Appendiceal Mucocele in A 54-Year-Old Woman: A Case Report

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Abstract

Introduction: Appendiceal mucocele is a clinically and pathologically rare condition. Mucinous secretions obstruct the lumen of the appendix leading to dilation.

Case presentation: A 54-year-old female presented with right lower quadrant pain for three days. The pain was described as cramping, with nausea and vomiting following it. The imaging studies revealed a well-circumscribed mass with a smooth thin wall, without calcifications arising from the inferomedial aspect of the caecum, highly suggestive of an appendiceal mucocele. An exploratory laparotomy was performed on the patient, which revealed a cystic mass (6x4x4 cm) of the appendix with no peritoneal tumors, seedlings, or metastases and no sign of perforation or an abscess. A simple appendectomy was performed because no pathogenic process was discovered in the base of the appendix, the caecum, or the mesenteric lymph nodes. The patient recovered uneventfully. The final diagnosis was mucinous cystadenoma with free margins.

Discussion: Appendicular mucocele is a rare condition with a wide range of symptoms. Although abdominal imaging is a valuable diagnostic tool, histopathology is the gold standard for determining a conclusive diagnosis. The long-term prognosis for surgery for benign appendicular mucoceles is excellent.

Keywords: Appendicitis; Mucocele; Mucinous cystadenoma; Appendix; Appendectomy.

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I. Introduction

Appendiceal mucocele (AM) is a rare disease from the appendix with a low incidence of 0,2-0,7% and higher occurrence in females and ages >50 years [1-3]. The appendix lumen is obstructed and dilated due to an intraluminal accumulation of the mucoid material. The clinical diagnosis is difficult because most patients are asymptomatic. Even if a patient has symptoms, they are nonspecific, and they mimic other diseases such as appendicitis and other gynecological entities [2, 4-6].

The clinical suspicion is crucial because perforation of an AM can lead to pseudomyxoma peritonei (PMP). PMP is a peritoneal malignancy treated by cytoreductive surgery with heated intraperitoneal chemotherapy. This neoplasm spreads rapidly to the peritoneum and rarely through the lymph nodes and the vessels. PMP outcomes are very poor, which means that the appropriate diagnosis and management before the perforation of the appendiceal mucocele is crucial [3-6].

Surgery is the only treatment of mucocele. The histologic types encompass mucinous retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. Mucinous cystadenomas are more common than mucinous cystadenocarcinomas [1, 2, 7].

II. Case presentation

A 54-year-old female visited the emergency department with a three-day history of abdominal pain localized in the right lower quadrant. The pain was described as cramping, associated with nausea and vomiting.

Her medical history was unremarkable. Body temperature was 37.6 °C. Physical examination revealed a soft, non-distended abdomen, with tenderness in the right lower quadrant and no evidence of peritonism. Digital rectal examination was normal.

Laboratory studies revealed elevated white blood cells and neutrophils (white blood cell count 14.1 K/Ul and neutrophils 89%) with normal C—reactive protein (3 mg/l). The Covid-19 test was negative. Abdominal ultrasound showed a cystic lesion in the right abdominal lower quadrant. Subsequently, the patient underwent a contrast-enhancement abdominal computed tomography, which revealed a well-circumscribed mass with a smooth thin wall, without calcifications arising from the inferomedial aspect of the caecum, highly suggestive of an appendicular mucocele.

The patient underwent an exploratory laparotomy, which revealed no evidence of peritoneal tumors, seedlings, or metastases. A cystic mass measuring $6 \times 4 \times 4$ cm was found in the right iliac fossa arising from the body of the appendix. There was no evidence of perforation or abscess formation. A simple appendectomy was performed because no pathologic process was found in the base of the appendix, the caecum, and the mesenteric lymph nodes. The patient recovered uneventfully, and she was discharged on the third postoperative day.

Histopathological examination confirmed the final diagnosis of a mucinous cystadenoma arising from the body of the appendix with free margins of resection. The patient remained well on regular follow-up visits over two years.

III. Discussion

Appendiceal mucocele (AM) is asymptomatic in 25% of the patients. The most common symptom is right lower quadrant pain, as in our patient. Other symptoms are palpable mass (50% of cases) and sporadic urinary symptoms [1, 3, 5, 7]. After the clinical examination, imaging is the next step. At first, an ultrasound shows the diameter of the appendix. Diameter > 15 mm is a threshold for diagnosis of mucocele (sensitivity 83%, specificity 92%). Also, ultrasound can differentiate benign and malignant mucoceles. The most critical imaging study to confirm the diagnosis and the disease extension is computed tomography, usually documenting a well-encapsulated cystic mass in the right lower abdominal quadrant, often associated with mural calcifications. Signs of ruptures such as mucinous ascites intraperitoneally are vital as a preoperative workup. For malignant mucoceles, further investigation with colonoscopy is essential for diagnosing synchronous or metachronous colon cancers [5-9].

The treatment for mucocele is surgery (conventional or laparoscopic) and may range from appendicectomy to right colectomy and cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy and early postoperative intraperitoneal chemotherapy. Conventional surgery is preferred to laparoscopic because there is a lower risk of rupture. Simple appendectomy is the surgery of choice in patients with benign mucocele with normal caecum and no evidence of perforation. Otherwise, a right hemicolectomy is recommended when malignant mucocele is suspected with signs of perforation, enlarged mesenteric lymph nodes or positive cytology. A thorough examination of the abdomen is recommended because of the well-known link between appendicular mucocele and other mucin-secreting cell malignancies, such as colon and ovarian cancers [1, 3, 7-11].

Pseudomyxoma peritonei (PMP) requires a multi-approach in referral centers because a high morbidity rate impacts these treatments. 5-year survival rates have been recorded, ranging from 50 to 96%. Based on prognosis, mucinous appendiceal neoplasms are divided as disseminated peritoneal adenomucinosis (least aggressive) and peritoneal mucinous adenocarcinoma (most aggressive) [8, 11, 12]. Patients with less aggressive cancer treated with cytoreductive surgery and perioperative intraperitoneal chemotherapy and achieve complete cytoreduction have a 70% survival rate after 20 years [9, 12, 13].

IV. Conclusion

Appendiceal mucocele is a rare condition with a wide range of symptoms. While ultrasound and CT scans of the abdomen are helpful diagnostic techniques, histology is required for a conclusive diagnosis. Surgery for benign appendicular mucocele has an excellent long-term prognosis.

Abbreviations:

AM: Appendiceal Mucocele

PMP: Pseudomyxoma Peritonei

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- 1. Stefanou SK: Study conception and design, drafting of manuscript.
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- 3. Tepelenis K: Literature search and acquisition of data.
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References

- Marudanayagam R, Williams GT, Rees BI. Review of the pathological results of 2660 appendicectomy specimens. J Gastroenterol. 2006;41(8):745-749. doi:10.1007/s00535-006-1855-5.
- [2]. Naraynsingh V, Hariharan S, Sammy I. Cecal volvulus and mucocele of the appendix. Acta Gastroenterol Latinoam. 2010;40(4):354-356.
- [3]. Jayakrishnan TT, Zacharias AJ, Sharma A, Pappas SG, Gamblin TC, Turaga KK. Role of laparoscopy in patients with peritoneal metastases considered for cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC). World J Surg Oncol. 2014;12:270. doi:10.1186/1477-7819-12-270.
- [4]. Morano WF, Gleeson EM, Sullivan SH, et al. Clinicopathological Features and Management of Appendiceal Mucoceles: A Systematic Review. Am Surg. 2018;84(2):273-281.
- [5]. Demetrashvili Z, Chkhaidze M, Khutsishvili K, et al. Mucocele of the appendix: case report and review of literature. Int Surg. 2012;97(3):266-269. doi:10.9738/CC139.1.
- [6]. Lien WC, Huang SP, Chi CL, et al. Appendiceal outer diameter as an indicator for differentiating appendiceal mucocele from appendicitis. Am J Emerg Med. 2006;24(7):801-805. doi:10.1016/j.ajem.2006.04.003.
- [7]. Saad EA, Elsamani EY, AbdElrahim WE, Elsiddig KE, Khalil EAG. Surgical treatment of mucocele of the appendix: a systematic review and case report. J Surg Case Rep. 2018;2018(6):rjy102. doi:10.1093/jscr/rjy102
- [8]. Pickhardt PJ, Levy AD, Rohrmann CA Jr, Kende AI. Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation [published correction appears in Radiographics. 2003 Sep-Oct;23(5):1340]. Radiographics. 2003;23(3):645-662. doi:10.1148/rg.233025134.
- Karakaya K, Barut F, Emre AU, et al. Appendiceal mucocele: case reports and review of current literature. World J Gastroenterol. 2008;14(14):2280-2283. doi:10.3748/wjg.14.2280.
- [10]. Dhage-Ivatury S, Sugarbaker PH. Update on the surgical approach to mucocele of the appendix. J Am Coll Surg. 2006;202(4):680-684. doi:10.1016/j.jamcollsurg.2005.12.003.
- [11]. Ronnett BM, Zahn CM, Kurman RJ, Kass ME, Sugarbaker PH, Shmookler BM. Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to "pseudomyxoma peritonei". Am J Surg Pathol. 1995;19(12):1390-1408. doi:10.1097/00000478-199512000-00006.
- [12]. Lorenzon L, De Dominicis C, Virgilio E, Balducci G. The appropriate management of an appendiceal mucocele. BMJ Case Rep. 2015;2015:bcr2014209045. doi:10.1136/bcr-2014-209045.
- [13]. Sugarbaker PH. New standard of care for appendiceal epithelial neoplasms and pseudomyxoma peritonei syndrome? Lancet Oncol. 2006;7(1):69-76. doi:10.1016/S1470-2045(05)70539-8.

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