Giant Mucocele Appendix: A Hybrid Laparoscopic Approach

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ABSTRACT

Appendicular mucocele is a rare but well-described clinicopathological diagnosis. It denotes dilatation of the appendix due to luminal obstruction by mucinous secretions. We report a case of a 71-year-old male patient with chronic kidney disease on regular hemodialysis with lump in right iliac fossa since 4 months with no history of pain abdomen. Ultrasonography abdomen followed by contrast enhanced computed tomography of abdomen suggested a diagnosis of mucocele appendix. Patient was taken up for laparoscopic appendectomy which revealed appendix that was hugely dilated with wide base impinging into the cecal wall. In order to prevent intraoperative rupture of mucocele and keeping malignancy as a differential, procedure was converted into open and cecum was repaired at the site of wide appendicular base. Surgery is the definitive treatment for mucocele and laparoscopic assist provides precision of dissection and minimizes injury to surrounding viscera. The aim of this publication is to demonstrate that laparoscopic surgery is as safe as traditional open approach. Also, surgeon should never hesitate to convert the procedure to open if there is risk of peritoneal spillage and doubt of malignancy in mind.

Keywords: Appendix, mucocele, malignancy, hemodialysis

Rokitansky was the first one to describe mucocele of appendix in 1842. It is an obstructive distention of the appendix due to mucoid secretions in the lumen, which can be either neoplastic or non-neoplastic. It is a rare entity with an incidence of 0.2-0.3% of all appendectomies and 8-10% of all appendiceal tumors. The clinical presentation ranges from asymptomatic to appendicitis like symptoms.

It can rarely present as intestinal obstruction. Four types of appendiceal mucocele have been explained on the basis of cause of obstruction: retention cysts, epithelial hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma. Appendiceal mucocele can be either benign or malignant. A preoperative diagnosis is crucial in order to choose the correct operative management. The correct surgical management depends on size and location of lesion.

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Laparotomy is the traditionally recommended approach, but minimally invasive surgical approach seems to be as safe as open surgery.

CASE REPORT

A 71-year-old male patient admitted to dialysis unit of our hospital with diagnosis of chronic kidney disease, on routine hemodialysis, complained of lump in right iliac fossa since 4 months with no history of pain abdomen. Abdominal examination revealed a nontender cystic mobile mass around 5 × 6 cm with smooth surface and well-defined margins in the right iliac fossa without any guarding or rigidity. Digital rectal examination was normal. Patient's blood investigations were within normal limits except renal functions.

Ultrasonography (USG) abdomen was suggestive of cystic mass in right iliac fossa followed by abdominal computed tomography (CT) scan which revealed a large well-defined, tubular-shaped hypodense lesion of near fluid attenuation seen in right iliac fossa posterior to cecum. Appendix could not be seen separately. It was extending superiorly along the right pararenal space. The lesion showed saccular dilatation in its mid part with peripheral calcification of its walls. It measured 5.7 cm in diameter and 11.7 cm in length. The radiologist gave differential diagnosis of mucocele appendix, cystic lymphangioma, hydatid cyst, cystic mesothelioma Figure 1 shows CECT abdomen of patient showing mucocele appendix.

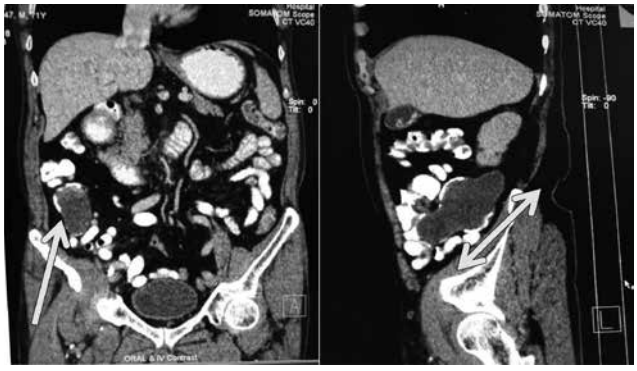

Figure 1. CECT abdomen of patient showing mucocele appendix.

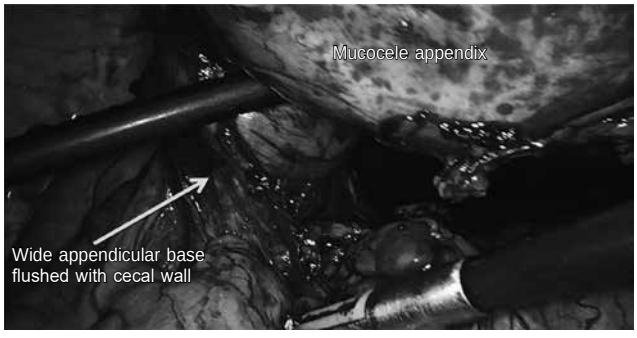

Figure 2. Laparoscopic view of mucocele appendix with wide base flushed with cecal wall.

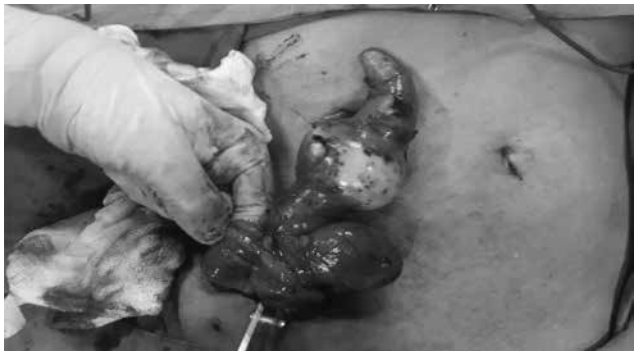

Figure 3. Mucocele appendix safely delivered out after giving skin incision.

Following informed consent, the patient was taken up for laparoscopic appendectomy after a cycle of hemodialysis. There was no evidence of peritoneal tumors, seedling or metastases. A grossly enlarged retrocecal appendix 15 cm long and 5 cm in diameter in the middle was found. Appendix was finger-like in distal part and ball-like in the middle with a wide base around 3 cm merging into the cecum (Fig. 2). Mesoappendix was swollen. The mesoappendix was coagulated and cut using bipolar cautery.

Since the appendix had larger diameter and in order to prevent iatrogenic rupture of mucocele and retrieve the specimen intact, procedure was converted into open. Port incision in right iliac fossa was widened and specimen delivered out as shown in Fig. 3. Appendicular base was held with intestinal clamp and appendix was cut. The mucocele had a lumen separate from cecal lumen (Fig. 4). Cecum was repaired in 2 layers using vicryl because appendix had a wide base of approximately 3 cm.

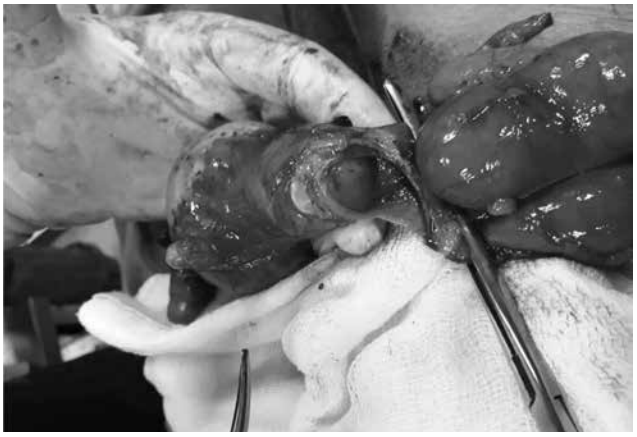

Figure 4. Mucocele with a lumen separate from cecum.

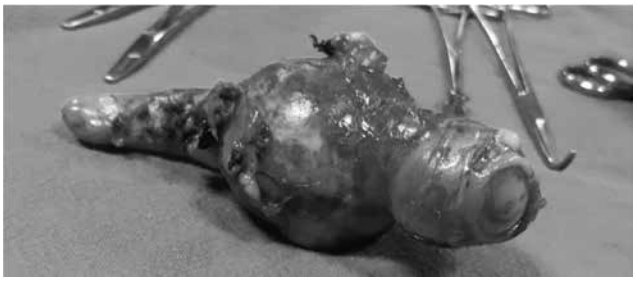

Figure 5. Gross specimen of mucocele appendix.

Postoperatively patient was started on oral liquids next day and the patient was discharged on Day 5 postoperatively in a good condition.

Grossly appendix measured 13 × 6 × 5 cm (Fig. 5), outer surface congested and cut surface showed dilated lumen filled with mucoid material with wall thickness 0.1-0.2 cm. Histopathological examination confirmed the final diagnosis of a benign mucocele appendix arising from the body of the appendix with free margins of resection. The patient remained well on regular follow-up visits over 2 months.

DISCUSSION

Clinical presentation of appendicular mucoceles is usually vague and furthermore, it can be asymptomatic in 25% of patients. Most commonly, patients present with right lower quadrant pain. Palpable masses have been reported in 50% of cases as seen in our patient. USG and CECT abdomen are most helpful in making

preoperative diagnosis. It helps in planning the choice of procedure and avoids complications. Appendicular mucocele can be benign or malignant and the World Health Organization (WHO) classifies them into four histological types: retention cysts, epithelial hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma. Mucinous cystadenoma is the most common of the four types. Size is an important factor to consider when dealing with appendiceal mucocele. An appendiceal mucocele that is <2 cm is rarely malignant and those >6 cm are more often associated with cystadenoma and cystadenocarcinoma and a higher rate of perforation. Rupture of either benign or malignant types is associated with pseudomyxoma peritonei, which is associated with a higher morbidity and mortality. Benign appendiceal mucocele has a 91-100% 5-year survival rate, while malignant forms have a 5-year survival rate of 25%. Historically, open surgery was an established procedure but with the advent of minimally invasive surgery, laparoscopic appendectomy has become the gold standard procedure. However, in our case, after total laparoscopic dissection and releasing the mucocele up to the base, we had converted the procedure into open due to its wide base burying into cecum and to retrieve the specimen intact a wide incision was needed. The procedure could have been completed laparoscopically with the help of endostaplers but this would also have required a wide incision to deliver the specimen out. Moreover, it would have added to the cost of procedure with no added advantage. That's why a midway path was adapted by completing the dissection laparoscopically and repairing the cecal defect with standard two-layer technique. Combining both the techniques helped us to cut the procedure cost, deliver the intact specimen out and at the same time avail advantages of minimally invasive surgery. Careful consideration should be given to minimize rupture of the appendiceal mucocele when making a decision on approach of choice. Evidence suggests that appendectomy is curative for benign, grossly intact mucoceles.

CONCLUSION

Appendiceal mucocele is a rare condition. The clinical presentation is often non-specific and the clinician should have appendiceal mucocele in mind in patients presenting with long-term right lower quadrant pain, adnexal masses and acute appendicitis picture. Often, the diagnosis is made incidentally during imaging or surgical procedure. Radiological imaging and careful analysis is critical in planning management. Surgical resection is potentially curative and rupture of the mucocele should be avoided as it may lead to pseudomyxoma peritonei, a condition with high morbidity and mortality. The aim of this publication is to show that hybrid approach with laparoscopic assisted open appendectomy helps in maintaining safety, feasibility as well as cuts the cost of procedure.

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