



International Journal of Surgery Science

E-ISSN: 2616-3470

P-ISSN: 2616-3462

© Surgery Science

www.surgeryscience.com

2020; 4(3): 333-336

Received: 04-05-2020

Accepted: 06-06-2020

Dr. B Sankararaman

Professor of General Surgery,
Vinayaka Missions Medical College
and Hospital, Karaikal, India

Dr. S Gurunathan

Director of Medical Services,
Tamil Nadu, India

Dr. M Arafath Natchiar

Assistant Professor, Vinayaka
Missions Medical College and
Hospital, Karaikal, India

Dr. Jassim AJ

Final year Post-Graduate,
Vinayaka Missions Medical College
and Hospital, Karaikal, India

Corresponding Author:

Dr. B Sankararaman

Professor of General Surgery,
Vinayaka Missions Medical College
and Hospital, Karaikal, India

Mucocele of appendix: A rare case study and review of literature

**Dr. B Sankararaman, Dr. S Gurunathan, Dr. M Arafath Natchiar and
Dr. Jassim AJ**

DOI: <https://doi.org/10.33545/surgery.2020.v4.i3f.515>

Abstract

Introduction: Mucocele of Appendix refers to the dilatation of the appendix due to mucus. It is a rare disorder with an estimated incidence of 0.2%-0.7% of all appendectomies performed [6] and 8% -10% of all appendiceal tumours [1]. It can be triggered by benign or malignant diseases which cause obstruction of the appendix resulting in accumulation of mucus secretion. Complications of Appendiceal Mucocele include among others obstruction, intussusception, and pseudomyxoma peritonei, which has a particularly poor prognosis [2].

Case Report: 69 year old post-menopausal female presented with complaints of right lower abdominal pain and nausea for 15 days. She was evaluated with Contrast Enhanced CT. CT showed a 9.3 x 3.8 x 3.5 cm tubular, thin-walled cystic mass extending from the base of the caecum to the right adnexa in pelvis suggestive of Mucocele of the appendix. Appendix was removed in the usual method. The stump was buried in caecum by purse string sutures using vicryl.

Conclusion: Mucoceles of the Appendix are rare. They have varied presentations. They should be properly evaluated and treated. Though right hemicolectomy is necessary in malignant lesions, it may be an over-treatment for benign mucoceles.

Keywords: Mucocele, appendicitis, appendix, mucinous cystadenoma, pseudomyxoma peritonei, mucinous cystadenocarcinoma

Introduction

Mucinous neoplasms of the appendix sometimes referred to as Mucocele include a spectrum of benign and malignant diseases, including simple cysts, mucinous cystadenoma, mucinous cystadenocarcinoma and pseudomyxoma peritonei. It is extremely rare, found in 0.2-0.7% of resected appendices [6]. A simple cyst results from non-neoplastic occlusion of the appendiceal lumen, is usually less than 2cms in diameter. In contrast, mucinous cystadenoma are benign tumours that represent the majority of 'Mucocele'. They can grow upto 8 cms or larger [5]. Patients typically remain asymptomatic due to slow-growing distention of the appendix and instead present incidentally with a mass on physical examination or abdominal imaging. On plain radiograph or CT, wall calcification is characteristic [8]. Early diagnosis is critical for favourable long-term outcomes because the operative management will differ from that of a dilated appendix secondary to acute appendicitis [3]. Dissemination of neoplastic cells and mucoid material in abdominal cavity, caused by appendiceal perforation, results in pseudomyxoma peritonei which is the dramatic evolution in 10-15% of cases [4]. Mucinous cystadenocarcinoma represents the malignant form of cystic neoplasms of the appendix. Unlike cystadenoma, patients are more likely to be symptomatic, with abdominal pain, weight loss, an abdominal mass or signs of acute appendicitis. In both cystadenomas and cystadenocarcinomas, the mucus material contains epithelial cells with low or high grade of dysplasia/frankly malignant cells. The rupture of this lesion may lead to the dissemination of the epithelium that causes multiple peritoneal deposits which produce mucin in the abdominal cavity, causing mucinous ascites or pseudomyxoma peritonei [21]. Right hemicolectomy should be performed in the setting of any suspicion of malignancy in an appendiceal mass for possible cure [5]. Physicians should consider it in the differential diagnosis of persistent enlarging ovarian cyst or adnexal mass [9]. The differential diagnosis of appendiceal Mucocele also includes mesenteric cyst and duplication cyst [16].

Case Report: 69-year-old post-menopausal female presented with complaints of right lower abdominal pain and nausea for 15 days. There was no history of vomiting or fever. Patient was the mother of 3 children with last child birth 35 years back. She had undergone tubectomy for sterilization. There was no history of tuberculosis, diabetes mellitus or hypertension. Examination revealed a right iliac fossa mass of about 8cm x 4 cm with indistinct edges. The mass was not mobile intra abdominally. Hernial sites were free. Transverse scar of tubectomy was present in the hypogastric region. Oral and IV contrast enhanced CT abdomen showed a 9.3 x 3.8 x 3.5 cm tubular, thin-walled cystic mass extending from the base of the caecum to the right adnexa in pelvis (Fig.1&2). There were no evidences of mural calcifications or thickening or solid components. This was

suggestive of Mucocele of the Appendix. The patient was taken up for elective surgery. Right lower Para Median incision was made. Mucocele of Appendix was confirmed intra-operatively (Fig 3 and 4). It was found extending into the pelvis. Base of appendix felt supple and caecum was free. No signs of inflammation were present. There were no adhesions of peritoneum. Uterus and adnexae were normal (Fig 5). Appendix was removed in the usual method. The stump was buried in caecum by purse string sutures using vicryl. The specimen (Fig.6) was sent for biopsy. Histopathological features were consistent with Mucocele of Appendix lined by flattened epithelial cells without any atypical features or inflammatory cells.



Fig 1: & Fig 2: (CT abdomen showing the mucocele of the appendix)

Operative Pictures



Fig 3: & Fig 4: showing mucocele of appendix



Fig 5: (Intra operative picture showing uterus, with attachments)



Fig 6: (showing resected specimen of mucocele of appendix)

Discussion

Mucocele of appendix refers to the dilatation of the appendix due to mucus. It is a rare disorder with an estimated incidence of 0.2%-0.7% of all appendectomies performed^[6] and 8% -10% of all appendiceal tumours^[1]. Reports are conflicting as to which sex is commonly affected^[11, 12, 19]. It is commonly encountered in patients between 26 and 83 years of age with a mean age of 52^[12]. Mucocele of the appendix is not a true histopathological entity. It is a macroscopic descriptive term for a distended and mucus-filled appendix of varied etiology. They are classified into four histological groups^[6, 15] (Chart 1). The first group consists of a simple retention cyst secondary to proximal occlusion of the appendix by e.g. a fecalith or scar tissue from previous inflammation, or in rare cases due to endometriosis^[6]. With rising pressure, degenerative changes in the appendiceal mucosa consequently develop. This type of mucocele is usually smaller than 2cm in diameter. The second group, called mucosal hyperplasia, has the same features as hyperplastic colonic polyps. Benign mucinous cystadenomas form the third group. Finally, the fourth group includes the malignant mucinous cystadenocarcinomas, characterized by stromal invasion by malignant cells without or with peritoneal implants i.e. pseudomyxoma peritonei. According to the histopathological examination, the incidence rate of mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma was found to be 23.1%, 61.4%, and 15.5%, respectively^[1]. In both cystadenomas and cystadenocarcinomas, the mucus material contains dysplastic epithelial cells or frankly malignant cells. The rupture of the appendix may lead to the dissemination of the epithelial cells that produce mucin in the abdominal cavity, causing mucinous ascites or pseudomyxoma peritonei^[21]. Clinical presentation and treatment options depend upon this classification. If the histological diagnosis is mucinous cystadenocarcinoma without metastasis, right hemicolectomy has to be carried out for oncological clearance.

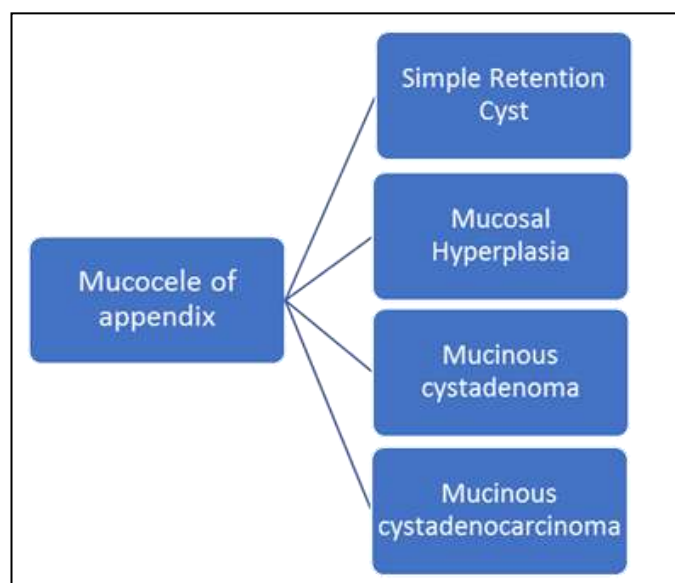


Chart 1: Classification of Mucoceles of Appendix

Review of Literature

Appendiceal Mucocele is a rare entity which was first described by Rokitansky in 1842^[7]. Reports are conflicting as to which sex is commonly affected^[11, 12, 19]. Many patients remain asymptomatic. It is diagnosed incidentally or presents as right lower quadrant abdominal pain mimicking acute appendicitis or urolithiasis or as right adnexal mass^[13]. Its association with

various conditions like ovarian tumor, endometriosis, and infertility or ovarian tumor has been found^[18]. Appendiceal mucocele has various complications but pseudomyxoma peritonei (due to rupture) is the most fearful complication. USG and CECT are important tools to diagnose. Surgery is the definitive management. Benign appendiceal mucocele has good prognosis while prognosis of malignant mucocele reduces significantly due to complications of pseudomyxoma peritonei^[2]. There is a continuous debate about the approach for surgery. Conventional surgery is generally preferred to laparoscopic approach as the latter increases the risk of rupture, but it is still performed for selected patients^[16]. Fine needle aspiration must be avoided as the risk of perforation is high that will also lead to pseudomyxoma peritonei^[9]. It is important to consider appendiceal mucocele as a differential diagnosis of any mass lesion in this area^[13]. AM may very rarely cause volvulus of caecum^[10]. Torsion of the appendiceal mucocele has been reported in certain rare cases^[2]. Appendiceal mucocele may mimic chronic tubo-ovarian abscess in certain cases^[16]. Postoperative chronic pain in an appendectomy patient may be due to appendicular stump Mucocele^[20]. Limited resources also show elevated serum carcinoembryonic antigen (CEA) in cases of mucocele of the appendix^[14]. Appendiceal mucocele may present as a porcelain appendix in certain patients^[16]. Rarely during a Caesarean section, a low grade mucinous appendiceal neoplasm may be an incidental finding^[17].

Conclusion

Though rare, Mucocele of Appendix presents in different ways. Surgeons should be aware of this entity and about its varied presentations. It should be differentiated from other clinical conditions and properly evaluated and treated. Rupture should be avoided to prevent the dreadful complication of pseudomyxoma peritonei. Though Right hemicolectomy is necessary in malignant lesions, it may be an over-treatment for benign mucoceles. Our case is unique in that the size of the AM is 9.3 x 3.8 x 3.5 cm and still the epithelial cells have not gone for atrophy as they are supposed to; the epithelial cells were flattened and did not show features of neoplasm. It was an unusually large simple retention cyst.

References

1. Abuoglu H, MK Yildiz *et al.* Clinicopathological analysis of patients operated for appendiceal mucocele. *Ulus Travma Acil Cerrahi Derg.* 23(3):230-234.
2. Aggarwal N, A Bhargava. Appendiceal mucocele secondary to torsion in an asymptomatic patient. *J Surg Case Rep.* 2019(8):rjz241.
3. Arrington D, B Jewett *et al.* Incidental mucocele of the appendix in a 15-year-old girl. *Pediatr Emerg Care.* 30(8):555-7.
4. Caracappa D, N Gulla *et al.* Appendiceal mucocele. A case report and literature review." *Ann Ital Chir.* 82(3):239-45.
5. Stocchi L, Wolff BG, Larson DR, Harrington JR. surgical treatment of appendiceal mucocele. *Arch Surg Chic Ill* 1960. 2003; 138(6):585-589; discussion 589-590.
6. Zaza Demetrashvili, Mamuka Chkhaidze *et al.* "mucocele of appendix: case report and review of literature. *Int Surg.* 2012; 97(3):266-269.
7. Rokitansky CFA. *Manual of Pathological Anatomy.* Vol. 2. Philadelphia: Blanchard & Lea, 1855.
8. Pickhardt PJ, Levy AD, Rohrmann CA, Kende AI. Primary neoplasms of the appendix: radiologic spectrum of disease with pathologic correlation. *Radiogr Rev Publ Radiol Soc N*

- Am Inc. 2003; 23(3):645-662.
9. Alghamdi AO, MY Aldossary *et al.* Low grade appendiceal mucinous neoplasm mimicking an ovarian cyst: A case report." *Int J Surg Case Rep.* 70:145-148.
 10. Costa V, JP Demuro. Low-grade appendiceal neoplasm presenting as a volvulus of the caecum." *Gastroenterol Rep (Oxf)* 1(3):207-10.
 11. Akbulut S, M Tas *et al.* Unusual histopathological findings in appendectomy specimens: a retrospective analysis and literature review." *World J Gastroenterol.* 17(15):1961-70.
 12. Basak F, M Hasbahceci *et al.* Does it matter if it is appendix mucocele instead of appendicitis? Case series and brief review of literature." *J Cancer Res Ther.* 14(6):1355-1360.
 13. Cristian DA, FA Grama *et al.* Low-grade appendiceal mucinous neoplasm mimicking an adnexal mass. *Rom J Morphol Embryol.* 56(2 Suppl):837-42.
 14. Khan AF, SH Waqar *et al.* "Mucocele of appendix with an elevated carcinoembryonic antigen: A case report." *J West Afr Coll Surg.* 7(4):120-127.
 15. Eltaib A Saad, Elamin Y Elsamani, Walid E AbdElrahim, Kamal E Elsiddig, Eltahir AG Khalil. Surgical treatment of mucocele of the appendix: a systematic review and case report". *J Surg Case Rep.* 2018; 2018(6):rjy102.
 16. Porcelain appendix:A Rare but Essential Diagnosis[J HK Coll Radiol 2002; 5:111-113]
 17. Eisuke Inubashiri, Yukio Watanabe, Noriyuki Akutagawa, Katamaru Kuroki, Masaki Sugawara, Keizou Deguchi, Nobuhiko Maeda *et al.* An incidental finding of low-grade appendiceal mucinous neoplasm during cesarean section: A case report". *JGH Open.* 2020; 4(2):306–308.
 18. Bharati V. Appendiceal Mucocele: A Rare Pathological Entity but an Important Differential Diagnosis. *Obstet Gynecol Int J.* 2016; 5(3):00159.
DOI: 10.15406/ogij.2016.05.00159
 19. Asenov Y, B Korukov *et al.* Appendiceal mucocele - Case Report and Review of the Literature. *Chirurgia (Bucur).* 2016; 110(6):565-9.
 20. Cama JK. Recurrent abdominal pain post appendectomy--a rare case. *Pac Health Dialog.* 16(2):78-81.
 21. Janduí Gomes De Abreu Filho, Erivaldo Fernandes De Lira. Mucocele of the appendix: appendectomy or colectomy?, *Journal of Coloproctology (Rio de Janeiro), J. Coloproctol. (Rio J.)* vol.31 no.3 Rio de Janeiro July/Sept, 2011.