Case Report

Giant appendiceal mucocele as a first manifestation in a patient with silent ulcerative colitis: A case report

Shareefa Abdulghaffar, MD*, Noor Badrawi, MD, Tarig Elnour Khairi, MD, Tasnim Riasath Keloth, MD, Paul Emmanuel Businge, MD

Department of Radiology, Department of Histopathology, Dubai Health Authority, Rashid Hospital, PO Box 4545, Dubai, United Arab Emirates

A R T I C L E   I N F O

Article history:
Received 21 December 2021
Revised 25 January 2022
Accepted 25 January 2022

Keywords:
Appendiceal mucocele
Ulcerative colitis
Computed tomography
Mucocele
Appendicular mucinous neoplasm

A B S T R A C T

Appendiceal mucocele (AM) is a rare lesion found in only 0.3% of all appendectomies and it is characterized by distention of the appendix by the accumulating intraluminal mucinous material. Four main histopathological subtypes of appendiceal mucocele has been described including: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and cystadenocarcinoma. We report a case of 48-year-old middle eastern female, previously healthy, presented to the primary health center with right iliac fossa discomfort, and referred to our hospital for further evaluation. Contrast enhanced computed tomography of the abdomen and pelvis and focused ultrasonography over the right iliac fossa showed the characteristic CT and sonographic appearance of appendiceal mucocele. An additional incidental note of lack of colonic haustral pattern involving the rectum and descending colon was noted on abdominal CT raising the suspicion of Ulcerative Colitis which was then confirmed by colonoscopy and histopathology. Although no causal relationship between appendicular mucocele and inflammatory bowel disease has been established yet and few case reports had reported appendicular mucocele in patients with ulcerative colitis, close surveillance in these patients, and early recognition of the characteristic imaging features is crucial in preventing dreadful complications like pseudomyxoma peritonei.

© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington.
This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

Appendiceal mucocele is a rare pathology of the appendix caused by accumulation of mucus within a dilated appendix [1]. It has been attributed to 2 main possible pathologic process: increased appendiceal intraluminal pressure secondary to obstruction caused by prior inflammation, mucosal hyperplasia, appendiceal fecoliths, or by an appendiceal tumor such as carcinoid, cystadenoma, and cystadenocarcinoma [2].

* Competing Interests: None declared.
+ Corresponding author
E-mail address: shareefa.yaqoob.90@gmail.com (S. Abdulghaffar).
https://doi.org/10.1016/j.radcr.2022.01.076
1930-0433/© 2022 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)
Appendiceal involvement in cases of ulcerative colitis is rare and most likely to be secondary to obstructive inflammatory process; however, clear causative relation between the 2 process has not been established.

**Case report**

A 48-year-old middle eastern female with no significant past medical or surgical history presented to the primary health center with mild discomfort on the right iliac fossa for the last few months. Patient denied any history of vomiting, weight loss, alteration in bowel habit, loss of appetite or rectal bleeding. No history of abnormal vaginal bleeding. Patient was not on any regular medications. Patient was vitally stable and during physical examination a palpable mass was felt in the right lower quadrant and abdominal ultrasound was done there and it showed a large complex right adnexal mass. Patient was referred to our hospital for further investigations. Basic hematological and laboratory work up as well as tumor markers including CA-125, CEA, and Ca19-9 were done and were within normal limits. Contrast enhanced computed tomography of the abdomen and pelvis (Fig. 1) showed an elongated cystic...
Focused ultrasound of the right lower abdomen shows a large elongated cystic mass at the base of the cecum with internal alternating echogenic multilayers resembling the onion skin appearance (red arrow) suggestive of appendiceal mucocele. Image (B) Color doppler image of the mass shows no internal vascularity (Color version of the figure is available online.)

Fig. 3 – Spot colonoscopy images showing superficial mucosal erosions and exudates with loss of underlying haustral pattern suggestive of ulcerative colitis.

Discussion

Appendicular mucocele (AM) is a rare entity that is seen in up to 0.3% of surgically removed appendix [1]. It is a cystic mass resulting from chronic mucinous collection within a dilated appendix and can be benign or malignant in nature [3]. There are 4 general histopathological subtypes including, a simple mucocele (retention cyst), mucosal hyperplasia,
mucinous cystadenoma, and mucinous cystadenocarcinoma [1,3,4]. Patients commonly manifest with features of acute appendicitis in up to 50% of the patients. Remaining of the cases present with non-specific symptoms or asymptomatic [5].

Typical CT findings of an appendiceal mucocele include a cystic well-encapsulated mass with wall calcifications containing homogenous low attenuation material and attached to the cecum at the expected location of the appendix [1,4,5].

Mural nodularity and irregular wall thickening are generally suggestive of malignant nature; however, differentiating benign from malignant AM may be difficult, and histologic examination is required [4].

Individuals with inflammatory bowel disease (IBD) and particularly ulcerative colitis, are infrequently diagnosed with appendicular neoplasms, and the relation remains unclear [2,6–9]. It is suggested that the inflammatory nature of the disease can involve the appendiceal orifice and prompt mucin retention within the appendix. Immunologic and genetic causes are also suspected as possible cause for developing AM in this particular group of individuals and is yet to be explored. We reported a case of appendicular mucocele with underlying undiagnosed inflammatory bowel disease (IBD) suggesting the relation between the 2 entities as a result of untreated inflammatory process leading to obstructive development of an AM.

**Conclusion**

Although rare, patients with IBD may develop appendiceal mucocele as a result of the inflammatory nature of the disease involving the appendiceal orifice leading to retention of mucin in the appendix. It is important to note that this group of individuals are at higher risk of developing malignancy of the colon and appendix may also be involved. Therefore, regular surveillance may be required for possible development of this entity.

**Patient consent**

A written consent was obtained from the patient for publication of this case and any accompanying images.
REFERENCES

[1] Wang H, Chen YQ, Wei R, Wang QB, Song B, Wang CY, et al. Appendiceal mucocele: a diagnostic dilemma in differentiating malignant from benign lesions with CT. AJR Am J Roentgenol 2013;201(4):W590–5. doi: 10.2214/AJR.12.9260.

[2] Takeda Y, Nakase H, Mikami S, Inoue T, Satou S, Sakai Y, et al. Possible link between ulcerative colitis and in situ adenocarcinoma of an appendiceal mucocele: importance of inflammation in the appendiceal orifice related to UC. Inflamm Bowel Dis 2008;14(6):873–4. doi: 10.1002/ibd.20386.

[3] Honnef I, Moschopulos M, Roeren T. Appendiceal mucinous cystadenoma. Radiographics 2008;28(5):1524–7. doi: 10.1148/rg.285075160.

[4] Bennett GL, Tanpitukpongse TP, Macari M, Cho KC, Babb JS. CT diagnosis of mucocele of the appendix in patients with acute appendicitis. AJR Am J Roentgenol 2009;192(3):W103–10. doi: 10.2214/AJR.08.1572.

[5] Leonards LM, Fahwa A, Patel MK, Petersen J, Nguyen MJ, Jude CM. Neoplasms of the appendix: pictorial review with clinical and pathologic correlation. Radiographics 2017;37(4):1059–83. doi: 10.1148/rg.2017160150.

[6] Matsumoto T, Nakamura S, Shimizu M, Iida M. Significance of appendiceal involvement in patients with ulcerative colitis. Gastrointest Endosc 2002;55(2):180–5. doi: 10.1067/mge.2002.121335.

[7] Lakatos PL, Gyori G, Halasz J, Fuszek P, Papp J, Jaray B, et al. Mucocele of the appendix: an unusual cause of lower abdominal pain in a patient with ulcerative colitis. A case report and review of literature. World J Gastroenterol 2005;11(3):457–9. doi: 10.3748/wjg.v11.i3.457.

[8] Zannoni U, Masci C, Bazzocchi R, Gandolfo F, Pezzi A, Alampi G, et al. Cancer of the appendix in long-standing ulcerative colitis: a case report. Tumori 1997;83(6):958–9.

[9] Noaki R, Kawahara H, Watanabe K, Kobayashi S, Uchiyama K, Yanaga K. Appendiceal mucocele detected under treatment of ulcerative colitis. Case Rep Gastroenterol 2009;3(3):360–5. doi: 10.1159/000242475.