

THE APPENDICEAL MUCOCELE AN ENTITY NOT TO BE UNDERESTIMATE.

M. Moujahid*, V. A. Blata, I. Talbi, Y. Mejdahoui, A. Laalou, R. Chouiba, F. Mohafid, M. Laaroussi, N. Njoumi, M. Najih, M. Yaka, A. Ehirchiou and A. Zentar

Visceral Surgery Service Ii, Military Training Hospital Mohamed V Rabat Mohamed V University, Faculty of Medicine and Pharmacy, Rabat-Morocco.

*Corresponding Author: M. Moujahid

Visceral Surgery Service Ii, Military Training Hospital Mohamed V Rabat Mohamed V University, Faculty of Medicine and Pharmacy, Rabat-Morocco.

Article Received on 05/05/2017

Article Revised on 26/05/2017

Article Accepted on 16/06/2017

ABSTRACT

Appendiceal mucocele is a rare disease. Sometimes it is discovered accidentally and sometimes it resembles acute appendicitis. Correct diagnosis before surgery is important for the selection of adequate surgical treatment to avoid intraoperative and postoperative complications. Ultrasonography, and particularly computed tomography (CT), should be used extensively for this purpose. If mucocele is treated incorrectly, pseudomyxoma peritonei may develop. We report a case of appendicular mucocele in a male patient aged 60 years old who was admitted to the emergency department of Military Hospital Mohamed V in Rabat, with the signs of acute appendicitis. As if it were an elderly patient, in whom the literature insists on performing CT, therefore appendicular mucocele was discovered by this imaging technique. Emergency open right hemicolectomy and ileo transverse-colon anastomosis termino lateral was performed because the base of the appendix was involved in the process. No free fluid was found in the peritoneal cavity. Histopathologic diagnosis was mucocele of appendix with simple mucous cyst. The follow-up was simple without any recurrence since five years.

KEYWORDS: Appendix, Appendicitis, Mucocele.

INTRODUCTION

The mucocele of the appendix (AM) was first described in 1842 by Rokitansky.^[1] AM is a rare entity that can present with a variety of clinical symptoms or occur as an incidental surgical finding. The incidence is 0.2%-0.4% of all appendectomied specimens.^[1-3] It affects preferentially the woman between 50 and 60 years.^[2] AM is a progressive dilatation of the appendix from the intraluminal accumulation of the mucoid substance.^[3,4] It may be a benign or malignant process that may come as a consequence of obstructive or inflammatory processes, cystadenomas or cystadenocarcinomas.^[5-7] Besides these causes, other tumor lesions in the appendix or cecum may present as mucocele.^[8] Its main complication is pseudomyxoma peritonei.

There are four histological types, which lead to individualized surgical treatment and course in each case.^[7,9]

- ✚ Simple mucocele (inflammatory, obstructive or retention cyst) is characterized by degenerative epithelial changes and results in the obstruction and the distension of the appendix. There is no evidence of hyperplasia or mucosal atypia.
- ✚ Hyperplastic mucocele, the appendix dilation occurs due to the hyperplastic growth of the appendix or

cecal mucosa, just like hyperplastic polyps in the colon.

Simple and hyperplastic mucoceles correspond to 5 to 25% of the cases, and mucus is usually acellular.

- ✚ The mucinous cystadenoma is an appendix neoplasm with dysplastic epithelium similar to colon adenomatous polyps, and corresponds to 63 to 84% of the cases.
- ✚ The mucinous cystadenocarcinoma presents high grade cellular dysplasia and stromal invasion, besides muscularis mucosae, and represents 11 to 20% of the cases.

Treatment is always surgery and determined by the organ's integrity, the dimensions of the base and histological type of the lesion.

Preoperative diagnosis that distinguishes AM from acute appendicitis (AA) as the others like mesenteric cysts, digestive duplications, complicated appendicitis, ovarian cysts and hydrosalpinx^[8] is essential for the best choice of surgical approach (open vs laparoscopic) to prevent peritoneal dissemination and perform the appropriate surgery.^[8-10]

CASE REPORT (Material and Method)

A 60-year-old man, with no significant pathological history, consulted in the emergency room for pains in the

right iliac fossa which has had evolution for 15 days. The patient was afebrile with a sensitivity of the right iliac fossa, the biological assessment showed a hyperleucocytosis without inflammatory syndrome. An abdomino-pelvic spiral computed tomography (CT) was performed before and after injection of contrast agent, axial cut (Fig. 1), with frontal reconstruction (fig. 2), which showed the presence of a cystic mass of the right iliac fossa juxta caecale, oblong, whose wall is enhanced after injection of contrast product, the diagnosis of appendicular mucocele has been posed and the patient was operated by a right hemicolectomy and ileo transverse-colon anastomosis termino lateral (Figure 3). The sequences were simple without any sign of recurrence.

FIGURES



Figure n°1: Abdominal scan showing cystic formation of the right iliac fossa.

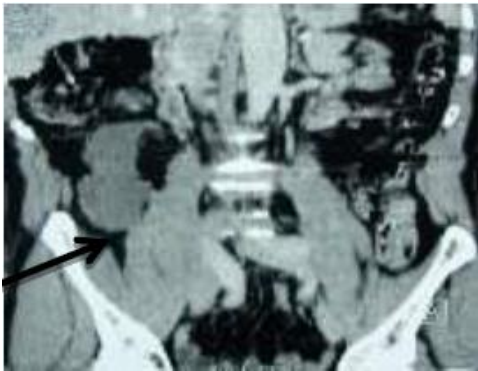


Figure n°2: frontal section showing the oblong nature of the cystic formation juxta caecale.

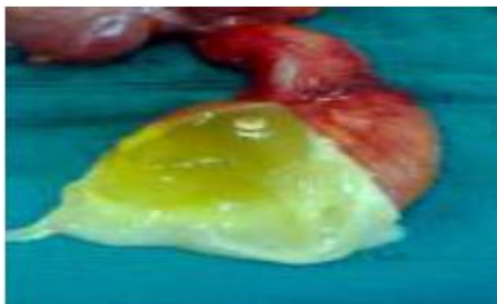


Figure n°3: right hemicolectomy piece showing the appendicular mucocele with gelatinous contents after opening the piece.

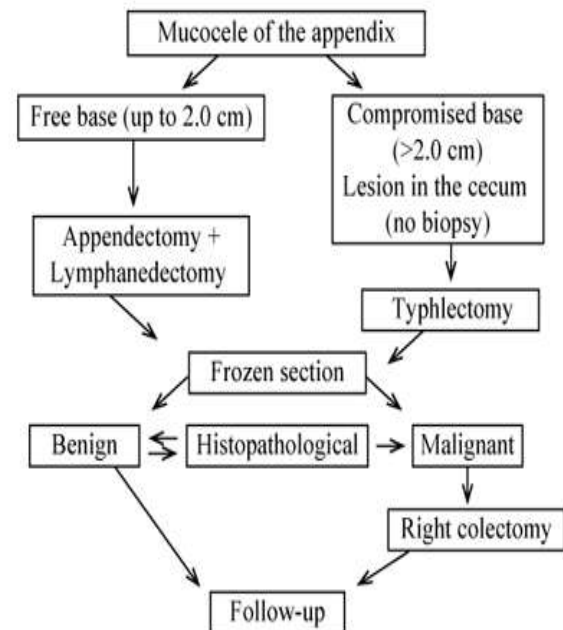


Figure 3. Treatment protocol of mucocele.

DISCUSSION

Mucocele of the appendix was first described by Rokitsky. This disease is characterized by dilatation of a lumen as a result of an accumulation of a large amount of mucus. The appendix is lined by epithelium containing more goblet cells than the colon. As a result, most appendiceal epithelial tumors are mucinous and start as mucoceles. It falls under the category of rare diseases. Its incidence ranges between 0.2% and 0.4% of all excised appendixes.^[10] This condition can have benign as well as malignant processes. According to modern classification, there are 4 histologic types: retention cyst, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.^[10,11]

The clinical flow of the disease does not have a specific picture. It often flows asymptotically. In about 50% of cases it is discovered accidentally during radiologic and endoscopic examinations or at surgery. A patient's clinical symptoms may include pain in the right lower quadrant of the abdomen, palpable abdominal mass, nausea, vomiting, weight loss, gastrointestinal bleeding, and signs of intussusception of the intestines.^[11,12]

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 complication, and repeated surgery.^[13] USG, computed tomography (CT), and colonoscopy is used for diagnostics. 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.^[14] 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.^[15] By colonoscopy an elevation of the appendiceal orifice is seen and a yellowish mucous discharge would be visible from this orifice. Furthermore, synchronous and metachronous tumors of colon can be identified. In our patient USG did not provide the correct information, so in this case especially in older patients in whom AM should be considered.^[16] We decided to perform CT that brought the diagnosis.

One of the cardinal principles of surgical treatment of this disease is that intact mucoceles do not pose a threat for the patient. Therefore, the selection of an adequate surgical method is very important. Some surgeons think that open surgery should be favored against laparoscopy. If the surgery was launched using a laparoscopic method and it appears that there is an appendiceal mucocele, it must be converted into open surgery. This has two objectives:

- To perform surgery carefully so the cyst is not ruptured and the filling is not scattered into the peritoneal cavity.
- With an open surgery compared to the laparoscopic method, it is possible to have a fuller inspection, palpation, and direct inspection of the spots in the abdomen where mucinous tumors are most common.^[17,18] Some surgeons consider that the operation can be performed using a laparoscopic method by adhering to safety rules, especially when removing the mucocele from the abdomen and an endobag must be used.^[19,20]

An algorithm for the selection of the type of surgery has been furnished by Dhage, Ivatury and Sugarbaker.^[20] It envisages several factors: 1) Whether mucocele is perforated or not. 2) Whether the base of the appendix (margins of resection) is involved in the process; and 3) whether there are positive lymph nodes of mesoappendix and ileocolic. As a result patients may require different operations: appendectomy to right colectomy, including cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy, early postoperative intraperitoneal chemotherapy.^[20] In our patient the mucocele was not perforated, base of the appendix was involved by the process, and no regional lymph node enlargement. Therefore, right hemicolectomy was performed, which is an adequate surgery in such a case.

CONCLUSION

Appendiceal mucocele is a rare disease and has a clinical picture that resembles acute appendicitis. A correct diagnosis before surgery is very important for the selection of surgical technique to avoid severe intraoperative and postoperative complications. USG, particularly CT, should be used extensively for this purpose. In our opinion, every patient more than 50 years old who arrives at the emergency department with

clinical symptoms of acute appendicitis must undergo CT and it would be prudent to always think about the diagnosis of AM.

No conflict of interest

REFERENCES

1. Pitiakoudis M, Tsaroucha AK, Mimidis K, Polychronidis A, Minopoulos G, Simopoulos C. Mucocele of the appendix: a report of five cases. *Tech Coloproctol*, 2004; 8: 109–112.
2. Zanati SA, Martin JA, Baker JP, Streutker CJ, Marcon NE. Colonoscopic diagnosis of mucocele of the appendix. *Gastrointest Endosc*, 2005; 62: 452–456.
3. Lien WC, Huang SP, Chi CL, Liu KL, Lin MT, Lai TI, Liu YP, Wang HP. Appendiceal outer diameter as an indicator for differentiating appendiceal mucocele from appendicitis. *Am J Emerg Med.*, 2006; 24: 801–805.
4. Jaffe BM, Berger DH. The appendix. In: Brunnicardi FC, Andersen DK, Billiar TR, Dunn DL, Hunter JG, Pollock RE, editors. *Schwartz's Principles of Surgery*. International edition: McGraw Hill Companies Inc, 2005; 1119–1137.
5. Gonzalez Moreno S, Shmookler BM, Sugarbaker PH. Appendiceal mucocele. Contraindication to laparoscopic appendectomy. *Surg Endosc*, 1998; 12: 1177–1179.
6. Creuzé N, Savoye-Collet C, Lemoine F, Tapon E, Ribeiro C et Thiebot J. Mucocèle sur moignon appendiculaire. *J Radiol*, 2013; 89: 57-9.
7. Higa E, Rosai J, Pizzimbono CA, Wise L. Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendiceal mucocele. *Cancer*, 1973; 32(6): 1525-41.
8. Driman DK, Melega DE, Vilos GA, Plewes EA. Mucocele of the appendix secondary to endometriosis. Report of two cases, one with localized pseudomyxoma peritonei. *Am J Clin Pathol*, 2000; 113(6): 860-4.
9. 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.
10. Rokitsky C. F. *A Manual of Pathological Anatomy*. Philadelphia: Blanchard & Lea, 1855; 2.
11. Sugarbaker P. H. Epithelial appendiceal neoplasms. *Cancer J.*, 2009; 15(3): 225–235.
12. Lien W. C., Huang S. P., Chi C. L., Liu K. L., Lin M. T., Lai T. I., et al. Appendiceal outer diameter as an indicator for differentiating appendiceal mucocele from appendicitis. *Am J Emerg Med.*, 2006; 24(7): 801–805.
13. 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.
14. Papaziogas B., Koutelidakis I., Tsiaousis P., Goula O. C., Lakis S., Atmatzidis S., et al. Appendiceal

- mucocele. A retrospective analysis of 19 cases. *J Gastrointest Cancer.*, 2007; 38(2–4): 141–147.
15. Haritopoulos K. N., Brown D. C., Lewis P., Mansour F., Eltayar A. R., Labruzzo C., et al. Appendiceal mucocele: a case report and review of literature. *Int Surg.*, 2001; 86(4): 259–262.
 16. Vriens B. H., Klaase J. M. Giant mucinous cystadenoma of the appendix. *Am J Surg.*, 2007; 194(3): 392–393.
 17. Ivatury S, Sugarbaker PH. Update on the surgical approach to mucocele of the appendix. *J Am Coll Surg.*, 2006; 202(4): 680-4.
 18. Karakaya K, Barut F, Emre AU, Ucan HB, Cakmak GK, Irkorucu O, et al. Appendiceal mucocele: case reports and review of current literature. *World J Gastroenterol*, 2008; 14(14): 2280-3.
 19. Khan MR, Ahmed R, Saleem T. Intricacies in the surgical management of appendiceal mucinous cystadenoma: a case report and review of literature. *J Med Case Reports*, 2010; 5(4): 129.
 20. Sugarbaker PH. Appendiceal epithelial neoplasms and pseudomyxoma peritonei, a distinct clinical entity with distinct treatments. In: Bland KJ, Buchler MW, Csendes A, Garden OY, Saar MG, Wong J (eds). *General Surgery. Principles and International Practice*. London-Limited: Springer, 2009; 885-893.