

Appendiceal mucocele : Case Report And Review of Literature

^{1*}Rajneesh Kumar, ²Ankur Hastir, ³Ramandeep Singh Walia

Corresponding author: ^{1*}Rajneesh Kumar *

Abstract: Mucocele of Appendix occurs when there is abnormal accumulation of mucus within the appendix leading to dilation of appendiceal lumen by mucus. The disease is considered as a rare lesion of the appendix, which is found in 0.2 to 0.3 % of the appendectomies. The course and prognosis of mucocele related to the histological subtypes. Preoperative diagnosis is difficult due to non-specific clinical manifestation and 50% are discovered incidentally at surgery. Patient may present with pain in lower right quadrant of abdomen and surgeon may mistake it for acute appendicitis. A combination of sonographic (namely the onion skin sign) and CT findings may aid in the correct preoperative diagnosis of the mucocele of appendix by an expert radiologist. The most dreaded complication of benign or malignant mucocele is pseudomyxoma Peritonei, so whenever you suspect mucocele of appendix during surgery extra care should be taken while handling the specimen. Treatment is always surgery and depends on the integrity and size of the appendix base and on the histological type of the disease.

Keywords: mucocele, appendix, pseudomyxomaperitonei, sonography.

I. Introduction

Mucocele of appendix is an uncommon disorder which is often asymptomatic and it has 0.2% to 0.4% prevalence among appendectomies (1-4). The term mucocele is widely used in diagnosing both benign and malignant lesions, but specific criteria are being proposed for definitive diagnosis and surgical management of appendiceal mucocele (5). There are 4 histological types of appendiceal mucocele: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma (6,7). Mucocele is a generic descriptive term for cystic dilatation of the appendix lumen (7).

Mucinous cystadenoma and cystadenocarcinoma comprise most of the case (8). The neoplastic mucocele and retention cysts are clinically and radiologically difficult to differentiate, but malignant mucoceles have additional findings (8). If not treated properly, perforation of appendix may lead to extravasation of its contents (epithelial cells) into peritoneal cavity may lead to pseudomyxomaperitonei which has very poor prognosis and has a high mortality (9). Mucocele of the appendix was first described by Rokitsansky in 1842, (10). And formally named by Feren in 1876. The clinical presentation of a mucocele is usually non-specific and upto fifty percent are discovered incidentally at surgery (11). This is characterized by the dilation of the organ lumen with mucus accumulation, being more frequent among individuals aged 50 years or more. Some articles confirm its prevalence among women (12, 13), others demonstrate a higher incidence among women (8, 14). Patient presenting with pain in right iliac fossa may be mistaken for acute appendicitis, one of the most common surgical diseases (3, 9, 15, and 16). It is important to differentiate between these two pathologies before surgery i.e. Acute appendicitis and mucocele of appendix by USG or CT scan if lumen is grossly distended.

Case Report

We present a case presenting to the gynecologist where initial clinical findings and investigations suggest an ovarian cyst. 70yrs. Female presented in Gynae department with prolapse uterus and a mass in right lower quadrant with dull pain of 2 years duration. Ultrasonography (USG) showed an elongated cystic lesion measuring 8x2cm is seen in pelvis right side? Ovarian cyst (Figure: 1). CEA was within normal limits.



Figure: 1



Figure: 2

Patient was operated for prolapse uterus and vaginal hysterectomy was done by Gynecologist and in the same sitting abdomen was opened through Pfannenstiel incision for suspected ovarian cyst which could not be assessed per vaginally and on exploration of abdomen gynecologist realized that cystic swelling was arising from iliocecal junction. Subsequently call was send to surgeon and on intraoperative examination we found the cystic mass of appendix (10cm x 4.5cm) present in the right iliac fossa with tense walls, but without perforation. No discharge was present in the peritoneal cavity. A mucocele was suspected. (Figure: 3, 4) Appendectomy with part of caecum along with base of appendix was removed. No Para aortic lymphadenopathy was seen. Appendix was cut open and a yellow gel- like content was observed (Figure:4).Patient's postoperative course was uneventful, and she was discharged home on postoperative day 5.Six months after surgery patient was doing well with no symptoms.Histologically diagnosis was mucinous cystadenoma.Final pathological analysis showed distension of the appendix with the flattening of mucosa. The base of the caecum showed normal intestinal histology with no signs of mucus deposition or cellular atypical on permanent tissue sections.



Figure: 3



Figure: 4

II. Discussion

Accumulation of mucous secretion is slow and gradual,with no signs of infection inside the organ leading to dilatation of appendicular lumen. It results from the lumen obstruction in the appendix, which is secondary to epithelial proliferation, either benign or malignant of the appendix mucosa, or of lesions in the caecum, adjacent to the appendicealostium.Much less frequently, inflammatory or obstructive causes, to include appendicitis and obstruction by a fecolith or appendicolith, are the cause of mucocele formation.The differential

diagnosis of mucocele of the appendix includes mesenteric cyst, duplication cyst, right ovarian cyst and hydrosalpinx. Intra-luminal bubble of gas or an air-fluid level within mucocele suggests the presence of superinfection, which can occur in both benign and malignant mucoceles. Mucocele of appendix is a descriptive term for an appendix distended by mucus, secondary to mucinous cystadenoma (63%), mucosal hyperplasia (25%), mucinous cystadenocarcinoma (11%) and retention cyst, (17).

The epithelial lining of the appendix consists of abundant exocrine goblet cells, and thus most tumor types seen in appendiceal samples are mucus producing (18). Excessive production of mucus by adenomatous tumors leads to formation of a mucocele and is usually caused by the entrapment of mucus and characterized by invasion of mucus into the appendiceal wall (19). The carcinoembryonic antigen (CEA) level at preoperative may suggest malignancy in the appendix or in the colon (20,21). Imaging techniques Ultrasonography, Computed tomography (CT), and less often MRI have been described for preoperative diagnosis (8, 29). On USG typical cystic mass with variable internal echogenicity. The presence of an "onion skin sign" (Sonographic layering within a cystic mass) is considered a highly suggestive feature (22). Appendix diameter 15mm or more in USG examination has been determined as the threshold for appendiceal mucocele diagnosis with a sensitivity of 83% and a specificity of 92%. (23).

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. (3,15,16,24,25,26) Colonoscopic findings show "Volcano sign", the appendiceal orifice seen in the centre of a firm mound covered by normal mucosa or a yellowish, lipoma-like submucosal mass (27). Patient may present with right lower quadrant pain, change in bowel habits, per rectal bleeding or a palpable mass. Approximately 23-50% of patients are asymptomatic, with the lesions being discovered incidentally during surgery, radiological evaluation or endoscopic procedures (28,29,30). So preoperative diagnosis of appendiceal mucoceles are difficult because of lack of clinical symptomatology.

Surgical excision of mucocele of an appendix can be done either by laparotomy or laparoscopic surgery. Laparoscopically, careful handling of the specimen is recommended as spillage of contents can lead to pseudomyxomateritonei. This can be achieved by atraumatic handling of the appendix and use of endobags for removal of specimens. Conversion to laparotomy should be considered if the lesion is traumatically grasped or if the tumor clearly extends beyond the appendix or if there is evidence of malignancy such as peritoneal deposits, (31). Laparoscopic approach has an increased risk of rupture and subsequent pseudomyxomateritonei formation (32, 33, 34). Moreno et al (33) suggest conversion to an open appendectomy in case of mucocele when laparoscopic appendectomy is intended. Some authors still recommend a minimally invasive approach in selected patients for this rare entity (35, 36, 37). Involvement of caecum or adjacent organs is an indication for right hemicolectomy and thorough exploration of the gastrointestinal tract and ovaries (38). Complications for appendiceal mucoceles consist of inflammation, invagination, intussusception, obstruction, and bleeding or fistula formation (36). A right hemicolectomy is a frequently performed if a malignant cause is suspected based on imaging or on intraoperative frozen section (39). Right hemicolectomy was not performed in these cases because frozen section analysis at the time of surgery showed no malignant characteristics. Since the risk of developing an adenocarcinoma of the colon is six times greater in patients with a mucocele than in the general population, colonic surveillance is warranted in these cases (40). The most dreaded complication of benign or malignant mucocele is pseudomyxomateritonei, which is difficult to treat surgically or medically. It has an uncertain prognosis, with a 5-year survival rate between 53% and 75%. (41, 42).

An algorithm for selection of the type of surgery has been furnished by Dhage-Ivatury and Sugarbaker (43). It envisages several factors: (a) whether or not a mucocele is perforated; (b) whether the base of appendix (margins of resection) is involved in the process; and (c) whether there are positive lymph nodes of mesoappendix and ileocolic. As a result patients may require different operations: appendectomy to the right colectomy, including cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy, early postoperative intraperitoneal chemotherapy. In our patient mucocele was not perforated (no discharge into peritoneal cavity), there was no pathological process in the base of appendix (negative margins of resection), and the regional lymph nodes were negative. Therefore only appendectomy was done and there were no late complications. Complete abdominal exploration during intraoperative is indicated due to the occurrence of mucocele in synchrony with other tumors, like colon and ovaries. This is necessary when emergency surgery is done with no preoperative workup (44).

Conclusion

Mucocele of the appendix can mimic an adenexal cyst and is difficult to diagnose. In a female presenting with right iliac fossa mass and with no clinical indication of any gynecological pathology, an appendiceal origin should be considered in the differential diagnosis.

Appendectomy whether open or laparoscopic does not guarantee the removal of all neoplastic tissue, including extensions into surrounding tissue and lymph nodes in case of malignant pathology (45,46). USG or CT findings of a cystic mass in right iliac fossa, adherent to the caecum, in a patient who has not undergone appendectomy,

are highly suggestive of mucocele of appendix. Presence of mural calcification can be valuable clue to the diagnosis. The presence of low attenuation ascities with peritoneal implants is suggestive of associated pseudomyxomaperitonei (47, 48, 49). Appendicealmucocele presents a challenge to the surgeon who does not know the pathological diagnosis on the operative procedure. Appendectomy alone should be performed only on mucinous lesions that are confirmed to be non-neoplastic after biopsy. If appendectomy done, precautions should be taken to minimize the risk of seeding the peritoneal cavity with tumorous mucin during manipulation (50, 51).

Conflict of interest

There is no conflict of interest.

References

- [1]. Rangarajan M., Palanivelu C., Kavalakat A. J., Parthasarathi R. Laparoscopic appendectomy for mucocele of the appendix. Report 8 cases. *Indian J Gastroenterol.* 2006;25(5):256–257. [PubMed]
- [2]. Marudanayagam R., Williams G. T., Rees B. I. Review of the pathological results of 2660 appendectomy specimens. *J Gastroenterol.* 2006;41(8):745–749. [PubMed]
- [3]. Ruiz-Tovar J., Teruel D. G., Gastineires V. M., Dehesa A. S., Quindos P. L., Molina E. M. Mucocele of the appendix. *World J Surg.* 2007;31(3):542–548. [PubMed]
- [4]. Smeenk R. M., van Velthuysen M. L., Verwaal V. J., Zoetmulder F. A. Appendiceal neoplasms and pseudomyxomaperitonei: a population-based study. *Eur J SurgOncol.* 2008;34(2):196–201. [PubMed]
- [5]. Stocchi L, Wolff BG, Larson DR, et al. Surgical treatment of appendicealmucocele. *Arch Surg.* 2003;138(6):585-590.
- [6]. Aho A., Heinonen R., Lauren P. Benign and malignant mucocele of the appendix. *ActaChir Scand.* 1973;139(4):392–400. [PubMed]
- [7]. Higa E., Rosai J., Pizzimbono C. A., Wise L. Mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendicealmucocele. *Cancer.* 1973;32(6):1525–1541. [PubMed]
- [8]. Kim SH, Lim HK, Lee WJ, Lim JH, Byun JY. Mucocele of appendix: Ultrasonographic and CT findings. *Abdominal Imaging* 1998; 23: 292-296. [PubMed] [FULL TEXT]
- [9]. Sugarbaker P. H. Appendiceal Epithelial Neoplasms and PseudomyxomaPeritonei, a Distinct Clinical Entity with Distinct Treatments. In: Bland K. J., Büchler M. W., Csendes A., Garden O. Y., Saar M. G., Wong J., editors. *General Surgery. Principles and International Practice.* London-Limited: Springer; 2009. pp. 885–893. (eds)
- [10]. Rokitansky CF. A manual of pathological anatomy. Vol 2. English translation of the Vienna edition (1842). Philadelphia: Blancard and Lea, 1855:89. [Links]
- [11]. Isaaca KL, Warshauer DM; Mucocele of the appendix; computed tomographic, endoscopic and pathologic correlation. *Am. J. Gastroenterol.* 1992; 87: 787-789.
- [12]. Misdraji J, Yantiss RK, Graeme-Cook FM, Balis UJ, Young RH. Appendiceal mucinous neoplasms. A clinicopathologic analysis of 107 cases. *Am J SurgPathol* 2003;27(8):1089-103. [Links]
- [13]. Stocchi L, Wolff BG, Larson DR, Harrington JR. Surgical treatment of appendicealmucocele. *Arch Surg* 2003;138:585-90. [Links]
- [14]. Ruiz-Tovar J, Teruel DG, Castineiras VM, Dehesa AS, Quindós PL, Molina EM. Mucocele of the appendix. *World J Surg* 2007;31(3):542-8. [Links]
- [15]. Pickhardt P. J., Levy A. D., Rohrmann C. A., Jr, Kende A. L. Primary neoplasms of the appendix manifesting as acute appendicitis: CT findings with pathologic comparison. *Radiology.* 2002;224(3):775–781. [PubMed]
- [16]. García Lozano A., VazquesTarrago A., Castro Garcia C., Richart Aznar J., Gomez Abril S., Martinez Abad M. Mucocele of the appendix: presentation of 31 cases. *Cir Esp.* 2010;87(2):108–112. [PubMed]
- [17]. Higa E, Rosai J, Pizzimbono CA, Wise L: Mucosal hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendiceal "mucocele".
- [18]. *Cancer* 1973, 32:1525-1541. PubMed Abstract | Publisher Full Text
- [19]. Sugarbaker PH: Epithelial appendiceal neoplasms. *Cancer J* 2009, 15:3:225-35.
- [20]. Deans GT, Spence RAJ: Neoplastic lesions of the appendix. *British J Surgery* 1995, 82:299-306. Publisher Full Text
- [21]. Yamane YD, Yamane H, Castro Júnior PC, Marsilac A, Mesquita RB, Paulo FL. Mucocele da apêndice - relato de caso e revisão da literatura. *Rev bras Coloproct* 2005;25(3):256-60. [Links]
- [22]. Fonseca LM, Sassiné GCA, Luz MMP, Silva RG, Conceição SA, Lacerda-Filho A. Cistoadenoma de apêndice - relato de caso e revisão da literaturasobretumoresmucinosos do apêndicevermiforme. *Rev bras Coloproct* 2008;28(1):89-94. [Links]
- [23]. Caspi B, Cassif E, Auslender R, Herman A, Hagay Z, Appelman Z: The onion skin sign: a specific sonographic marker of appendicealmucocele. *J Ultrasound Med* 2004, 23(1):117-121. PubMed Abstract | Publisher Full Text
- [24]. Lien WC, Huang SP, Chi CL, Liu KL, Lin MT, Lai TI, Liu YP, Wang HP. Appendiceal outer diameter as an indicator for differentiating appendicealmucocele from appendicitis. *Am J Emerg Med.* 2006;24:801–805. [PubMed]
- [25]. Karakaya K., Barut F., Emre A. U., Ucan H. B., Cakmak G. K., Irkorucu O., et al. Appendicealmucocele: case reports and review of current literature. *World J Gastroenterol.* 2008;14(14):2280–2283. [PMC free article] [PubMed]
- [26]. Francica G., Lapicciarella G., Giardibello C., Scarano F., Angelone G., De Marino F., et al. Giant mucocele of the appendix: clinical and imaging finding in 3 cases. *J Ultrasound Med.* 2006;25(5):643–648. [PubMed]
- [27]. Benett G. L., Tanpitukponqse T. P., Macari M., Cho K. C., Babb J. S. CT diagnosis of mucocele of the appendix in patients with acute appendicitis. *AJR Am J Roentgenol.* 2009;192(3):103–110. [PubMed]
- [28]. Hamilton DL, Stormont JM: The volcano sign of appendicealmucocele. *GastrointestEndosc* 1989, 35:453-456. PubMed Abstract
- [29]. Aho AJ, Heinonen R, Laurén P: Benign and malignant mucocele of the appendix.Histological types and prognosis.*ActaChirScand* 1973, 139(4):392-400. PubMed Abstract
- [30]. Dachman AH, Lichtenstein JE, Friedman AC: Mucocele of the appendix and pseudomyxomaperitonei. *AJR Am J Roentgenol* 1985, 144:923-929. PubMed Abstract | Publisher Full Text
- [31]. Soweid AM, Clarkston WK, Andrus CH, Jannet CG: Diagnosis and management of appendicealmucoceles. *Dig Dis* 1998, 16:183-186. PubMed Abstract | Publisher Full Text
- [32]. Navarra G, Asopa V, Basaglia E, Jones M, Jiao LR, Habib NA: Mucous cystadenoma of the appendix: is it safe to remove it by a laparoscopic approach? *SurgEndosc* 2003, 17(5):833-4. PubMed Abstract | Publisher Full Text

- [33]. Zanati SA, Martin JA, Baker JP, Streutker CJ, Marcon NE. Colonoscopic diagnosis of mucocele of the appendix. *GastrointestEndosc*. 2005;62:452–456. [PubMed]
- [34]. Gonzalez Moreno S, Shmookler BM, Sugarbaker PH. Appendicealmucocele. Contraindication to laparoscopic appendectomy. *SurgEndosc*. 1998;12:1177–1179. [PubMed]
- [35]. Rampone B, Roviello F, Marrelli D, Pinto E. Giant appendicealmucocele: report of a case and brief review. *World J Gastroenterol*. 2005;11:4761–4763. [PubMed]
- [36]. Lau H, Yuen WK, Loong F, Lee F. Laparoscopic resection of an appendicealmucocele. *SurgLaparoscEndoscPercutan Tech*. 2002;12:367–370. [PubMed]
- [37]. Rudloff U, Malhotra S. Volvulus of an appendicealmucocele: report of a case. *Surg Today*. 2007;37:514–517. [PubMed]
- [38]. Miraliakbari R, Chapman WH 3rd. Laparoscopic treatment of an appendicealmucocele. *J LaparoendoscAdvSurg Tech A*. 1999;9:159–163. [PubMed]
- [39]. Kahn M, Friedman JH: Mucocele of the appendix: Diagnosis and surgical management.
- [40]. Dis Colon Rectum 1979, 22:267-269. PubMed Abstract | Publisher Full Text
- [41]. Dhage-Ivatury S, Sugarbaker PH. Update on the surgical approach to mucocele of the appendix. *J Am CollSurg* 2006;202(4):680–684.
- [42]. Federle MP, Anne VS. Mucocele of the appendix. In Federle MP, ed. *Diagnostic imaging: Abdomen*. Salt Lake City, UT: Amirsys, 2004:26–27.
- [43]. Lam CW, Kuo SJ, Chang HC, et al. Pseudomyxomaperitonei, origin from appendix: report of cases with images. *Int Surg*. 2003;88(3):133-136.
- [44]. Hinson FL, Ambrose NS. Pseudomyxomaperitonei. *Br J Surg*. 1998;85(10):1332-1339.
- [45]. Dhage-Ivatury S., Sugarbaker P. H. Update on the surgical approach to mucocele of the appendix. *J Am Coll Surg*. 2006;202(4):680–684. [PubMed]
- [46]. Fujiwara T, Hizuta A, Iwagaki A, Matsuno T, Hamada M, Tanaka N, et al. Appendicealmucocele with concomitant colonic cancer. Report of two cases. *Dis Colon Rectum* 1996;39(2):232-6. [Links]
- [47]. Gonzalez Moreno S, Shmookler BM, Sugarbaker PH. Appendicealmucocele. Contraindication to laparoscopic appendectomy. *SurgEndosc*. 1998;12(9): 1177-1179.
- [48]. Chiu CC, Wei PL, Huang MT, et al. Laparoscopic resection of appendiceal mucinous cystadenoma. *J LaparoendoscAdvSurg Tech A*. 2005;15(3): 325-328.
- [49]. Yoshida Y, Sato K, Tada T, et al. Two Cases of Mucinous Cystadenoma of the Appendix Successfully Treated by Laparoscopy. *Case Reports in Gastroenterology* 2013;7:44-48.
- [50]. Rabie ME, Al Shraim M, Al Skaini MS, et al Mucus containing cystic lesions “mucocele” of the appendix: the unresolved issues. *Int J SurgOncol* 2015doi:10.1155/2015/139461
- [51]. Akagi I, Yokoi K, Shimanuki K, et al. Giant appendicealmucocele: report of a case. *J Nippon Med Sch* 2014;81:110-113.
- [52]. Srihari V, Jayaram J, Baleswari G, et al. Mucinous cystadenoma of the appendix. *J NTR Univ Health Sci* 2015;4:182-184.
- [53]. Rikki SINGAL, Muzzafar ZAMAN, BhanuPartap SHARMA. Unusual Entities of Appendix Mimicking appendicitis clinically Emphasis on Diagnosis and Treatment. *MAEDICA- a journal of Clinical Medicine*. 2017;12(1); 23-20.

*Rajneesh Kumar. "Appendicealmucocele : Case Report And Review of Literature." IOSR Journal of Dental and Medical Sciences (IOSR-JDMS) 16.7 (2017): 45-49.