



Minimally invasive appendectomy for resection of appendiceal mucocele: Case series and review of the literature

Sonia T. Orcutt^a, Daniel A. Anaya^{a,b}, Mokenge Malafa^{a,*}

^a Department of Gastrointestinal Oncology, H. Lee Moffitt Cancer Center and Research Institute, Tampa, FL, 33612, USA

^b Section of Hepatobiliary Tumors, Department of Gastrointestinal Oncology, H. Lee Moffitt Cancer Center and Research Institute, Tampa, FL, 33612, USA

ARTICLE INFO

Article history:

Received 20 April 2017

Accepted 23 May 2017

Available online 29 May 2017

Keywords:

Mucocele

Appendix

Robotic surgical procedure

Surgical procedures

Minimally invasive

Laparoscopy

Case report

ABSTRACT

INTRODUCTION: Appendiceal mucocèles encompass neoplastic and non-neoplastic causes of a distended Appendix filled with mucus. Appendectomy is recommended when an appendiceal mucocele is identified, incidentally or otherwise, in the event it is secondary to a malignancy. For an intact mucocele, it is critically important to avoid rupturing the mucocele during resection, as rupture of a neoplastic mucocele can result in pseudomyxoma peritonei, or mucin deposits in the peritoneum, which is associated with long-term morbidity and mortality. For this reason, laparotomy is the traditionally recommended surgical approach for treatment.

PRESENTATION OF CASES: In our case series, we describe two patients, a 49-year-old woman and a 79-year-old man, with incidentally identified appendiceal mucocèles. These patients were successfully treated with minimally invasive approaches to appendectomy, one with a robotic approach and one with a hand-assisted laparoscopic approach. The mucocèles were removed without rupture, and both patients recovered well postoperatively without complication.

DISCUSSION: While laparotomy is the traditionally recommended surgical approach for resection of appendiceal mucocèles, certain minimally invasive techniques allow for safe removal of the mucocèles while minimizing the morbidity of laparotomy.

CONCLUSION: Minimally invasive approaches to appendectomy, specifically the robotic-assisted approach and the hand-assisted laparoscopic approach, can be considered for safe resection of appendiceal mucocèles.

© 2017 Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Appendiceal mucocele is a rare diagnosis, accounting for less than 1% of appendiceal pathologies [1]. The term “mucocele” is nonspecific and describes a distended Appendix whose lumen is filled with mucus but does not represent a pathological description. Mucocèles of the Appendix are diagnosed incidentally about 50% of the time [1], often as a result of imaging for synchronous malignancies or benign conditions. Symptoms of mucocèles can include abdominal pain or mass, weight loss, nausea, or a change in bowel habits [2]. In addition, patients with a mucocele may present with appendicitis, intussusception, appendiceal torsion, gastrointestinal bleeding, or increasing abdominal girth from rupture of a neoplastic mucocele [1]. Traditionally, resection is recommended for mucocèles, as it can be unclear from imaging which mucocèles harbor

malignancy. However, resection must be done with caution, as the rupture of a neoplastic mucocele can give rise to pseudomyxoma peritonei, or the deposition of mucin throughout the peritoneal cavity, which can result in significant morbidity from mucinous ascites and bowel obstruction [1]. Due to this concern for rupture, open resection through laparotomy has been advocated [2].

There have been case reports and case series describing the use of standard laparoscopy for appendiceal mucocèles [3–7], although there is reticence to adopt this technique due to the technical difficulty of performing a laparoscopic appendectomy without causing iatrogenic rupture. We present two patients with appendiceal mucocèles who were successfully resected using minimally invasive approaches other than standard laparoscopy at a tertiary cancer center by surgical oncologists with significant experience with these approaches. One was resected with a hand-assisted laparoscopic technique and the other with a robotic-assisted technique. Both methods allowed for safe removal of the mucocèles without rupture, while minimizing hospital stays and morbidity from laparotomies. This case series is compliant with the SCARE and PROCESS guidelines [8,9].

* Corresponding author at: Department of Gastrointestinal Oncology, H. Lee Moffitt Cancer Center and Research Institute, 12902 Magnolia Drive, Tampa, FL 33612, USA.

E-mail addresses: Sonia.Orcutt@moffitt.org (S.T. Orcutt), Daniel.Anaya@moffitt.org (D.A. Anaya), Mokenge.Malafa@moffitt.org (M. Malafa).



Fig. 1. Computed tomography of the abdomen demonstrating a cystic mass in the right lower quadrant.

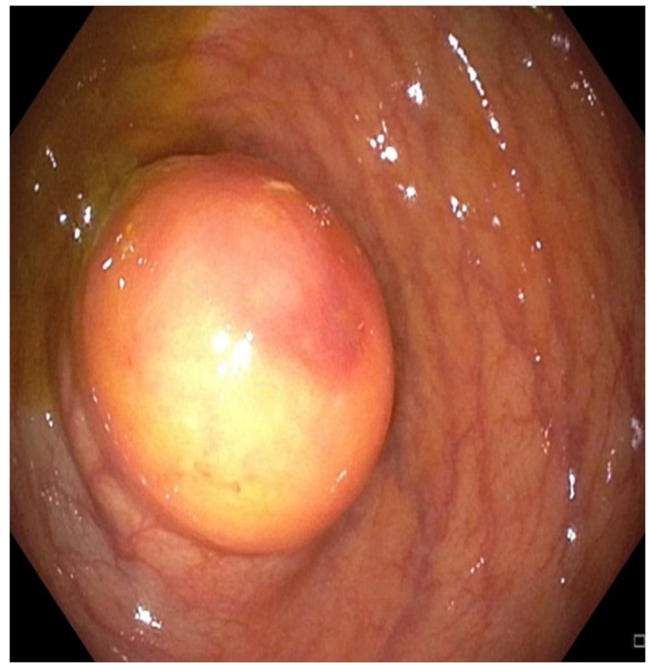


Fig. 2. Colonoscopic findings of the appendiceal mass protruding through the appendiceal orifice into the cecum.

2. Presentation of cases

2.1. Case 1

An otherwise healthy 49-year-old woman presented to her primary care physician with low-grade fevers and elevated liver function tests. She underwent abdominal ultrasonography to assess for a biliary etiology of her symptoms and was found to have an incidental pelvic cystic mass. After her liver function tests had returned to normal levels, she saw a gynecologic oncologist for further workup. Transvaginal ultrasonography demonstrated a large right ovary at 8.3 cm in maximum dimension, with a complex cystic mass measuring 7.2 cm. Due to a concern for malignancy, her gynecologist ordered a computed tomography scan, which identified an appendiceal mass (9.5×3.7 cm) with no other abnormalities (Fig. 1). CEA and CA 125 were within normal limits. She was then referred to surgical oncology. A colonoscopy identified a tumor at the appendiceal orifice, which was found upon biopsy to contain intestinal mucosa with hyperplastic changes, without evidence of dysplasia or carcinoma (Fig. 2). With the diagnosis of appendiceal mucocele, she was offered a laparoscopic appendectomy. At the time of the operation, there was some difficulty visualizing the base of the Appendix clearly, and given the size and somewhat tenuous tissue harboring the mucocele, a hand port was placed. This allowed the cecum to be mobilized and the base of the Appendix to be clearly identified during careful manipulation of the Appendix and mucocele. The cecum was divided just distal to the appendiceal base, to ensure removal of the entire mass. The Appendix was maintained intact, and there was no spillage of mucin during the operation. There was also no evidence of mucin deposits on exploration. The patient was discharged in good condition and experienced no complications. Final pathology demonstrated a low-grade mucinous neoplasm with copious mucin production, negative margins, and no invasive component.



Fig. 3. Computed tomography of the abdomen demonstrating a cystic mass in the right lower quadrant.

2.2. Case 2

A 79 year-old man, with a history of intraductal papillary mucinous neoplasm of the pancreas, melanoma, and prostate cancer, was undergoing surveillance for his previous cancers when he was identified on computed tomography to have a 2.2×2 cm lesion abutting his appendix (Fig. 3). He was asymptomatic. He had undergone a colonoscopy in 2014, which had not identified any malignancy. CEA and CA 19-9 were within normal limits. He was offered a robotic appendectomy. There was no evidence of intraperitoneal mucin deposits. The robotic approach allowed the Appendix to be meticulously dissected and the base of the Appendix to be clearly identified without manipulating the appendiceal mucocele itself. The cecum was divided just distal to the appendiceal base, again to ensure removal of the entire mass. The specimen was intact and not ruptured (Fig. 4). The patient was discharged in good condition without complications. Final pathology

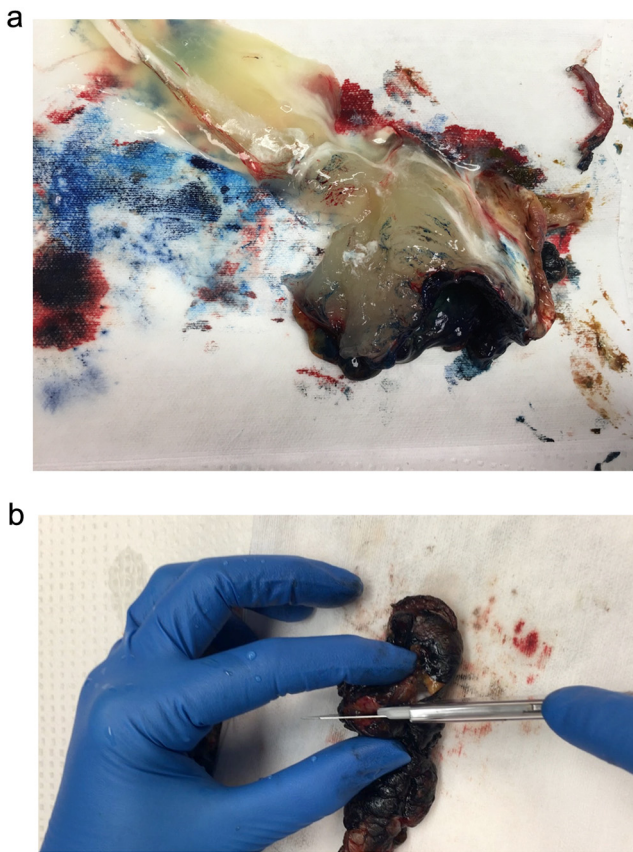


Fig. 4. Pathologic sectioning of the masses, with mucus noted extruding from the masses. (a) Case 1 and (b) Case 2.

demonstrated a low-grade mucinous neoplasm in the background of a serrated adenoma, with no evidence of dysplasia or carcinoma.

3. Discussion

In this report, we present two patients with appendiceal mucocèles, both of which were identified incidentally. Each patient's appendectomy was successfully performed using minimally invasive approaches, and neither mucocèle was disrupted. In addition, the potential morbidity of the traditionally recommended approach of laparotomy was minimized with the use of minimally invasive approaches.

Mucocèles represent a variety of histologies, both neoplastic and non-neoplastic, that can result in a dilated, mucus-filled appendix. Historically, mucocèles are described as arising from four different processes. Benign causes of mucocèles include mucosal hyperplasia or a simple cyst secondary to obstruction by a fecalith or stricture. Neoplastic causes include mucinous cystadenomas, which are noninvasive, and mucinous cystadenocarcinomas, which are malignant. Simple cysts represent about 25% of mucocèles, hyperplasia and mucinous cystadenomas each represent about 33%, and mucinous cystadenocarcinomas represent 5%–10% [1]. Despite their rarity, mucinous cystadenocarcinomas account for 37% of appendiceal neoplasms [10]. There is a female predominance to cystadenocarcinomas (as opposed to other appendiceal neoplasms), and diagnosis is typically in the seventh decade [1].

There have been inconsistencies in the literature in the description and reporting of mucinous appendiceal neoplasia and pseudomyxoma peritonei. A recent international Delphi consensus was convened to better clarify and standardize these definitions [11]. The consensus for appendiceal tumors was that the term

"cystadenoma" should no longer be used. "Mucinous adenocarcinoma" should be used to describe mucinous tumors with infiltrative invasion, "low-grade appendiceal mucinous neoplasm" is now used to describe tumors with spreading growth but without destruction, "high-grade appendiceal mucinous neoplasm" describes tumors with low-grade architectural features but high-grade cytological features, and "signet ring carcinoma" describes tumors with over half of cells demonstrating signet ring morphology [11]. The term "pseudomyxoma peritonei" is used in reference to the presence of mucinous deposits in the peritoneal cavity secondary to a mucinous neoplasm and can have low-grade, high-grade, or signet ring cell histological features [11].

Imaging characteristics are relatively consistent and can help in developing a diagnosis prior to an operation. Computed tomography scan can distinguish cystic masses associated with the Appendix or cecum from other cystic masses, such as ovarian masses. Neoplastic processes tend to be larger, measuring at least 2 cm [2]. Calcification of the wall of the mucocèle may be seen [12], with neoplastic processes being suggested by irregularity or a solid component of the wall. A cystic mass can be noted on ultrasonography, often with wall calcification. Peritoneal deposits and omental thickening, with or without associated ascites, may be seen if there is pseudomyxoma peritonei. If the patient is presenting with appendicitis, signs of inflammation or gas bubbles may be seen [12]. Mucocèles of the Appendix can also be identified on colonoscopy. Cecal masses described as submucosal or extrinsic in the region of the appendiceal orifice are often seen [13].

Resection is recommended when identified (whether incidentally or not) due to the potential for the mucocèle to harbor a neoplasm. It is imperative during resection to avoid rupture of the mucocèle and potential spillage of mucin in the peritoneal cavity, which can result in pseudomyxoma peritonei with associated morbidity and mortality. The 5-year survival rate for mucinous cystadenocarcinoma is 32%–58% [10], although once it has progressed to pseudomyxoma peritonei, survival is reported at 23% [1]. However, with newer techniques, including the use of cytoreductive surgery and heated intraperitoneal chemotherapy, 10-year survival can improve to close to 50% [14]. Appendectomy is recommended for non-neoplastic causes of mucocèle as well. A more extensive right hemicolectomy is additionally recommended for patients with mucinous adenocarcinoma without peritoneal disease, to obtain negative margins and remove potentially involved lymph nodes. Once there is evidence of peritoneal disease, right hemicolectomy no longer offers a survival benefit and is not recommended [2].

Historically, laparoscopy has been considered a contraindication for the surgical treatment of appendiceal mucocèles. This is due to reports of patients undergoing laparoscopic appendectomy for intact mucocèles who later developed peritoneal implants, suggesting that maintaining the integrity of the wall of the mucocèle is challenging [15]. In addition, some authors have recommended laparotomy for improved exploration of the abdomen for mucin deposits [2]. However, several case reports and small case series have described laparoscopy for appendiceal mucocèles without intraoperative rupture of the mucocèle [3–7], although there is limited data on long-term recurrence. While none of the cases reported resulted in rupture of the mucocèle, it is possible that unreported cases exist which involve rupture. The surgical approaches used in the current study, namely hand-assisted laparoscopy and robotic-assisted laparoscopy, have specific benefits over standard laparoscopy that may allow for safer, atraumatic removal of the Appendix for this particular pathology. Hand-assisted laparoscopy allows a hand to aid in dissection intraperitoneally, providing tactile sensation, as well as a less traumatic method to grasp tissues than with instruments [16]. This potentially decreases the risk of mucocèle rupture, particularly for cases as the one presented with

large cystic masses with distended and tenuous walls and distorted anatomy. Robotic-assisted surgery allows for improved dexterity, with wrist-like motion of the instruments and a three-dimensional camera [17], again offering the potential for a safer resection. With these minimally invasive approaches, both patients in the current report were able to undergo successful appendectomies, with negative margins and no disruption of the mucocoeles.

4. Conclusion

In conclusion, appendiceal mucocoele is a nonspecific term that encompasses a range of diagnoses, including neoplastic and non-neoplastic causes of appendiceal dilation and mucin production. Resection is recommended for mucocoeles due to their potential for malignancy. It is imperative to maintain the integrity of the mucocoele wall to prevent the risk of pseudomyxoma peritonei. Although laparoscopy has traditionally been considered contraindicated for appendectomy for mucocoeles due to the risk of perforation, we have presented two alternative minimally invasive approaches: hand-assisted laparoscopy and robotic-assisted laparoscopy. These approaches afford similar benefits to laparoscopy, while being potentially safer and allowing for the effective surgical treatment for patients with appendiceal mucocoeles.

Conflicts of interest

No conflicts of interest.

Funding

No funding.

Ethical approval

Our institution does not require institutional review board or other approval for small-volume case series which are deidentified and collected retrospectively (University of South Florida IRB policy #311 allows for the unique reporting of 3 patients or less without IRB approval).

Consent

Written informed consent was obtained from both patients for this study. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request. The information in this case report is completely de-identified.

Author contribution

Conception or design of work: Orcutt, Malafa.

Acquisition, analysis, or interpretation of data for the work: Orcutt, Anaya, Malafa.

Drafting the work or revising it critically for important intellectual content: Orcutt, Anaya, Malafa.

Final approval of the version to be published: Orcutt, Anaya, Malafa.

Agreement to be accountable for all aspects of the work: Orcutt, Anaya, Malafa.

Registration of research studies

Researchregistry.com UIN researchregistry2368.

Guarantor

Mokenge Malafa MD.

References

- [1] B. Rymer, R.O. Forsythe, G. Husada, Mucocoele and mucinous tumours of the appendix: a review of the literature, *Int. J. Surg.* 18 (2015) 132–135.
- [2] S. Dhage-Ivatury, P.H. Sugarbaker, Update on the surgical approach to mucocoele of the appendix, *J. Am. Coll. Surg.* 202 (2006) 680–684.
- [3] K.J. Park, H.J. Choi, S.H. Kim, Laparoscopic approach to mucocoele of appendiceal mucinous cystadenoma: feasibility and short-term outcomes in 24 consecutive cases, *Surg. Endosc.* 29 (2015) 3179–3183.
- [4] H. Lau, W.K. Yuen, F. Loong, F. Lee, Laparoscopic resection of an appendiceal mucocoele, *Surg. Laparosc. Endosc. Percutan. Tech.* 12 (2002) 367–370.
- [5] Y.T. Ju, S.T. Park, W.S. Ha, et al., Laparoscopic resection of a appendiceal mucocoele, *J. Korean Surg. Soc.* 80 (Suppl. 1) (2011) S21–25.
- [6] K.B. Park, J.S. Park, G.S. Choi, et al., Single-incision laparoscopic surgery for appendiceal mucocoeles: safety and feasibility in a series of 16 consecutive cases, *J. Korean Soc. Coloproctol.* 27 (2011) 287–292.
- [7] M. Rangarajan, C. Palanivelu, A.J. Kavalakat, R. Parthasarathi, Laparoscopic appendectomy for mucocoele of the appendix: report of 8 cases, *Indian J. Gastroenterol.* 25 (2006) 256–257.
- [8] R.A. Agha, A.J. Fowler, A. Saeta, et al., The SCARE Statement: consensus-based surgical case report guidelines, *Int. J. Surg.* 34 (2016) 180–186.
- [9] R.A. Agha, A.J. Fowler, S. Rajmohan, I. Barai, D.P. Orgill, P. Group, Preferred reporting of case series in surgery: the PROCESS guidelines, *Int. J. Surg.* 36 (2016) 319–323.
- [10] K.K. Turaga, S.G. Pappas, T. Gamblin, Importance of histologic subtype in the staging of appendiceal tumors, *Ann. Surg. Oncol.* 19 (2012) 1379–1385.
- [11] N.J. Carr, T.D. Cecil, F. Mohamed, et al., A consensus for classification and pathologic reporting of pseudomyxoma peritonei and associated appendiceal neoplasia: the results of the Peritoneal Surface Oncology Group International (PSOGI) modified delphi process, *Am. J. Surg. Pathol.* 40 (2016) 14–26.
- [12] P.J. Pickhardt, A.D. Levy, C.A. Rohrmann Jr., A.I. Kende, Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation, *Radiographics* 23 (2003) 645–662.
- [13] S.A. Zanati, J.A. Martin, J.P. Baker, C.J. Streutker, N.E. Marcon, Colonoscopic diagnosis of mucocoele of the appendix, *Gastrointest. Endosc.* 62 (2005) 452–456.
- [14] T.C. Chua, B.J. Moran, P.H. Sugarbaker, et al., Early- and long-term outcome data of patients with pseudomyxoma peritonei from appendiceal origin treated by a strategy of cytoreductive surgery and hyperthermic intraperitoneal chemotherapy, *J. Clin. Oncol.* 30 (2012) 2449–2456.
- [15] S. Gonzalez Moreno, B.M. Shmookler, P.H. Sugarbaker, Appendiceal mucocoele: Contraindication to laparoscopic appendectomy, *Surg. Endosc.* 12 (1998) 1177–1179.
- [16] H. Moloo, F. Hagggar, D. Coyle, et al., Hand assisted laparoscopic surgery versus conventional laparoscopy for colorectal surgery, *Cochrane Database Syst. Rev.* (2010) CD006585.
- [17] S.K. Baek, J.C. Carmichael, A. Pigazzi, Robotic surgery: colon and rectum, *Cancer J.* 19 (2013) 140–146.

Open Access

This article is published Open Access at [sciencedirect.com](https://www.sciencedirect.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.