Mucocele of Appendix with Appendicocolic Intussusception into the Caecum : A Rare Case Report

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Abstract: Appendiceal mucocele is a rare clinical entity. It can be caused by benign or malignant diseases resulting in obstruction of the appendix and consequent intraluminal accumulation of mucus secretions. Mostly it is discovered accidentally presenting as acute appendicitis. Appendiceal intussusception into the caecum associated with mucocele of the appendix is a rare cause of abdominal pain and interestingly difficult to diagnose. However, other possible differential diagnoses should be kept in mind. A 71 year-old male presented to emergency department with mild pain in right lower quadrant of abdomen for 15 days with mild tenderness in right iliac fossa on examination. On further investigation and work up, a diagnosis of mucocele of appendix was obtained. Subsequently, elective open appendicectomy was performed. Intraoperatively, it was found to be mucocele of appendix with intussusception into the caecum (appendicocolic). Open appendicectomy with reduction of appendicocolic intussusception was done. Postoperative HPE confirmed the diagnosis. Patient recovered well with uneventful hospital stay. The patient is still on regular follow-up till date.

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I. Introduction

Appendiceal mucocele is a rare entity caused by obstructive dilatation of the appendix due to intraluminal accumulation of mucoid material.¹ The incidence is 0.2% to 0.7%.¹ There are four histologic types of appendiceal mucocele namely; retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.²

If left untreated, it may progress to develop into pseudomyxoma peritonei which has a high mortality.² Appendicular intussusception into the caecum is an uncommon type of intussusception, with an incidence of approximately 0.01% in patients undergoing appendicectomy.³ Intussusception associated with appendiceal mucocele is extremely rare and they are rarely associated with clinical manifestations.

Here, we are reporting a rare case of of appendiceal mucocele with appendicocolic intussusception into the caecum.

II. Case Report

A 71-year old male presented to the emergency department with mild pain in the right lower abdomen associated with nausea. His bladder and bowel habits were normal. He had history of similar episodes of off and on pain in the past 5 years. No other significant past medical or surgical history. On general physical examination, he was afebrile with minimal constitutional symptoms. On palpation, a mildly tender mass was present in the right like fossa. Other systemic examinations were normal.

All routine baseline investigations were normal.USG W/A report showed appendicitis with B/L renal cysts.

Barium enema double contrast study showed smooth indentation over the medial aspect of caecum and adjacent terminal ileal loop. CECT Abdomen showed a blind ending tubular structure measuring 98mm in length showing irregular lumen measuring upto 38mm in diameter in the RIF with luminal content showing homogenous fluid attenuation, all s/o mucocele of appendix, and b/l renal simple cortical cysts.

The patient was managed conservatively at the initial presentation and was put up for elective open appendicectomy. At the time of surgery, cystic mass was found at the base of appendix with intussusception into the lumen of caecum. No surrounding free fluid and no lymphadenopathy with minimal adhesion was noted. Reduction of the intussusception followed by appendicectomy was done. HPE showed mucosal appendiceal hyperplasia producing mucocele of appendix with no evidence of nuclear atypia .Patient recovered well with uneventful hospital stay and is on regular follow up till date.

III. Discussion

Appendiceal mucocele was first described by Rokitansky in 1842.³ It is caused by progressive dilatation of a lumen as a result of an accumulation of a large amount of mucus. It can be caused by benign or malignant diseases resulting in obstruction of the appendix and consequent intraluminal accumulation of mucus secretions.⁴ Mucocele in the appendix may be classified according to the histological characteristics of lumen obstruction.² Rupture of the appendix containing epithelial adenoma cells with low or high grade dysplasia may lead to the dissemination of the epithelium causing mucinous ascites or pseudomyxoma peritonei.⁵

The clinical presentation of mucocele in the appendix is usually nonspecific with difficult preoperative diagnosis. *Stocchi L et al* in their study found that the most common complaint in appendiceal mucocele is pain in the right lower quadrant of the abdomen which may last for months, being noticed in up to 50% of the cases, associated or not with a palpable tumor.⁶ Similar finding was noted in another study by *Ruiz-Tovar J et al.*⁷ Similarly in our study, the patient presented with mild pain in the right lower abdomen with history of similar episode in the past with a palpable mildly tender right iliac fossa mass on examination.

Preoperative diagnosis is crucial for adequate surgical intervention. Imaging tests such as ultrasound, computed tomography, enema and colonoscopy etc may suggest the presence of mucocele of the appendix which help to deliver the definitive treatment.^{8,9} In our study, USG whole abdomen showed features of appendicitis, barium enema showed smooth indentation over the medial aspect of caecum and adjacent terminal ileal loop and findings of CECT whole abdomen were all suggestive of mucocele.

Despite growing evidence favoring the laparoscopic approach, open surgery is currently the standard surgical treatment of appendiceal mucocele. *Gonzalez et al* reported a case of laparoscopic mucocele resection that was followed by early peritoneal progression, forcing them to conclude that this entity was a contraindication to laparoscopic resection.¹⁰ Data from other studies indicate that laparoscopic appendicectomy for the management of appendix neoplasms is associated with long-term results comparable to those obtained with open appendectomy.¹¹ Open appendicectomy was done for our case and and a cystic mass was found at the base of appendix with intussusception into the lumen of caecum. No surrounding free fluid and no lymphadenopathy with minimal adhesion was noted.

Association of appendiceal mucocele with intussusception as found in our case is very uncommon with estimated incidence of upto 3.3% of adult intussusception.^{12,13} No case of appendicular mucocele as a lead point of intussusception was found in the series published in 2003 at the Mayo Clinic with 132 patients,⁶ and another in 2007 with 35 cases in a 21 year period.⁷ In 2004, *Yamaguchi et al* noted only 13 cases of appendicular mucocele as a lead point among 400 cases of adult intussusception over the past 10 years from Japanese domestic report.¹³ Our patient underwent open appendectomy with reduction of appendicocolic intussusception without subsequent complications.

IV. Conclusion

Appendiceal mucocele is a rare entity caused by obstructive dilatation of the appendix due to intraluminal accumulation of mucoid material. If left untreated, it may progress to develop into pseudomyxoma peritonei which has a high mortality. In about 50% of cases it is discovered accidentally during radiologic examinations or at surgery. Preoperative accurate diagnosis is crucial for adequate surgical intervention to prevent peritoneal dissemination and to avoid intraoperative and postoperative complications and avoidable reoperation.

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Figure 1. Mucocele of appendix with appendicocolic intussusception into the caecum.

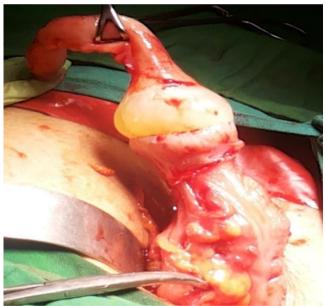


Figure 2. Base of appendix being dissected out of the caecum.

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