

## Uncommon pathology in a common surgical emergency: giant appendiceal mucocele with malignancy mimicking appendicitis

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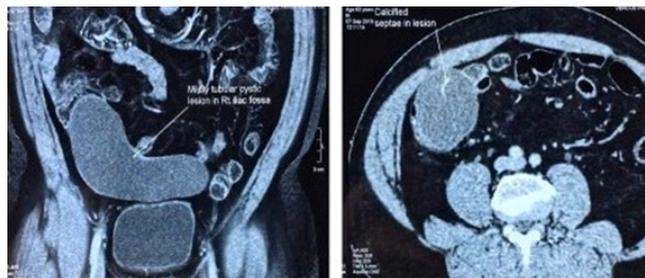
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### Introduction

Acute appendicitis with its classic symptoms of right lower abdominal pain, fever and vomiting is well-known, however, symptoms arising from the appendix can occasionally be atypical particularly when there are some rare underlying pathologies within. One such pathology is an Appendiceal Mucocele. It is usually discovered incidentally at the surgery or during radiological evaluation done for unrelated complaints. Surgical options can be a simple standard appendicectomy, partial caecectomy, ileocaecectomy or even right hemicolectomy, the ultimate goal being the attainment of clear margins. It is recommended that surgery be offered for all appendiceal mucoceles because of the risk of underlying malignancy. Here we present a case of a giant appendiceal mucocele with an underlying malignancy which was successfully treated by a laparoscopic limited right hemicolectomy.

### Case presentation

A 62-year-old gentleman presented with lower abdominal pain and increased frequency of micturition for a few days. An ultrasound scan of the abdomen revealed a large cyst in the appendix with prostatomegaly. His vital signs were normal. Abdominal examination revealed a mildly tender, well-defined lump in the right iliac fossa. Contrast-enhanced computed tomography of the abdomen revealed a 15 cm x 6.2 cm x 6.3 cm tubular cystic lesion in the right iliac fossa arising from the ileocaecal junction suggestive of appendiceal mucocele (Figure 1). We, therefore, proceeded with laparoscopy. At laparoscopy, the findings were confirmed (Figure 2). Given the possibility of malignancy in such a large appendiceal mucocele, a limited right hemicolectomy was performed. His postoperative course was uneventful. Histopathological examination confirmed mucinous cystadenoma of the appendix, both proximal and distal resection margins were clear and there was no regional lymph



**Figure 1.** CECT abdomen showing a giant tubular fluid-filled mucocele of the appendix (Left – coronal view, Right – axial view)



**Figure 2.** Large tubular structure in right iliac fossa (Left – Laparoscopic view, Right – Extracorporeal view after delivery of ileocaecal junction)

node involvement. The patient is currently doing well and is under follow up.

### Discussion

Appendicular mucocele is a rare condition where there is a cystic tubular dilatation of the appendiceal lumen. It occurs in patients in their 5th or 6th decades and there is a slight female preponderance. They are often asymptomatic, however, they can present as acute appendicitis. Rarely they can present with bowel obstruction or gastrointestinal bleeding due to intussusception, genitourinary symptoms due to obstruction of the right ureter or bladder or generalized peritonitis from a rupture [1,4]. The relevance of appendiceal mucocele in the spectrum of appendiceal tumours was not clearly defined until recently when the Peritoneal Surface Oncology Group International (PSOGI) developed a consensus classification that has helped to resolve much of the confusion surrounding the diagnostic terminology. Accordingly, mucinous lesions of the appendix are divided into 1) Non-neoplastic appendiceal mucinous lesions (also referred to as inflammatory or obstructive mucoceles) like simple mucoceles or retention

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cysts characterized by degenerative epithelial changes secondary to obstruction due to faecolith and 2) Neoplastic appendiceal mucinous lesions which include a) Serrated polyp with or without dysplasia, b) Mucinous neoplasms – either a low-grade appendiceal mucinous neoplasm (LAMN) or as high-grade appendiceal mucinous neoplasm (HAMN) and c) Mucinous adenocarcinomas which can be well, moderately or poorly differentiated mucinous adenocarcinomas [1].

Radiological studies, in particular, Contrast-enhanced computed tomography and ultrasound abdomen can diagnose appendiceal mucocoeles, however, they cannot definitively distinguish between non-neoplastic and neoplastic lesions. On most occasions, neoplastic lesions are generally larger (measuring more than 2 cms) compared to non-neoplastic lesions. The presence of soft tissue thickening, wall calcifications and wall irregularity, but not an increase in wall thickness, are suggestive of malignancy. The presence of ascites with hypodense peritoneal lesions and scalloping of the liver surface suggests the intraperitoneal spread of neoplastic cells from a ruptured mucinous neoplasm [2].

Surgical resection is recommended for all appendiceal mucinous lesions. As there are no reliable criteria to exclude benign from malignant lesions, surgery should be pursued even for a benign-appearing appendiceal mucocoele on imaging studies. An earlier laparoscopic approach was not advised because of the risk of rupture but now with increasing expertise, more surgeons are favouring the laparoscopic approach. The decision on whether to perform the surgery, laparoscopic or open is largely dependent on the surgeon's expertise [3]. In either case, the principles followed have to be the same, which include resection of the appendix, wide excision of mesoappendix to facilitate retrieval of all peri-appendiceal lymph nodes and careful assessment of the base of the appendix to exclude any extension into the caecal wall. In patients with a positive margin at the appendix base or positive peri-appendiceal lymph nodes, a right hemicolectomy is warranted [3,4]. A more aggressive approach such as a radical resection, removal of all gross implantations and hyperthermic intraperitoneal chemotherapy (HIPEC) is recommended in cases of ruptured appendiceal mucinous neoplasms that have to lead to Pseudomyxoma Peritonei (PMP) [1].

The prognosis of appendiceal mucinous lesions is closely associated with the histopathology, presence and extent of peritoneal spread. Prognosis in benign lesions is excellent with a 5-year survival rate of 91% to 100% after standard appendicectomy. Neoplastic lesions, particularly high-grade appendiceal mucinous neoplasms (HAMNs) have a more

guarded prognosis even after curative resection. Histopathologic features such as the presence of extra-appendiceal neoplastic epithelium, high-grade cytology, architectural complexity and invasion are important predictors of recurrence. Prognosis is worse for mucinous adenocarcinomas, particularly if the appendiceal mucocoele ruptures into the peritoneal cavity. Therefore all of these patients need to be under surveillance. Tumour markers such as Carcinoembryonic antigen (CEA), Ca 19-9 and Ca 125 can be elevated in patients with advanced appendiceal mucinous lesions and the levels correlate with treatment outcomes [5]. Currently, there are no guidelines for post-treatment surveillance but it is advised according to the histology, grade and completeness of surgery [1,5].

### Conclusion

Appendiceal mucocoeles or appendiceal mucinous lesions can present as appendicitis. They are difficult to diagnose clinically. Computed Tomography of the abdomen is essential. Large sizes of mucocoeles should raise suspicion of underlying malignancy. Appropriate and timely surgical resection with clear margins leads to excellent post-op recovery and oncological outcomes, however, these patients require close follow-up.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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**Learning Points:**

- Appendiceal mucoceles, both benign and malignant can present as acute appendicitis
- There are no pre-op tests to convincingly differentiate benign from malignant appendiceal mucoceles
- Surgical excision with negative margins should be the treatment of choice for all appendiceal mucoceles and these patients require close postoperative follow-up.
- Laparoscopic excision can be offered in safe and experienced hands