

AUTHORS' CONTRIBUTIONS

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