



APPENDICULAR MUCOCELE ACT AS TOURNIQUET AND PRESENTED AS GANGRENOUS PERFORATED INTESTINE.

General Surgery

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ABSTRACT

Mucocele of the appendix is a rare condition. Its presentation, like intestinal gangrene and perforation by making a tourniquet around the mesentery, is extremely rare. We reported a case of a female in her 60s who presented in a surgical emergency with a complaint of pain in the abdomen. Her X-ray showed free air, suggesting a hollow visceral perforation. An emergency explorative laparotomy was planned without further delay. Appendix having a 3x2 cm cystic mass at its tip that loops around the ileum mesentery caused ischemic necrosis of the distal ileum and perforation. Extra care was given to avoid rupture of cystic swelling in the peritoneal cavity. Appendicectomy along with resection of the gangrenous bowel and a double barrel ileostomy, was performed. On cut sections, mucin fluid is present in cystic swelling, and histopathology suggests this is a mucous retention cyst arising from the tip of the appendix. Bowel continuity restored after 3 months and patient remains disease-free during the 2-year clinical follow-up period. Appendicular mucocele act as tourniquets and can present as intestinal obstruction; only 10 such cases are available in the literature, but none presented as perforation peritonitis. The present case provides an understanding of such a common emergency presentation as perforation peritonitis for a rare disease of appendicular mucocele and measures taken during emergency surgery to prevent spillage of mucin in the peritoneal cavity that prevent any chance of pseudomyxoma peritonei.

KEYWORDS

Appendiceal mucocele; Appendicular tourniquet; Intestinal gangrene; Intestinal perforation

INTRODUCTION:

Appendicular mucocele is a rare condition; only 0.3–0.6% of appendix specimens have a diagnosis of mucocele of the appendix. 1, 2 Right lower quadrant pain mimics acute appendicitis, and a palpable mass is present in 50% of appendix mucocoeles. Very rarely, urinary dysfunction or hematuria can also present. 3, 4 Preoperative diagnosis can be done with ultrasonography, computed tomography, and colonoscopy. 5, 6 But most of the time, the diagnosis is intraoperative. Elevated levels of carcino-embryonic antigen are also helpful in diagnosing neoplastic origin mucocoeles. 7 In most cases, appendiceal mucocele is an intraoperative finding of emergency explorative laparotomy. It is very unusual for an appendicular mucocele to present like an intestinal perforation. The diagnosis of this rare aetiology is often missed preoperatively, as the majority of cases of perforation peritonitis require an emergency laparotomy. More radiological investigations will delay the definitive management. Early surgical intervention is the treatment of choice. Our case will provide an understanding of the unusual clinical presentation of a rare disease like appendicular mucocele and the steps taken for successful management.

CASE REPORT:

A lady aged 60 years presented at Surgical Emergency with an acute onset of abdominal pain for the last 5 days. There was no relevant medical or surgical history. On examination, the abdomen was distended and tense, and diffuse severe tenderness with guarding was present. Blood investigations revealed a total leukocyte count of 19800/microliter. An erect abdominal X-ray showed free air under the right dome of the diaphragm, suggesting a hollow viscus perforation. On the basis of history, clinical, and x-ray findings, a diagnosis of intestinal perforation was made. □ An emergency explorative laparotomy with written consent was planned without further delay. Intraoperative findings were spillage of faecal content and a gangrenous loop of a distal ileum, 40 cm from the ileocecal junction. Twenty-five centimetres of ileum were gangrenous, with a single perforation in it. On further exploration, an appendicular tourniquet around the mesentery of the distal ileum was identified as the cause of this gangrene (Fig. 1).

size 3x2 cm at its tip. A diagnosis of mucocele was suspected and planned for appendectomy due to the noninvolvement of the appendix base and the absence of enlargement of lymph nodes or hepatic metastases. The rest of the abdominal and pelvic cavity was free of tumours. The peritoneal cavity was packed with saline-soaked sponges; careful handling and ligation of the appendix were done to prevent spillage of its contents into the peritoneum. An appendicectomy with resection of perforated and gangrenous ileum was done along with a double barrel ileostomy. On the cut section of the specimen, a collection of mucinous fluid was observed at the tip of the appendix; the rest of the lumen was not dilated and was not communicating with the cyst (Fig. 2).

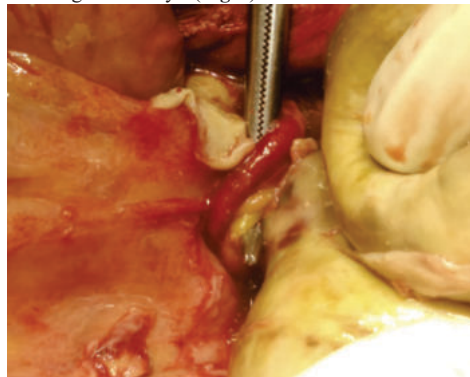


Fig.1 Appendix Working As Appendicular Tourniquet

On histopathological examination, no mucus-producing epithelium lining the cyst cavity and no malignant cells were observed. The rest of the appendix shows signs of appendicitis. A mucous-containing cystic swelling without mucin-producing epithelial and neoplastic cells was diagnosed as a mucous retention cyst arising from the tip of the appendix. Colonoscopy does not reveal any colonic carcinoma. Recovery was uneventful, and the patient was discharged on the fifth postoperative day. Bowel continuity was restored after three months and she remained disease-free during two years of postoperative clinical follow-up.

This tourniquet was 5 mm in diameter, tubular, and had a cystic mass of

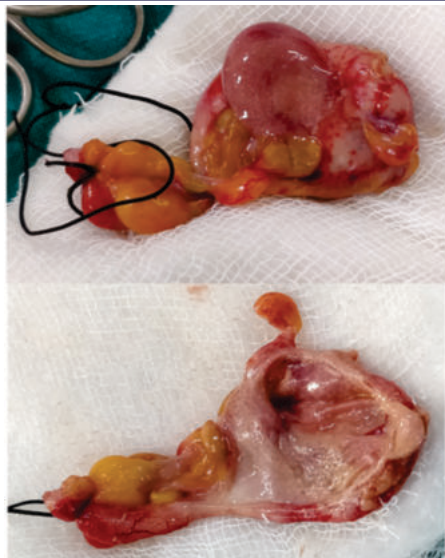


Fig.2 Mucous Coming Out Of Tip Of Appendix And Mucocoele Of Appendix Dilate Tip Of Appendix Cut Open Specimen

DISCUSSION:

Mucocoele of the appendix is more common in females than males and more common in patients older than 50 years. 8 Most common symptoms of mucocoele are right lower quadrant pain and palpable mass in 50% of cases; a few cases of urinary dysfunction and hematuria were also reported.^{3, 4} While 25% of patients do not have any symptoms.⁹ Preoperative diagnosis can be possible with USG and CT scans. CT is more specific in its diagnosis.⁵ Colonoscopy is also helpful in some cases.⁶ CEA raised value is helpful in malignant mucocoele cases.⁷ The preoperative diagnosis of appendicular mucocoele presented with bowel strangulation or perforation is difficult as these conditions require an emergency laparotomy because more delay in radiological investigations will delay definitive management. In the present case, the intestine was gangrene and perforated due to an appendicular tourniquet having a mucocoele at its tip, which presented as perforation peritonitis. Only 10 cases (Table I) with appendicular mucocoele acting as tourniquet and presented as intestinal obstruction have been found in the literature to date.¹⁰⁻¹⁹

Table I. Reported Cases Of Intestinal Obstruction Secondary To A Mucocoele Of The Appendix.

Serial no.	Author	Year	Age	Sex	Preoperative diagnosis as mucocoele	Size of mucocoele	Surgery performed	Pathological diagnosis
1	Mourad et al. ¹⁰	1999	57	M	Yes	12x7x7	Ileocecal resection	Mucinous cystadenoma
2	Chong et al. ¹¹	2001	65	F	No	8x5	Ileocecal resection	Mucinous cyst adenoma
3	Garg et al. ¹²	2011	70	M	No	7.5	Appendectomy	Mucinous cystadenoma
4	Miyakura et al. ¹³	2012	89	F	No	9x3	Ileocecal resection	Mucous containing cystic appendix without mucin-producing, neoplastic epithelial cells
5	Zaharie et al. ¹⁴	2012	39	F	No	Unkown	Appendectomy Ileal segment Resection	Mucinous cystadenoma

6	Opreanu et al. ¹⁵	2013	51	F	No	3x2.5x1.4	Ileocecal resection	Benign Mucocoele
7	Valverde et al. ¹⁶	2016	76	M	No	19x8	Appendectomy and part of cecum	Papillary cyst adenoma of appendix with mucocoele
8	Shui Xu et al. ¹⁷	2017	76	M	No	4x2.5x2.5	Appendectomy	Mucinous cystadenoma
9	Komo et al. ¹⁸	2018	79	F	No	3x1.5	Appendectomy	LAMN
10	Takei et al. ¹⁹	2020	92	M	No	5x2.5	Ileocecal resection with LN dissection	LAMN
11	Our case	2022	60	F	No	3x2	Appendectomy Ileal segment Resection	Mucous containing cystic appendix without mucin-producing, neoplastic epithelial cells

Small bowel perforation due to an appendicular mucocoele tourniquet is very rare, and this is the only available case.

Mucocoele can be caused by an inflammatory or neoplastic pathology of the appendiceal mucosa. They are mucosal hyperplasia, retention cysts, mucinous cystadenomas, and mucinous cystadenocarcinomas. The histopathological examination in the present case showed no morphologically well-defined, mucin-producing neoplastic epithelial cells in the wall of the appendix. The diagnosis of a retention cyst was thus made. An algorithm for surgery has been designed by Dhage-Ivatury and Sugarbaker.^{20, 21} Simple appendectomy is for benign mucocoele with normal caecum and appendicular base and no evidence of perforation in the cyst; the same was true in our case. Right hemicolectomy is the preferred choice for suspected malignant mucocoele (perforated mucocoele, enlarged mesenteric lymph node, or positive cytology for malignant cells). In the present case, the patient underwent appendicectomy and resection of a gangrenous bowel segment, as only the tip of the appendix was involved and there was no perforated mucocoele or enlarged mesenteric lymph node. Exploration of the whole abdominal cavity should be done due to the high association of the appendicular mucocoele and other mucin-secreting cell cancers with colon and ovarian cancers.²⁰ Extra care must be taken when handling a mucocoele to prevent its spillage into the peritoneal cavity, which can cause pseudomyxoma peritonei. In this case, the peritoneal cavity was packed with normal saline soaked gauze before appendicectomy, and careful handling and ligation of the appendix were performed to prevent spillage of mucous in the peritoneum. The present case provides an understanding of a common clinical presentation of a rare disease like appendicular mucocoele and measures taken during emergency surgery to ensure successful management without any complications.

CONCLUSION:

Mucocoele of the appendix can be presented clinically as perforation peritonitis. Most of the time, the diagnosis of mucocoele is intraoperative. There should not be any further delay in emergency surgery, as early intervention is always helpful in such cases. Extra care must be taken during appendix holding, ligation, and peritoneum packing with saline-soaked gauze to prevent spillage of mucous inside the peritoneal cavity, which can cause pseudomyxoma peritonei. Only appendicectomy can be done as a definitive surgery in cases of no involvement of the appendix base and no mesenteric lymphadenopathy intraoperatively.

Author contributions:

1st author drafted the manuscript. 1st, 2nd, 3rd, 4th and 5th author reviewed and edited the manuscript. Emergency laparotomy was performed by the 1st and 2nd. All authors read and approved the final manuscript.

Conflict Of Interest:

It is declared that they have no conflict of interest in this study.

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Consent :

Informed and written consent for surgical procedures, consent for publication and presentation was obtained from the patient.

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