EDITORIAL

Mucocele of the Appendix: Two Case Reports.

Mohammed E H Azoz[1] , Abdelmoniem M M Makkawi[1] , Ahmed Abdelrahman Fadolalnabi[1] ,Moataz Mohamed Alhasan Ali[2]

1. Department of Surgery, Faculty of Medicine, University of Alemam Almahdi .
2. Department of Pathology, Faculty of Medicine, University of Alemam Almahdi .

Correspondence: Mohammed Eltoum Hamed Azoz MD, F.MAS , Associate professor, Department of Surgery, Faculty of Medicine, University of Alemam Almahdi .General director of *AbdalaMohamed Salih Specialized Centre*. Head department of Gastrointestinal Endoscopic Unit, *Kosti Teaching Hospital*. Kosti, White Nile State,Sudan, P.O.Box 209, Tel:+29912247390. E mail: mohammedazoz61@hotmail.com.

Abstract:

Introduction: Appendiceal mucocele is a rare condition which was described as an obstructive dilatation of the appendix caused by intraluminal accumulation of mucoid material.

Case Reports: We present a case of 26 years old male who came for elective appendectomy which intraoperatively found to have mucocele of appendix which was treated successfully by appendicectomy. The second case was a 55 years female who presented with a right iliac fossa mass with mesenteric lymph nodes enlargement ,underwent right hemicolectomy and the histopathological report showed mucinous cystadenoma of the appendix.

Conclusion: In conclusion, appendiceal mucocele is a rare disease and has a clinical presentation that resembles acute appendicitis or RIF Tumour.

Keywords: Mucocele, Appendix .

Introduction:

Appendiceal mucocele is a rare condition which was described as an obstructive dilatation of the appendix caused by intraluminal accumulation of mucoid material. It has an incidence that ranged between 0.2% to 0.7% of all appendectomied specimens[1-4]. Mucocele of the appendix was first described by Rokitansky in 1842 [5]. Mucocele of appendix is a macroscopic clinical diagnosis and considered a benign disorder when the obstructive cause found to be a foreign body, bile stone or faeces. On the other hand, mucinous tumours (adenoma or adenocarcinoma) could also obstruct the lumen of the appendix, which is called pseudomyxoma peritonei. This disorder is lethal without treatment, because the enormous amount of gelatinous ascitis results in the dysfunction of the abdominal organs [6]. The prognosis of appendiceal mucocele depends on the histopathological findings[6]. The surgical management depends on the histopathological type of the appendiceal mucocele, involvement of the base of the appendix, involvement of the mesenteric lymph nodes and spread of the disease to peritoneal cavity.

EDITORIAL

We describe two rare cases of a mucocele of the appendix and to advocate successful management with satisfactory outcome.

Case Report 1:

A 26-year-old male, who presented with a medical history of appendicular mass 6 weeks prior to time of the surgery, came for elective appendicetomy. The review of systems was within normal limits and he had no family history of malignancy.

On physical exam, he was well-appearing and afebrile with normal vital signs. His abdomen was soft. His laboratory values were normal, including a white blood cell count. Intraoperatively,the appendix was found significantly dilated and tense and filled with clear fluid in its tip and distal third and its base was not involved (Fig 1). There was no free fluid or any mucinous deposits either in periappendeceal nor in the peritoneal cavity. Also there was no evidence of enlarged lymph nodes in the mesoappendix . The diagnosis of mucocele of appendix was made intraopartively and appendectomy was done.

On the third postoperative day, the patient was discharged without complications.The histopathological report of the appendix showed appendicular mucosal hyperplasia with mucin pooling (mucocele) with evidence of chronic appendicitis.

Case Report 2:

A 55 years old house – wife presented with painless right lower abdominal quadrant lump for 2 years. The lump gradually increased in size till it reached an orange size. This lump was not associated with pain, nausea, vomiting or any other gastrointestinal symptom. The patient was not diabetic, hypertensive or had any other systemic illnesses.

On examination she looked unwasted, not dehydrated or toxic, abdomen was soft and there was10cm×6cm palpable mass, slightly mobile side to side and slightly tender to touch, with well defined margins, in the right side of lower abdomen. There was no organomegaly, palpable paraortic lymph nodes or ascitis.

The routine laboratory reports of complete blood count ESR, blood sugar and urea, and urine examination were all within normal limits. The ultrasound report showed a cystic mass taking the shape of a kidney in the right iliac fossa without knowing its origin. Differential diagnosis of mesenteric cyst, ovarian cyst or caecal tumor was put and hence a decision of an exploratory laparotomy was made. At laparotomy, a firm mass was found in the caecal region with enlarged mesenteric lymph node, so decision of right hemicoloectomy was made. The patient had uneventful postoperative course. The speciment sent for histopathology which confirmed mucinous cystadenoma of the appendix.

Discussion:

The mucocele of the appendix is described as a cystically dilated appendix filled with mucin secondary to non-acuteappendiceal obstruction. The obstruction can be secondary to anappendicolith, endometriosis, extrinsic compression, inflammation,or

EDITORIAL

neoplasm, and therefore surgical resection and pathologic evaluation are recommended[7].

The mucocele of the appendix clinically can present mimicking acute appendicitis with right lower quadrant pain, as an abdominal mass, with an intussusception, or incidentally. They are often diagnosed during a work-up for abdominal pain by ultrasound or cross sectional imaging of the abdomen and pelvis for other indications[7].

Appendiceal mucinous neoplasms are seldom seen in the pediatric population, as they are typically present in the sixth decade of life [8, 9]. Alemayehu et al. reported the incidence of unexpected pathology findings after appendectomy in 3602 children aged over 16- years, and none were found to have a mucinous neoplasm [10].

Pathologically, the condition may be classified into neoplastic and non neoplastic variants. The neoplastic variant results from overproduction of mucus by a mucinous tumour of the appendix. It includes three main categories: mucosal hyperplasia, cystadenoma, and cystadenocarcinoma [11, 12, 13].In mucosal hyperplasia, there is no epithelial atypia, whereas in mucinous cystadenoma, there is some degree of epithelial atypia and, additionally, acellular mucous may be present in the periappendiceal region or free in the peritoneal cavity. These two variants are benign and simple appendectomy is curative. Pseudomyxoma peritonei did not follow these two categories inHiga series [12].The mucinous cystadenocarcinoma is characterized by the presence of stromal invasion by malignant glands and/or the presence of mucous and mucous secreting cells in the peritoneal cavity [11,12].

Contrary to the a fore mentioned categories, the nonneoplasticvariant results from chronic insidious obstruction ofthe appendiceal lumen by any process other than mucinousneoplasia [11,14,15]. This leads to retention of mucous behind the obstruction and finally its seepage to the out side as the intraluminal pressure increases. It encompasses different types according to the obstructive lesion and was thus termed inflammatory, obstructive, simple mucocele or retention cyst of the appendix [16].

Preoperative diagnosis of appendicular mucocele is very important for the selection of an adequate surgical method to prevent peritoneal dissemination, to prevent intraoperative and postoperative complications, and repeated surgery. Ultrasonography(USG), computed tomography (CT), and colonoscopy are used for diagnosis. 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 the case of acute appendicitis, the outer diameter threshold of the appendix is 6 mm to 15 mm and if more may indicate the presence of a mucocele, with 83% sensitivity and 92% specificity. CT is regarded as the most accurate method of diagnosis. CT can be used to discover the signs specific to mucocele with high accuracy: these signs include 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 may be visible from this orifice. Furthermore, synchronous and metachronous tumors of colon can be identified[17].

EDITORIAL

Concerning the surgical treatment, a useful algorithm has been suggested by Filho and associates [16]. When the appendiceal base is not involved, appendectomy is done with excision of all the mesoappendiceal fat and contained lymph nodes. If the base is involved, typhlectomy, using the linear cutting stapler (GIA), or partial right hemicolectomy, is done well away from the appendix base. Frozen section is performed, and if no malignancy is found, as in cases of hyperplasia and cystadenoma, the operation is terminated. On the other hand, in case of proved malignancy, cystadenocarcinoma, necessitates an oncologic right hemicolectomy.

Our first case was a 26 years old male who came for elective appendectomy which, intraoperatively, found to be a mucocele of the appendix, not involving the base of the appendix nor causing accumulation of mucinous fluid either peri-appendeceal or in the peritoneal cavity and had no enlarged lymph nodes in mesoappendix. The second case was 55 years female presented with right iliac fossa mass that proved to be mucinous cystadenoma of the appendix with mesenteric lymph nodes enlargement. The first case was treated by appendicectomy and the second one by oncologic right hemicolectomy which were adequate surgical tactics for these types of the mucoceles of the appendix.

In conclusion, appendiceal mucocele is a rare disease and has a clinical presentation that resembles acute appendicitis. The suspicion of the mucocele of the appendix should be kept in mind when an ultrasound report shows elongated cystic mass in right iliac fossa in a patient with right lower quadrant abdominal pain. A correct diagnosis before surgery is very important for the selection of the optimum surgical technique to avoid severe intraoperativeand postoperative complications.

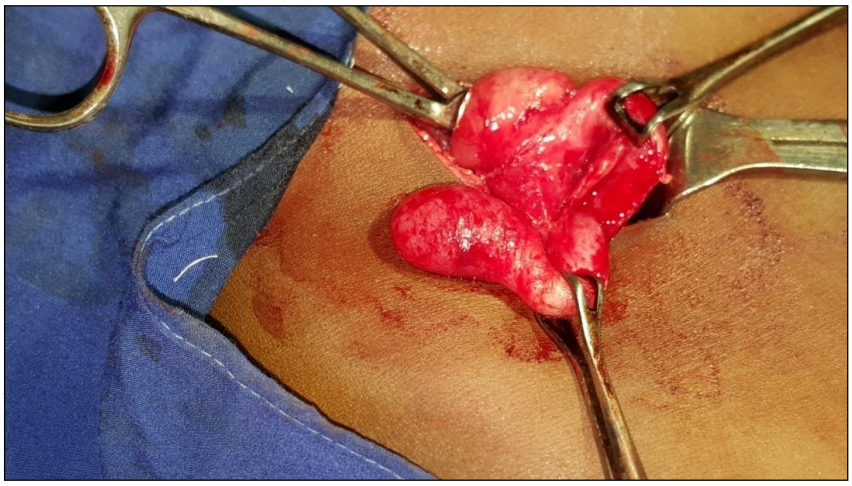

Figure 1: Mucocele of appendix involving the tip and the distal third

EDITORIAL

References:

1. Rangarajan M, Palanivelu C, Kavalakat AJ, Parthasarathi R. Laparoscopic appendectomy for mucocele of the appendix. Report 8 cases. Indian J Gastroenterol 2006; 25(5):256–257.
2. Marudanayagam R, Williams GT, Rees BI. Review of the pathological results of 2660 appendectomy specimens. J Gastroenterol 2006; 41(8):745–749.
3. Ruiz-Tovar J, Teruel DG, Gastineires VM, Dehesa AS, Quindos PL, Molina EM. Mucocele of the appendix. World J Surg 2007; 31(3):542–548.
4. Smeenk RM, van Velthuysen ML, Verwaal VJ, Zoetmulder FA. Appendiceal neoplasms and pseudomyxoma peritonei: a population-based study. Eur J Surg Oncol 2008; 34(2):196–201.
5. Jay V: The legacy of Karl Rokitansky. Arch Pathol Lab Med 124, 345–346 (2000).
6. Balint I, M Nad ,A KIRALY ,O BALI ,A RASHED ,L VIZSY. Giant appendix or an appendiceal mucocele? Case report of an 11-year-old child. Interventional Medicine & Applied Science, 2014. 6 (4): 187–190.
7. B S Rich, S. Roychoudhury, A K. Williamson, R D. Glick. Pediatric mucinous neoplasm of the appendix presenting as a mucocele: A case report and review of the literature, Journal of Pediatric Surgery Case Reports, 2017(17):11-14.
8. Aho AJ, Heinonen R, Lauren P. Benign and malignant mucocele of the appendix. Histological types and prognosis. Acta Chir Scand 1973; 139(4): 392-400.
9. Pai RK, Beck AH, Norton JA, Longacre TA. Appendiceal mucinous neoplasms clinicopathologic study of 116 cases with analysis of factors predicting recurrence. Am J Surg Pathol 2009; 33(10):1425-39.
10. Alemayehu H, Snyder CL, St Peter SD, Ostlie DJ. Incidence and outcomes of unexpected pathology findings after appendectomy. J Pediatr Surg 2014; 49: 1390-3.
11. J. Rosai, “Tumours of the gastrointestinal tract appendix,” in *Rosai and Ackerman’s Surgical Pathology*, pp. 757–775, Mosby, Philadelphia, Pa, USA, 9th edition, 2004.
12. E. Higa, J. Rosai, C. A. Pizzimbono, and L. Wise, “Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix. A reevaluation of appendiceal ‘mucocele’,” *Cancer*1973; 32(6): 1525–1541.
13. A. H. Qizilbash, “Mucoceles of the appendix.Their relationship to hyperplastic polyps, mucinous cystadenomas, and cystadenocarcinomas,” *Archives of pathology* 1975; 99(10): 548– 555.
14. H. S. Cooper, “Intestinal neoplasms,” in *Sternberg’s Diagnostic Surgical Pathology*, S. E. Mills, D. Carter, J. K. Greenson, V. E.Reuter, andM.H.Stoler, Eds., chapter 34,pp. 1368–1431,WoltersKluwer/LippincottWilliams &Wilkins, Philadelphia, Pa, USA, 5th edition, 2004.
15. D. K. Driman, D. E. Melega, G. A. Vilos, and E. A. Plewes, “Mucocele of the appendix secondary to endometriosis: report of two cases, one with localized pseudomyxoma peritonei,” *American Journal of Clinical Pathology* 2000; 113(6): 860– 864.
16. J. G. D. A. Filho and E. F. D. Lira, “Mucocele of the appendix: appendectomy or colectomy?” *Journal of Coloproctology* 2011; 31(3): 276–284.

17. Z . Demetrashvil et al . Mucocele of the Appendix: Case Report and Review of Literature. Int Surg 2012; 97:266–269.