JSCR Journal of Surgical Case Reports

Journal of Surgical Case Reports, 2018;6, 1–3

doi: 10.1093/jscr/rjy102
Case Report

CASE REPORT

Surgical treatment of mucocele of the appendix: a systematic review and case report

Eltaib A. Saad[1], Elamin Y. Elsamani[2], Walid E. AbdElrahim[1], Kamal E. Elsiddig[1], and Eltahir A.G. Khalil[3,*]

[1]Department of Surgery, Faculty of Medicine, University of Khartoum, Khartoum, Sudan, [2]Sudan Medical Specialization Board, Sudan, and [3]Department of Clinical Pathology and Immunology, Institute of Endemic Diseases, University of Khartoum, Khartoum, Sudan

*Correspondence address. Clinical Pathology and Immunology, Institute of Endemic Diseases, University of Khartoum, PO Box 102, 11111 Khartoum, Sudan. Tel/Fax: +249 83793267; E-mail: eltahirk@iend.org

Abstract

Introduction: Appendicular mucocele is a rare well-described clinico-pathological occurrence. It denotes an obstructive dilatation of the appendicular lumen by mucinous secretions.

Case report: A 60-year-old patient presented with right lower abdominal pain and nausea for 2 years. Abdominal CT scan suggested a diagnosis of a appendicular mucocele. Following informed consent, surgical exploration revealed a cystic mass arising from the body of the appendix with inflamed walls with no evidence of perforation. Simple appendectomy was performed as the caecum and the mesenteric nodes were free of pathological involvement. The final diagnosis of mucinous cystadenoma was confirmed by histopathology. Postoperative course was uneventful. The patient was in good health during a four years regular follow-up.

Discussion: Appendicular mucocele is a rare disease with vague symptoms. Abdominal imaging is an important diagnostic tool, but histopathology is the standard for definitive diagnosis. Surgery for benign appendicular mucoceles has an excellent long-term prognosis.

INTRODUCTION

Mucocele of the appendix is a rare but well-known clinico-pathological entity that has non-specific symptomology mimicking several common diseases. It is often detected as an incidental finding during surgery, routine radiological investigations or endoscopic examination. Appendicular mucocele has an incidence of 0.2–0.3% with a higher occurrence in females and people >50 years. It is an obstructive dilatation of the appendix caused by intra-luminal accumulation of mucoid material [1–3]. Four histologic types exist: retention mucinous cyst, mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma [2]. In mucinous cystadenomas, the luminal dilatation can reach up to 6 cm. Mucinous cystadenocarcinomas are less common than mucinous cystadenomas. This neoplasm rarely spreads through the lymphatic or vascular routes, but has a peculiar tendency of penetration and spread well beyond the appendix to the peritoneum. Moreover, in advanced cases the whole peritoneal cavity becomes distended with adhesive semi-solid mucous; a condition termed *pseudomyxoma peritonei* [4]. Complications of appendicular mucocele include: intestinal bleeding/obstruction, melena and pyonephrosis [4]. The worst complication is *pseudomyxoma peritonei* caused by spontaneous or iatrogenic perforation of the mucocele.

Received: March 25, 2018. **Accepted:** May 1, 2018

CASE REPORT

A 60-year-old male presented with a history of right lower abdominal pain, nausea with occasional vomits and no change in bowel habits for 2 years. The pain was dull, increased progressively and was relieved by simple analgesics. His appetite was good with no appreciable changes in his weight. There was no rectal bleeding, melena, mucoid stool, dyspoea or palpitations. His past medical and surgical histories were insignificant. He was generally unwell, not pale, not febrile or jaundiced and with stable vital signs. There were no stigmata of chronic liver or kidney diseases. Abdominal examination revealed minor tenderness over the right iliac fossa (RIF) without localized guarding or rigidity. There was a palpable cystic mass measuring $6 \times 4\,cm^2$ over the RIF, with a smooth surface and well-defined margins. Digital rectal examination was normal. His blood counts, glucose, liver and renal profiles were normal. Tumor markers (CEA, CA 19.9) levels were within normal ranges. Abdominal sonography showed a cystic mass measuring $6.5 \times 5.5\,cm^2$ arising in the RIF and displacing the urinary bladder to the left of the midline. Abdominal CT scan with oral and intravenous contrast revealed an oval-shaped cyst with a tubular fluid-density arising from inferomedial aspect of the caecum and extending down to the pelvic cavity in the right side posterior to the urinary bladder pushing it to the left, highly suggestive of an appendicular mucocele. No caecal masses or enlarged mesenteric lymph nodes seen. The liver was normal in size and texture with no evidence of metastatic deposits. Colonoscopy showed no luminal abnormality but the appendicular orifice could not be visualized clearly due to edema. Following informed consent, the patient had an exploratory laparotomy, which showed no evidence of peritoneal tumors, seedling or metastases. A cystic mass measuring $8 \times 6 \times 6\,cm^3$ was found in the RIF arising from the body of appendix. The appendicular wall was mildly inflamed, but there was no evidence of perforation or abscess formation. Simple appendectomy was performed as the caecum, base of the appendix and mesenteric node were free of any pathological involvement. The postoperative course was uneventful and the patient was discharged on Day 5 postoperatively in a good condition. Histopathological examination confirmed the final diagnosis of a benign mucinous cystadenoma arising from the body of the appendix with free margins of resection. The patient remained well on regular follow-up visits over a 4-year period. The patient consented in writing to the publication of the data.

DISCUSSION

Clinical presentation of appendicular mucoceles is usually vague as in this case; furthermore, it can be asymptomatic in a quarter of patients. Most commonly, patients present with right lower quadrant pain as seen in our patient. Palpable masses have been reported in 50% of cases, whereas urinary symptoms are rare [1, 3, 4]. The mass in this patient markedly displaced the urinary bladder but there were no urinary symptoms. Pre-operative diagnosis of appendicular mucocele is difficult because of the rarity of the condition and the non-specific nature of the presenting symptoms. Nevertheless, pre-operative diagnosis is important for the selection of the appropriate surgical procedure in order to prevent intra-operative complications especially particular peritoneal dissemination [1]. Sonographic examination is considered the first-line diagnostic modality that can probably differentiate benign and malignant mucoceles [5]. An appendicular diameter of 15 mm or more has been determined as a threshold for diagnosis of mucocele with a sensitivity of 83% and a specificity of 92%. Computed tomography (CT) scan is important to confirm the diagnosis and to evaluate the extent of the disease. Fine needle aspiration cytology is not usually recommended it increases the risk of perforation and dissemination into peritoneal cavity [6]. Colonoscopy usually reveals an elevation of the appendicular orifice; in addition, a yellowish mucous discharge would be visible as well. Colonoscopy is also important for the diagnosis of synchronous or metachronous colon cancers when present [7, 8].

Conventional surgery is generally preferred to laparoscopic approach as the latter increases the risk of rupture, but it is still performed for selected patients. An algorithm for the selection of the type of surgery has been formulated by Dhage-Ivatury and Sugarbaker [1, 3, 4, 9, 10]. Simple appendectomy is the choice for patients with benign mucocele as suggested by the presence of a normal caecum and appendicular base and no evidence of perforation, as in our patient. Right hemicolectomy is recommended when malignant mucocele is suspected by the presence of a perforated mucocele, enlarged mesenteric lymph node or a positive cytology. An accurate exploration of the abdomen is advised due to the well-known association between the appendicular mucocele and other mucin-secreting cells cancers, such as colon and ovarian cancers [7]. In conclusion, mucocele of the appendix is a rare disease with vague symptoms. Abdominal US and CT scans are important diagnostic tools, but histopathology is needed for definitive diagnosis. Surgery for benign appendicular mucoceles has an excellent long-term prognosis.

CONFLICT OF INTEREST STATEMENT

None declared.

REFERENCES

1. Demetrashvili Z, Chkhaidze M, Khutsishvili K, Topchishvili G, Javakhishvili T, Pipia I, et al. Mucocele of the appendix: case report and review of literature. *Int Surg* 2012;**97**:266–9.
2. Marudanayagam R, Williams GT, Rees BI. Review of the pathological results of 2660 appendectomy specimens. *J Gastroenterol* 2006;**41**:745–9.
3. Rampone B, Roviello F, Marrelli D, Pinto E. Giant appendiceal mucocele: report of a case and brief review. *World J Gastroenterol* 2005;**11**:4761.
4. Sugarbaker PH. Appendiceal epithelial neoplasms and pseudomyxoma peritonei, a distinct clinical entity with distinct treatments. In: Bland KJ, Büchler MW, Csendes A, Garden OY, Saar MG, Wong J, eds. *General Surgery. Principles and International Practice*. London-Limited: Springer, 2009;885–93.
5. Lien WC, Huang SP, Chi CL, Liu KL, Lin MT, Lai TI, et al. Appendiceal outer diameter as an indicator for differentiating appendiceal mucocele from appendicitis. *Am J Emerg Med* 2006;**24**:801–5.
6. Pickhardt PJ, Levy AD, Rohrmann CA Jr, Kende AI. Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. *Radiographics* 2003;**23**:645–62.

7. Karakaya K, Barut F, Emre AU, Ucan HB, Cakmak GK, Irkorucu O, et al. Appendiceal mucocele: case reports and review of current literature. *World J Gastroenterol* 2008;**14**:2280.
8. Zanati SA, Martin JA, Baker JP, Streutker CJ, Marcon NE. Colonoscopic diagnosis of mucocele of the appendix. *Gastrointest Endosc* 2005;**62**:452–6.
9. Lau H, Yuen WK, Loong F, Lee F. Laparoscopic resection of an appendiceal mucocele. *Surg Laparosc Endosc Percutan Tech* 2002;**12**:367–70.
10. Dhage-Ivatury S, Sugarbaker PH. Update on the surgical approach to mucocele of the appendix. *J Am Coll Surg* 2006; **202**:680–4.