

Mucocele of Appendix: Case Report and Review of Literature

Mangilal V¹, Suryanarayana Reddy², Sumalatha², Prashanth S³, Manoj Kumar D³

¹ Associate Professor

² Professor and HOD

² Assistant Professor of Pathology

³ Post Graduate students

Department of General Surgery
Chalmeda Anand Rao
Institute of Medical Sciences
Karimnagar - 505 001.
Telangana, India.

CORRESPONDENCE :

¹ Dr. Mangilal V,
MS (Gen.Surgery)
Associate Professor
Department
of General Surgery
Chalmeda Anand Rao
Institute of Medical Sciences
Karimnagar-505001 Telangana,
India.
Email:vankdothmangilal@gmail.com

ABSTRACT

Appendiceal mucocele is a rare disease. Sometimes it is discovered accidentally and sometimes it resembles acute appendicitis. Correct diagnosis before surgery is very important for the selection of adequate surgical treatment to avoid intraoperative and postoperative complications. Ultrasonography, and particularly computed tomography, should be used extensively for this purpose. If mucocele is treated incorrectly pseudomyxoma peritonei, which is characterized by malignant process, may develop.

We present a case of a 64-year-old woman who was admitted to the emergency department with the signs of acute appendicitis. Open surgery was performed. At the time of surgery cystic mass of appendix with dimensions 6.9X2.5cm, with inflamed walls, but without perforation was discovered in right iliac fossa. Only appendectomy was performed because no pathologic process was found in the base of the appendix and lymph nodes were not increased in size. Histopathologic diagnosis was mucosal hyperplasia of Appendix.

Key words: Mucocele , pseudomyxoma peritonei, appendicitis

INTRODUCTION

The mucocele of the appendix was first described in 1842 by Rokitansky.^[1] This disease is considered as a rare lesion of the appendix, which is found in 0.2 to 0.3% of the appendectomies.^[2] It is characterized by the dilation of the organ lumen with mucus accumulation, being more frequent among individuals aged 50 years or more.^[3,4] Gender prevalence is controversial.

Appendix mucocele may come as a consequence of obstructive or inflammatory processes, cystadenomas or cystadenocarcinomas.^[5] Besides these causes, other tumor lesions in the appendix or cecum may present as mucocele.^[6] Its main complication is pseudomyxoma peritonei. Treatment is always surgery and determined by the organ's integrity, the dimensions of the base and histological type of the lesion.

CASE REPORT

A 64-year-old woman was referred to the emergency department. Her complaints were pain in the right lower

quadrant of the abdomen and nausea. The symptoms started 3 days prior. When palpating the lower right quadrant of the abdomen the patient felt pain, muscles were moderately rigid. She was diagnosed as carcinoma breast (Stage 4) 4months ago and she underwent neoadjuvant chemotherapy for the same.

Body temperature was 37.5uC.Hb was 11gr% Leukocytosis (13.8X 10⁹ /L) was notable from laboratory test results. Ultrasonography (USG) showed inflamed appendix with mucocele and fat stranding open surgery was performed. At the time of surgery, a cystic mass of the appendix (6.8X2.5cm), with inflamed walls, but without perforation, was discovered in the right iliac fossa. No discharge was found in the peritoneal cavity.

Only appendectomy was performed because no pathologic process was found in the base of the appendix and the lymph nodes were not increased in size. Histopathologic diagnosis was mucosal hyperplasia of appendix. There were no complications in the postoperative period. The patient was discharged from the seventh post operative day.

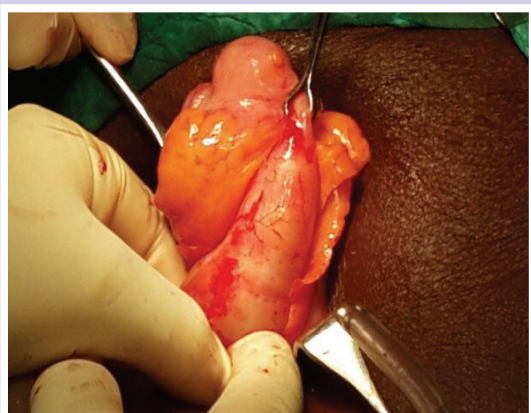


Figure-1: Intraoperative image showing dilated cystic appendix

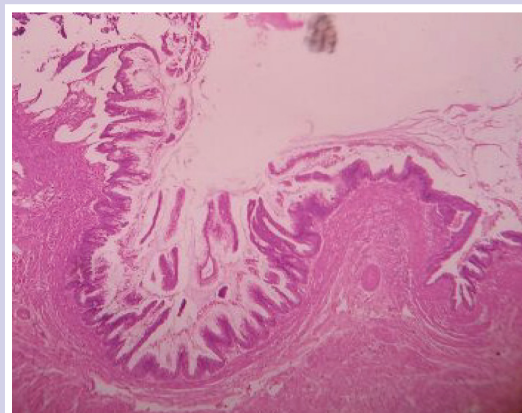


Figure-2: Histopathology showing mucosal hyperplasia

The mucocele of the appendix is a descriptive and unspecific term to define the cystic dilation of the appendix caused by the accumulation of mucus secretion. This process is slow and gradual, with no signs of infection inside the organ.

It results from the lumen obstruction in the appendix, which is secondary to the inflammatory or neoplastic proliferation of the appendix mucosa, or of lesions in the cecum, adjacent to the appendiceal ostium. Mucocele in the appendix may be classified according to the histological characteristics of lumen obstruction.^[7]

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.

In 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.

The clinical presentation of the disease does not have a specific picture. Mostly it is asymptomatic. In about 50% of cases it is discovered accidentally during radiologic and laproscopic 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.^[3,8,9,11]

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.^[8,11] 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.^[10] 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.^[3,7,9] 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.^[3,9]

In our patient as USG abdomen was clearly giving evidence of presence of mucocele of appendix of size 6.9X2.5cm, we did not perform CT.

One of the cardinal principles of surgical treatment of this disease is that intact mucoceles do not pose a threat for the patient. If it is perforated and the filling turns up in the peritoneal cavity, there is a high probability that pseudomyxoma peritonei will develop, for which treatment is very problematic and long-term results are quite unsatisfactory.^[8,11] 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 2 objectives: (1) to perform surgery carefully so the cyst is not ruptured and the filling is not scattered into the peritoneal cavity and (2) 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.^[8,11]

An algorithm for the selection of the type of surgery has been furnished by Dhage-Ivatury and Sugarbaker.^[11] It envisages several factors: (1) whether or not a mucocele is perforated; (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 the right colectomy, including cytoreductive surgery, heated intraoperative intraperitoneal chemotherapy, early postoperative intraperitoneal chemotherapy.^[11]

In our patient the mucocele was not perforated (no discharge into the peritoneum cavity), there was no pathologic process in the base of the appendix (negative margins of resection), and the regional lymph nodes were negative. Therefore, only appendectomy was performed, which is an adequate surgery in such a case. Also, according to the algorithm, no long-term follow-up is advised for our patient.

In conclusion, appendiceal mucocele is a rare disease and has a clinical picture that resemble 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 open surgery should be favoured against laparoscopic surgery.

CONFLICT OF INTEREST :

The authors declared no conflict of interest

FUNDING : None

REFERENCES

1. Rokitansky CF. A manual of pathological anatomy. Vol 2 English translation of the Vienna. edition (1842). Philadelphia: Blizard and Lea, 1855:89.
2. Woodruff R, McDonald J. Benign and malignant cystic tumors of the appendix. *Surg Gynecol Obstet.* 1940; 71:751-5.
3. Misdradi J, Yantiss RK, Graeme-Cook FM, Balis UJ, Young RH. Appendiceal mucinous neoplasms. A clinicopathologic analysis of 107 cases. *Am J Surg Pathol.* 2003; 27 :1089-103.
4. Stocchi L, Wolff BG, Larson DR, Harrington JR. Surgical treatment of appendiceal mucocele. *Arch Surg.* 2003; 138:585-90.
5. 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 :1525-41.
6. 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 :860-4.
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 :1525-41.
8. 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
9. Garcia Lozano A, Vazquez Tarrago 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 : 108-112
10. Lien WC, Huang SP, Chi CL, Liu KL, Lin MT, Lai TI et al. Appendiceal outer diameter as an indicator for differentiating appendiceal mucocele from appendicitis. *Am J Emerg Med.* 2006; 24 : 801-805
11. Dhage-Ivatury S, Sugarbaker PH. Update on the surgical approach to mucocele of the appendix. *J Am Coll Surg.* 2006; 202 : 680-684.