



EVOLUTION OF MANAGEMENT OF MUCOCELE OF APPENDIX OVER TIME- CASE SERIES AND REVIEW OF LITERATURE

General Surgery

Dr. Priya Gupta* Fellow, Zen Multispeciality Hospital, Chembur, Mumbai *Corresponding Author

Dr. Vishakha Kalikar Junior consultant, Zen Multispeciality Hospital, Chembur, Mumbai

Dr. Prasad Bhukebag Junior consultant, Zen Multispeciality Hospital, Chembur, Mumbai

Dr. Roy Patankar Professor, Zen Multispeciality Hospital, Chembur, Mumbai

KEYWORDS

INTRODUCTION:

Mucocele of the appendix occurs due to obstruction of lumen of appendix resulting in abnormal accumulation of mucous in the lumen, giving the appendix an enlarged appearance with thinned out wall. Preoperative clinical diagnosis may be challenging due to non-specific clinical manifestations of the disease or may mimic acute appendicitis. Radiological imaging is essential for diagnosis. When diagnosed pre-operatively, it is always treated by surgery which depends on size and integrity of the appendix base and on the histology of the lesion. During surgery, care should be taken to avoid spillage of its contents into the abdomen which may lead to pseudomyxoma peritonei (PP), which has very poor prognosis.

METHODS:

This is a retrospective study carried out at a tertiary care multispeciality hospital in Mumbai. Four hundred and sixty laparoscopic appendicectomy surgeries were carried out at our centre from February 2018 to February 2023, out of which 4 patients had preoperative diagnosis of mucocele on radiology, which was confirmed on histopathology examination.

RESULTS:

All 4 patients presented with complaints of chronic lower abdominal pain and blood reports showed mild leucocytosis. Ultrasound of the abdomen reported presence of blind ending dilated tubular structure which was non-peristaltic, non-compressible and non-vascular arising from cecum with diameter of 3-5 cm and 8-10 cm in length in right lumbar region with probe tenderness. Contrast-enhanced computed tomography (CE-CT) was done in patients with suspicion of mucocele on ultrasound, which was suggestive of irregular wall thickening of base of appendix (maximum width 20-26 mm) showing post-contrast enhancement. Dilatation of distal appendix was seen (measuring 23 mm in its maximum diameter) with few intraluminal air foci within. Minimal intraluminal oral contrast opacification was seen in proximal part of appendix with no oral contrast opacification of distal part with no periappendiceal fat stranding or collection. Irregular thickening of cecum at the base of appendix was observed likely suggestive of appendicular mucocele involving adjacent cecal wall (Figure 1).

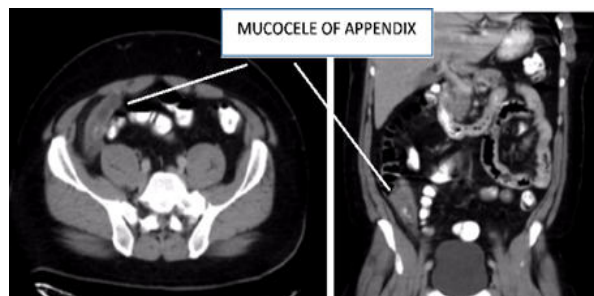


Figure 1: Mucocele of appendix on axial and coronal CT scan

Operative Details:

All patients were operated by laparoscopic appendicectomy, using standard laparoscopic ports, by the same surgeon, at the same centre.

Intra-operatively inflamed, enlarged, and edematous appendix was seen (Figure 2). In two patients out of four, the tumor could be palpated distally (within the tip of appendix) with normal base, while in other 2 patients, base of appendix was also edematous and dilated and the tumor could be palpated at the base of appendix (Figure 2).

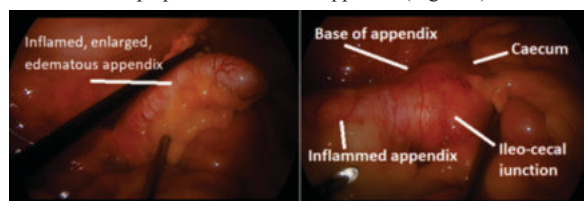


Figure 2: Inflamed appendix with enlarged base.

Laparoscopic appendicectomy was done in all 4 patients. Results were as follows: (Table 1)

Table 1: Results of laparoscopic appendicectomy

Patients	Base of Appendix Involved	Frozen Section/ Histopathology	Surgery Performed
1.	No	LAMN- Tip of Appendix	LA
2.	No	LAMN- Tip of Appendix	LA
3.	Yes	LAMN- Resection margin free	LA with cuff of caecum resection
4.	Yes	LAMN- Lymph node positive	LA f/b right hemicolectomy

In 2 patients where base of appendix was involved, laparoscopic appendicectomy was done including resection of 2 cm margin of cecal wall using ultrasonic shears, keeping the ileo-cecal valve intact. Intracorporeal anastomosis of cecum was done in 2 layers using silk. Specimen was extracted through a 2.5 cm grid-iron incision in right iliac fossa, using a locally made plastic wound protector, taking care not to spill the contents. Mucous was seen extruding from the tumour in the specimen (Figure 3).

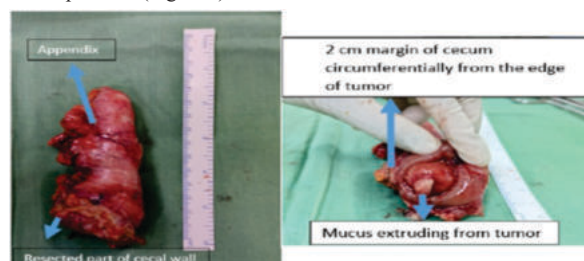


Figure 3: Specimen including appendix and part of cecal wall, showing tumor and mucinous exudate from the tumor

Frozen section was sent for all 4 patients which showed low grade mucinous neoplasm with free margins. Specimen was sent for histopathology examination.

Histopathology report suggested mucin secreting epithelium with areas of mucosal ulceration, mild to moderate cytologic atypia of epithelium. Foci of dystrophic calcification were seen within the mucin pools. The colonic margin of resection was free of tumor and extracellular mucin. Pools of acellular mucin were seen over the ulcerated luminal surface, dissecting up to the muscularis propria, with no evidence of extravasated mucin in the subserosal adipose tissue in all four patients and a diagnosis of Low grade Appendiceal Mucinous Neoplasm (LAMN) was made.

In one patient, lymphnodes of mesoappendix were positive, hence decision was taken to do right hemicolectomy subsequently after 2 weeks.

Post-operatively, patient with appendicectomy were started on liquid diet after 6 hours and soft diet the following day and discharged on post-operative day 1.

Patients with cecal cuff resection and right hemicolectomy were started orally on liquids from day 3, soft diet from day 4 and discharged on day 5.

All 4 patients were routinely followed up till 6 months. There were no post-operative complications and no recurrence in 6 months follow-up period.

DISCUSSION:

Mucocele of appendix is a pathological condition which occurs due to mucosal inflammatory or neoplastic diseases that result in the formation of an obstructive, distended appendix due to intraluminal accumulation of mucous. Rokitsansky first reported mucocele of the appendix in 1842⁽¹⁾. It is about twice more common in females, and is reported in only 0.2%-0.7% patients on histopathological examination of appendicectomy specimens⁽²⁻⁴⁾. According to the World Health Organization, there are four formations that result in mucocele of the appendix: simple mucocele or retention mucocele, mucosal hyperplasia (5%-25%), mucinous cystadenoma (63%-84%), mucinous cystadenocarcinoma (11%-20%)⁽⁵⁻⁸⁾.

Mucocele of the appendix usually presents clinically with non-specific symptoms, pain in right lower quadrant being the most common symptom among symptomatic patients⁽⁹⁾. Other symptoms may include lump in abdomen, weight loss, nausea, vomiting, altered bowel habits, unexplained anemia, and lower gastrointestinal bleeding⁽⁹⁾. Rarely it may also present as intussusception or bowel obstruction⁽¹⁰⁾. If appendiceal mucocele ruptures spontaneously, it may present as acute appendicitis, leading to PP syndrome.

Though pre-operatively, mucocele may be diagnosed with Ultrasonography, CT scan, or Magnetic Resonance Imaging (MRI); diagnosis is confirmed intraoperatively due to macroscopic appearance and a definite diagnosis can only be made after histopathological examination of the specimen⁽¹¹⁻¹³⁾. On CE-CT, features differentiating mucocele from acute appendicitis include cystic dilatation of the appendix, mural calcification, and a luminal diameter more than 1.3 cm⁽¹⁴⁾. In this study, mucocele of appendix was anticipated in all four patients on the basis of radiological findings, diagnosed intra-operatively, and confirmed on frozen section and histopathological examination.

Surgical management for mucocele of appendix has evolved over decades. The key principle in surgical management of mucinous tumors of appendix is the need of excising all mesoappendix including appendix and its lymph nodes⁽¹¹⁾.

An algorithm was created by Dhage-Ivatury and Sugarbaker in 2005 for the selection of the type of surgery, wherein it was proposed that when a mucocele was identified at the time of laparoscopy, the definitive maneuver was to convert to an open laparotomy, even if it was possible to remove it intact without trauma⁽¹⁵⁾.

Miraliakbari et al have described one of the initial laparoscopic surgeries done for mucocele of appendix, using an endobag for retrieval of specimen to prevent port-site implantation⁽¹⁶⁾.

To determine the surgical extent, base of the appendix is checked during surgery. In an unruptured appendix, if lesion is palpated near the base of the appendix, a cuff of caecum is resected along with the

appendix and included in specimen without compromising the ileocaecal valve and it ensures complete resection⁽¹⁷⁾. In case of involvement of base and when a clear margin is unavailable, a partial caecectomy, ileocectomy, or right colectomy can be done following the oncological principles with high ligation of ileocolic pedicle at its origin⁽¹⁷⁾. Appendicectomy with lymphadenectomy is the definite treatment in simple and benign mucocele. If the histological diagnosis reports bowel or mucinous adenocarcinoma, carcinoid or adenocarcinoid tumor greater than 2.0 cm, with positive nodes or high mitotic rate, the resection is complemented with right colectomy^(18,19).

In our study, all four patients were operated laparoscopically and the specimen was removed using endobag to prevent port site implantation. Due to involvement of base of appendix and cecum, alongwith appendicectomy, a cuff of cecum was resected and closure of cecum was done laparoscopically to prevent the dissemination of contents into the abdominal cavity.

If lesion has ruptured, findings of peritoneal cavity spillage should be documented and patient should be treated by cytoreductive surgery and heated intraperitoneal chemotherapy (HIPEC)⁽²⁰⁾.

Although many authors advocate that laparoscopic appendicectomy in cases of mucocele is not indicated due to the manipulation and risk of appendicular rupture and the dissemination of mucus and possibility of peritoneal implants⁽¹³⁾, we propose that laparoscopic appendicectomy with cecal cuff resection (if required), is a feasible option of mucinous neoplasms of appendix and may not need conversion to laparotomy if extreme care is taken to prevent spillage of contents.

CONCLUSION:

Radiological suspicion of mucocele of appendix based on USG and CT-scan necessitates frozen section biopsy during appendicectomy. Low grade mucinous tumors can safely be managed with cecal resection, open or laparoscopically. Presence of enlarged lymph nodes, or spillage of mucous or mucinous adenocarcinoma necessitates right hemicolectomy. In absence of lymph node enlargement and negative frozen section report, cecal resection can be considered as completion surgery.

REFERENCES:

- Rokitansky C.F. Vol. 2. Blancard and Lea; Philadelphia: 1855. p. 89. (A Manual of Pathological Anatomy). English translation of the Vienna edition (1842).
- Woodruff R, Mc Donald J. Benign and malignant cystic tumors of the appendix. *Surg Gynecol Obstet*. 1940;71:751-755.
- Emre A, Akbulut S, Bozdog Z, Yilmaz M, Kanlioz M, Emre R, et al. Routine histopathologic examination of appendectomy specimens: retrospective analysis of 1255 patients. *Int Surg*. 2013;98:354-362. <https://doi.org/10.9738/INTSURG-D-13-00098.1>.
- Yilmaz M, Akbulut S, Kutluturk K, Sahin N, Arabaci E, Ara C, et al. Unusual histopathological findings in appendectomy specimens from patients with suspected acute appendicitis. *World J Gastroenterol*. 2013;19:4015-4022. <https://doi.org/10.3748/wjg.v19.i25.4015>.
- Tărcoveanu E, Vasilescu A, Hee RV, Moldovanu R, Ursulescu C, Ciobanu D, et al. Appendicular Mucocele: Possibilities and Limits of Laparoscopy. Brief Series and Review of the Literature. *Chirurgia (Bucur)* 2015;110:530-537.
- Kılıç MÖ, İnan A, Bozer M. Four mucinous cystadenoma of the appendix treated by different approaches. *Türk J Surg*. 2014;30:97-99. <https://doi.org/10.5152/UCD.2014.2397>.
- Higa E, Rosai J, Pizzimbono CA, Wise L. Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendiceal "mucocele" *Cancer*. 1973;32:1525-1541.
- Pai RK, Longacre TA. Pseudomyxoma peritonei syndrome: classification of appendiceal mucinous tumours. *Cancer Treat Res*. 2007;134:71-107. https://doi.org/10.1007/978-0-387-48993-3_5.
- Ryder B, Forsythe RO, Husada G. Mucocele and mucinous tumours of the appendix: A review of the literature. *Int J Surg*. 2015;18:132-135. <https://doi.org/10.1016/j.ijsu.2015.04.052>.
- Sun P, Jiang F, Sun H, Zhao X, Ma J, Li C, et al. Minimally invasive surgery for appendiceal intussusceptions caused by mucocele of appendix: Case report and review of the literature *J Gastrointest Oncol*. 2020;11:102-7.
- Kılıç MÖ, İnan A, Bozer M. Four mucinous cystadenoma of the appendix treated by different approaches. *Türk J Surg*. 2014;30:97-99. <https://doi.org/10.5152/UCD.2014.2397>.
- Malva FU, Hasbahceci M, Sertar A, Cipe G, Karatepe O, Kocakoc E, et al. Appendiceal mucocele: clinical and imaging features of 14 cases. *Chirurgia (Bucur)* 2014;109:788-793.
- Stocchi L, Wolff BG, Larson DR, Harrington JR. Surgical Treatment of Appendiceal Mucocele. *Arch Surg*. 2003;138:585-590.
- Bennett, Genevieve L.; Tanpitukpongse, Teerath P.; Macari, Michael; Cho, Kyunghye C.; Babb, James S. (2009). *CT Diagnosis of Mucocele of the Appendix in Patients with Acute Appendicitis*. *American Journal of Roentgenology*. 192(3), W103-W110. doi:10.2214/AJR.08.1572
- Dhage-Ivatury, S., & Sugarbaker, P. H. (2006). *Update on the Surgical Approach to Mucocele of the Appendix*. *Journal of the American College of Surgeons*, 202(4), 680-684. doi:10.1016/j.jamcollsurg.2005.12.003.
- MIRALIAKBARI, REZA; CHAPMAN, W.H.H. (1999). *Laparoscopic Treatment of an Appendiceal Mucocele*. *Journal of Laparoendoscopic & Advanced Surgical Techniques*, 9(2), 159-163. doi:10.1089/lap.1999.9.159
- Jandui Gomes et al, Mucocele of the appendix: appendectomy or colectomy. *J. Coloproctol.* (Rio J) 31 (3) Sept 2011. <https://doi.org/10.1590/S2237-9363.20110003.00008>.

18. Pahlavan PS, Kanthan R. Goblet cell carcinoid of the appendix. *World J Surg Oncol* 2005;3(36):1-11.
19. Fornaro R, Frascio M, Sticchi C, De Salvo L, Stabilini C: Appendectomy or right hemicolectomy in the treatment of appendiceal carcinoid tumors? *Tumori* 2007;93(6):587-90.
20. Loungnarath R, Causeret S, Bossard N, Faheez M, Sayag-Neaujard AC, Brigand C, et al Cytoreductive surgery with intraperitoneal chemohyperthermia for the treatment of pseudomyxoma peritonei: A prospective study *Dis Colon Rectum*. 2005;48:1372-9