ORIGINAL RESEARCH PAPER

General Surgery

APPENDICEAL MUCOCELE: CASE SERIES AND BRIEF REVIEW

KEY WORDS:

Dr. Sunil Kumar B. B. Associate Professor , Department Of Surgical Gastroenterology , Jss Hospital , Mysuru

Dr. Mahesh S. Shetty Associate Professor , Department Of Surgical Gastroenterology , Jss Hospital , Mysuru

Dr. Ankit Raj* Postgraduate , Department Of General Surgery, Jss Hospital , Mysuru
*Corresponding Author

ABSTRACT

INTRODUCTION: Appendiceal mucocele is a rare condition which has a clinical picture resembling acute appendicitis. A correct diagnosis before surgery is important for the selection of surgical technique which helps in avoiding intraoperative and postoperative complications.

MATERIALS AND METHODS: In our case series, we present three isolated cases of appendiceal mucocele complicated by the development of acute inflammation of the appendix.With the help of Ultrasonography followed by Computed Tomography preoperative diagnosis of Appendiceal Mucocele was made. For two of the cases patients were directly taken up for midline incision Laparotomy and for one case patient initially underwent laparoscopic appendicectomy and after histopathological confirmation of adenocarcinoma , patient was taken up for a midline incision laparotomy .The mucoceles were removed without rupture, and all patients recovered well postoperatively without any complications.

DISCUSSION: Mucocoele of the appendix is a rare diagnosis. However, given the possibility of neoplastic peritoneal dissemination, it may be considered as a differential diagnosis, especially in elderly patients with non-specific symptoms similar to appendicitis. Diagnosis relies on CT, tumour markers and intra-operative findings. Treatment is surgical resection. Correct initial treatment can prevent iatrogenic progression to pseudomyxoma peritonei in malignant cases.

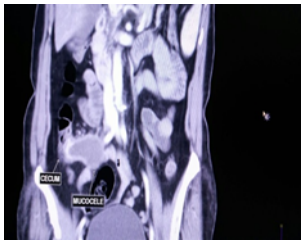
INTRODUCTION-

Appendiceal mucocele(AM) is a rare diagnosis, accounting for less than 1% of appendiceal pathologies . The term "mucocele" describes a distended Appendix whose lumen is filled with mucus but does not represent a pathological description. AM are diagnosed incidentally about 50% of the time [1] , mostly via imaging for synchronous malignancies or benign conditions. Symptoms may include abdominal pain or mass, weight loss, nausea, or a change in bowel habits [2] . Patients with a mucocele may present with associated appendicitis, intussusception, appendiceal torsion, gastrointestinal bleeding, or increasing abdominal girth from rupture of a neoplastic mucocele . Ideally , resection is recommended for mucoceles, as imaging may not confirm which mucoceles are associated with malignancy. However, precautionary measures should be taken while performing resection as the rupture of a neoplastic mucocele can give rise to pseudomyxoma peritonei, or the deposition of mucin throughout the peritoneal cavity, resulting in significant morbidity from mucinous ascites and bowel obstruction [1] . Hence open resection through laparotomy has been recommended [2] .

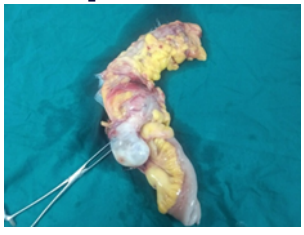
PRESENTATION OF CASES

CASE 1

A 70 year old female presented to our OPD with 1 month history of pain abdomen, more localized to right lower abdomen, associated with generalized weakness, nausea and decreased appetite from last 6 months and no history of surgeries in the past. Patient reported mild right iliac fossa tenderness on palpation. She was afebrile. Laboratory investigations showed Leucocytosis with neutrophilia. Abdominal Ultrasonography showed encapsulated cystic lesion in the lower quadrant of the abdomen with a liquid content of variable echogenicity -? Appendicular Abscess /Mucocoele Appendix. CECT abdomen and pelvis showed well circumscribed low attenuating tubular mass contiguous with the base of the caecum showing thin curvy linear mural calcifications with few low attenuating areas along the surface of the lesion f/s/o Mucocele of Appendix. Vertical Midline Incision Laparotomy was performed. Intraoperatively a cystic mass of appendix with dimensions 8 cm × 5 cm with broad base and inflamed walls communicating with caecum but without perforation was discovered in right iliac fossa. Multiple significant lymph nodes of mesoappendix and ileocolic region were seen. With suspicion of malignancy and non-availability of frozen section, Right hemicolectomy with ileotransverse anastomosis was done Histopathologically Mucinous Cystadenoma with Mucocele was reported. After 6 months of surgery patient is doing well with no postoperative complications.



1.CECT Abdomen and pelvis

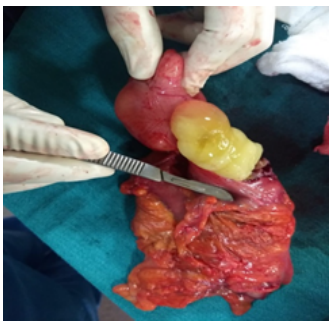


2. Hemicolectomy specimen showing mucocele

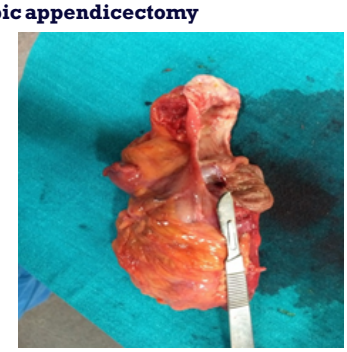
CASE 2

A 65 year old male presented to our OPD with complaints of pain abdomen on and off since past 3 months in the right hypochondriac and right iliac fossa region since past 3 months which had gradually increased in the past 10 days.

Patient gave history of fever and vomiting since past 1 week which was intermittent and associated with decreased appetite from last 3 months . Patient did not give history of any other previous surgeries. On examination patient had tenderness in the right iliac fossa with voluntary guarding . Blood investigations showed leukocytosis with increased neutrophils . Ultrasonography showed a cystic lesion in the lower quadrant of the abdomen with heteroechogenicity alongwith cholelithiasis . Patient was followed up preoperatively with CECT abdomen and pelvis which showed a circumscribed mass in the right iliac fossa near the base of caecum of maximum diameter of 35 mm mostly s/o Mucocoele of appendix . Patient underwent laparoscopic cholecystectomy with appendicectomy in the same sitting and specimen was sent for histo pathological examination . Histopathology confirmed the cystic lesion as mucin secreting adenocarcinoma (pT2NxMx). After due consent patient was taken up for a Midline Incision Laparotomy and a right hemicolectomy was performed. Histopathological examination confirmed free resected margins . Patient is now asymptomatic post 1 year of surgery .



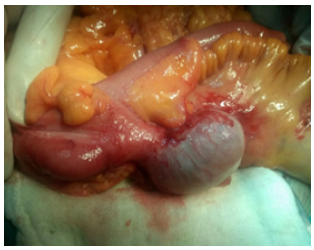
3.Showing mucinous material from the appendix post laparoscopic appendicectomy



4.Showing same specimen after removing mucinous material

CASE 3.

A 70 year old male patient presented to our opd with c/o pain abdomen since past 4 months more on the right side on and off , non radiating with incomplete evacuation of stools and increased frequency of micturition. Patient also gave history of single episode of dark colored stools 1 month back . Colonoscopy was done which showed a benign colonic polyp . Patient gave history of completing Antitubercular therapy 6 months back and underwent laparoscopic cholecystectomy 5 years back . On examination tenderness was present in the right iliac fossa with no guarding / rigidity .Ultrasound showed a cystic lesion in the appendix measuring approximately 3x2 cm ? abscess/ mucocoele of appendix . Hence CECT Abdomen and pelvis was done which showed tubular smooth walled cystic lesion with blind ending arising from the caecum , measuring 30 mm in maximum diameter with no post contrast enhancement alongwith tiny specks of peripheral wall calcification suggestive of Mucocoele of Appendix . CEA was found to be normal. Patient was taken up for Midline laparotomy with limited ileocaecal resection with side to side anastomosis. Intraoperatively a distended Histopathological examination confirmed the specimen as a benign mucinous lesion of appendix . 1year post surgery patient is asymptomatic with no complaints .



5.Intraoperative photo of specimen with mucocele of appendix

DISCUSSION:

Mucocele of the appendix is a rare entity, with a reported incidence ranging from 0.07 to 0.3% of all appendectomies and a female-to-male ratio of 3–4 to 1 .The external appearance is gross enlargement of an appendix whose lumen is distended by mucin. The causes are heterogeneous and include retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma . The important point is that all appendiceal neoplasams, both histologically benign and malignant, may result in diffuse mucinous peritoneal tumors. These mucinous peritoneal tumors carry high mortality even with extensive treatment. [3] In this case series, we present three patients with appendiceal mucoceles, all of which were identified incidentally. Each patient's Mucoceles represent a variety of histologies, both neoplastic and non-neoplastic, that can result in a dilated, mucus-filled appendix . Historically, mucoceles are described as arising from four different processes. Benign causes of mucoceles 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 mucoceles, hyperplasia and mucinous cystadenomas each represent about 33%, and mucinous cystadenocarcinomas represent 5%–10% [4] . Despite their rarity, mucinous cystadenocarcinomas account for 37% of appendiceal neoplasms [5] . There is a female predominance to cystadenocarcinomas (as opposed to other appendiceal neoplasms), and diagnosis is typically in the seventh decade [4] .There have been inconsistencies in the literature in the description and reporting of mucinous appendic ealneoplasia and pseudomyxomaperitonei. A recent international Delphi consensus was convened to better clarify and standardize these definitions . The consensus for appendicealntumors 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 . The term "**pseudomyxomaperitonei**" 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 [6]USG is the first-line diagnostic method for patients with acute abdominal pain. USG can be used to differentiate between mucocele and acute appendicitis. In case of acute appendicitis, the outer diameter threshold of the appendix is 6 mm, and 15 mm and more indicates the presence of a mucocele, with 83% sensitivity and 92% specificity [7 8]. CT is regarded as the most accurate method of diagnostics. CT can be used to discover the signs specific to mucocele with

high accuracy: appendix lumen more than 1.3 cm, its cystic dilatation, and wall calcification. By colonoscopy an elevation of the appendiceal orifice is seen and a yellowish mucous discharge would be visible from this orifice. One of the cardinal principles of surgical treatment of this disease is that intact mucoceles do not pose a threat for the patient. If it is perforated and the filling turns up in the peritoneal cavity, there is a high probability that pseudomyxoma peritonei will develop, for which treatment is very problematic and long-term results are quite unsatisfactory. Therefore, the selection of an adequate surgical method is very important .Resection is recommended when identified (whether incidentally or not) due to the potential for the mucocele to harbor a neoplasm. It is imperative during resection to avoid rupture of the mucocele 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% [5] , although once it has progressed to pseudomyxoma peritonei, survival is reported at 23% [9] . However, with newer techniques, including the use of cytoreductive surgery and heated intraperitoneal chemotherapy, 10-year survival can improve to close to 50% [10] . Appendectomy is recommended for non-neoplastic causes of mucocele 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 does not offer any survival benefit .

CONCLUSION :

Mucocele of the appendix is a descriptive term for mucinous distension of the appendiceal lumen. It refers to the progressive retrograde dilatation of the vermiform appendix. Due to lack of specific signs or its quiet presentation, this condition is mostly diagnosed only at an advanced stage. Currently, the evaluation of appendiceal lesions depends mainly on Ultrasound which might be unable to identify the origin of the tumour. Given the significance of preoperative diagnosis for planning management, it is essential that this condition may be taken into account in the differential diagnosis, in order to increase the suspicion when assessing such patients and hence reduce delays in diagnosis and treatment .In our opinion every patient more than 50 years presenting with clinical symptoms of acute appendicitis must undergo CT. Despite recent research on the therapeutic strategies against appendiceal neoplastic disorders, surgical resection appears as the only potentially curative approach.

COMPLIANCEWITH ETHICAL STANDARDS

CONFLICT OF INTEREST

Authors declare that they have no conflict of interest.

DISCLAIMER

The authors are solely responsible for the data and the content of the paper. In no way, the Honorary Editor-in-Chief, Editorial Board Members, or the printer/publishers are responsible for the results/findings and content of this article.

INFORMED CONSENT

Informed consent was obtained from all individual participants included in the study. A copy of the consent is available for review on request .

REFERENCES:

1. Mastoraki, A., Sakorafas, G., Vassiliu, P., Contopoulou, C., & Arkadopoulos, N. (2016). Mucocele of the Appendix: dilemmas in differential diagnosis and therapeutic management. Indian journal of surgical oncology, 7(1), 86-90.
2. Moreno, S. G., Shmookler, B. M., & Sugarbaker, P. H. (1998). Appendiceal mucocele. Surgical Endoscopy, 12(9), 1177-1179. doi:10.1007/s004649900811
3. Demetrashvili, Z., Chkhaidze, M., Khutsishvili, K., Topchishvili, G., Javakhishvili, T., Pipia, I., & Qerqadze, V. (2012). Mucocele of the Appendix: Case Report and Review of Literature. International Surgery, 97(3), 266-269. doi:10.9738/cc139.1
4. Rymer, B., Forsythe, R. O., &Husada, G. (2015). Mucocoele and mucinous tumours of the appendix: a review of the literature. International Journal of Surgery, 18, 132-135.
5. Turaga, K. K., Pappas, S. G., &Gamblin, T. C. (2012). Importance of histologic subtype in the staging of appendicealtumors. Annals of surgical oncology, 19(5), 1379-1385.
6. Carr, N. J., Cecil, T. D., Mohamed, F., Sobin, L. H., Sugarbaker, P. H., González-Moreno, S., ...& Moran, B. J. (2016). A consensus for classification and pathologic reporting of pseudomyxomaperitonei and associated appendicealneoplasia. The American journal of surgical pathology, 40(1), 14-26.
7. Lien, W. C., Huang, S. P., Chi, C. L., Liu, K. L., Lin, M. T., Lai, T. I., ... & Wang, H. P. (2006). Appendiceal outer diameter as an indicator for differentiating appendicealmucocele from appendicitis. The American journal of emergency medicine, 24(7), 801-805.
8. Francica, G., Lapiccirella, G., Giardiello, C., Scarano, F., Angelone, G., De Marino, F., &Molese, V. (2006). Giant mucocele of the appendix: clinical and imaging findings in 3 cases. Journal of ultrasound in medicine, 25(5), 643-648
9. Rymer, B., Forsythe, R. O., &Husada, G. (2015). Mucocoele and mucinous tumours of the appendix: a review of the literature. International Journal of Surgery, 18, 132-135.
10. Chua, T. C., Moran, B. J., Sugarbaker, P. H., Levine, E. A., Glehen, O., Gilly, F. N., ... &Liauw, W. (2012). Early-and long-term outcome data of patients with pseudomyxomaperitonei from appendiceal origin treated by a strategy of cytoreductive surgery and hyperthermicintraperitoneal chemotherapy. Journal of Clinical Oncology, 30(20), 2449-2456.