

Case Report

Double trouble: a rare presentation of mucocele of appendix

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ABSTRACT

Appendix is a blind ending tubular structure present at the convergence of the three tenia coli the base of the caecum. Most commonly presented to the out-patient department with features of appendicitis, but mucocele of the appendix secondary to malignant etiology is a rare clinical finding, more than half of the patients are asymptomatic in their clinical presentation. An accurate diagnosis and proper treatment protocols are necessary for its management. If mucocele of appendix is treated incorrectly may lead pseudomyxoma peritonei. A case of a 54-year-old man with right lumbar pain, duration of 1 week, clinical findings and ultrasonography (USG) abdomen reported as ill-defined collection in front of the psoas muscle (psoas abscess). Hence contrast-enhanced computed tomography (CECT) abdomen and pelvis was performed and was reported as features suggestive of mucocele of appendix and a right proximal ureteric calculus (8 mm) with hydronephrosis. After obtaining consent, patient underwent open appendectomy with right ureteroscopic lithotripsy (URSL) + double J stenting. Post-operative period was uneventful but histopathology of the specimen revealed a malignant origin. In patients presenting with a right sided abdominal pain, a broad spectrum of differential diagnosis should be considered and evaluated accordingly.

Keywords: Mucocele, Appendix, Mucinous cystadenoma, Appendicitis, Pseudomyxoma peritonei

INTRODUCTION

Appendiceal mucocele occurs as a result of obstruction and dilatation of the appendiceal lumen with mucoid material. Mucocele of the appendix is a rare clinical finding. The incidence is 0.2% to 0.7% in all appendix specimens.^{1,2} Mucocele can occur due to many pathology and one being a rare occurrence secondary to mucinous cystadenoma, hence it is important to differentiate acute appendicitis and mucocele of appendix before surgery and select adequate surgical tactics and post-operative patient management.

If treated improperly, the mucocele may progress or may rupture which causes spillage of the malignant cells into the peritoneal cavity, leading to pseudomyxoma peritonei.^{3,4}

CASE REPORT

A 54-year male with no co-morbidities, presented with complains of abdominal pain of 1-week duration which was localised to the right lower abdomen, non-radiating, colicky pain, with no history of vomiting, fever, burning micturition. On examination his vitals and general condition was stable, per abdominal examination, tenderness was noted in the right lumbar and iliac fossa, no guarding or rigidity, no palpable mass or organomegaly. Routine investigations showed no leucocytosis and renal function test showed a derangement in serum creatinine levels (1.75 mg/dl) and a deranged glycated haemoglobin A1c (HbA1c) (7.7%). Initially in the out-patient department he underwent ultrasound abdomen and pelvis which was reported as a well-defined collection noted with debris in the right lumbar and right iliac fossa region, anterior to psoas muscle (psoas abscess), therefore a contrast-enhanced computed tomography

(CECT) abdomen and pelvis was done and reported as mucocele of appendix measuring 7×4 cm, with bilateral renal calculi and right upper ureteric calculi with hydronephrosis measuring 8 mm. After explaining the diagnosis and obtaining consent he underwent open appendectomy and right ureteroscopic lithotripsy (URSL) + double J stenting under spinal anaesthesia. Intra operative findings were, a well-defined tubular cystic seen arising from the 2 cm above the base of the cecum measuring 7×4 cm, adhesions were present around the appendix and was directed to the sub hepatic region, mucoid content was present within the lumen of the appendix, no contamination was present, base of the caecum was not involved, hence only a routine appendectomy was done.



Figure 1: Appendix identified and dissected.



Figure 2: Base of appendix identified.



Figure 3: Appendix specimen.

Appendicular base was transfixed and ligated with polyglactin 910 Trusynth sutures and skin closed with polyamide Trulon sutures. Histopathology reported as features suggested of low grade appendiceal mucinous neoplasm extending beyond the muscularis propria and invading into subserosal tissue.

DISCUSSION

Mucocele of the appendix refers to an appendix distended by mucus. It was first described by Rokitansky.⁵ The epithelium the appendicular lumen contains more goblet cells than the rest of the colon. Hence, most of the epithelial tumours arising from the appendix are mucinous and start as mucocele.⁶ But endometriosis or carcinoid tumour are some of the other conditions which can lead to mucocele in the appendix. The overall occurrence of appendiceal mucocele on histopathology examination of appendix specimen is about 0.2–0.3%. Mucocele of the appendix are classified into 4 histologic types: retention cyst, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma.^{7,8}

Mucocele of appendix are asymptomatic in a majority of cases and is diagnosed during radiologic and endoscopic examinations or intra operatively. But in few cases, patients present with clinical symptoms like pain in the right iliac fossa, nausea, vomiting, palpable abdominal mass and intussusception.^{9,10}

Early preoperative diagnosis and selection of appropriate surgical method is the key in management of mucocele of the appendix to prevent peritoneal dissemination and further complications in the postoperative period.^{9,11} Ultrasonography, computed tomography (CT), and colonoscopy are the most commonly used for diagnostic tools. Ultrasonography is the basic investigation done for patients coming to the emergency room with acute abdominal pain. In cases of mucocele of the appendix, if the outer diameter of the appendix is 15 mm or more, is diagnostic with 83% sensitivity and 92% specificity.^{12,13} Another sign is considered to be specific for diagnosis of mucocele of the appendix is onion skin appearance.¹⁴ CT can be used to detect the signs specific to mucocele of appendix with high accuracy which includes presence of appendix lumen more than 1.3 cm, cystic dilatation, and wall calcifications.¹⁵ The 'volcano sign' on colonoscopy is specific to mucocele which presents as the appendiceal orifice in the centre of a firm mound covered by normal mucosa or a yellowish lipoma like submucosal mass.¹⁶ On colonoscopy synchronous and metachronous tumours of colon can be identified.

The goal of surgical treatment if mucocele of the appendix is to remove the mucocele intact without rupturing it to avoid complications like pseudomyxoma peritonei, which has a high morbidity and poor survival rates.¹⁸ The treatment options to surgically resect a mucocele of appendix can either be by laparotomy or laparoscopy with the main objectives of the surgery being: avoiding the

rupture of the mucocele into the peritoneal cavity and to have a detailed examination of the abdomen where mucinous tumours are most common.¹⁹ Dhage-Ivatury and Sugarbaker proposed an algorithm for the selection of patients for type of surgery (open/laparoscopy).¹³ It comprises of several factors: mucocele is perforated or not; base of the appendix (margins of resection) is free of tumour or not; and presence of positive lymph nodes of mesoappendix and ileocolic. Hence patients may require different operative procedures varying from a simple appendectomy to the right colectomy, and cytoreductive surgery, with additional heated intra-operative intraperitoneal chemotherapy and early post-operative intraperitoneal chemotherapy if there is involvement of the base.⁹ In this patient the mucocele was not perforated (no discharge into the peritoneum cavity), there was malignant changes in the base of the appendix (positive margins of resection) and the regional lymph nodes were negative. Therefore, patient was advised for further management with right hemicolectomy and chemotherapy.

CONCLUSION

To conclude, appendiceal mucocele is a rare disease with asymptomatic presentation. Early and accurate diagnosis preoperatively with history, clinical examination, ultrasonography, particularly CT, should be done for the selection of appropriate surgical technique to avoid post-operative complications. In our opinion, every patient more than 50 years' old who arrives at the emergency department with clinical symptoms of acute appendicitis must undergo CT and managed accordingly.

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