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APPENDICEAL MUCOCELE: A RARE CASE REPORT

General Surgery

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ABSTRACT

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 (appendicectomy/colectomy) to avoid severe intraoperative and postoperative complications. Here, we are presenting a case of 50 years old female admitted with chief complaints of right iliac fossa pain for 4 months. With the help of USG and CECT preoperative diagnosis of Appendiceal Mucocele was made. Appendicectomy was performed. Intraoperatively a appendiceal mucocele with dimensions 6 cm × 5 was found and appendicectomy was done. Histopathological diagnosis reported, recurrent appendicitis with mucin flecks in the luminal surface. After 6 months of surgery patient is doing well with no postoperative complications. **Conclusion:** In our opinion every patient more than 50 years old presenting in emergency department with clinical symptoms of acute appendicitis must undergo CT and open surgery should be favored against laparoscopic surgery.

KEYWORDS

Appendiceal lump, appendicitis, appendicular mucocele, appendicular mass

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 0.3 to 0.7% of the appendectomies [2]. It is characterized by the dilation of the organ lumen with mucus accumulation. Appendix mucocele may come as a consequence of obstructive or inflammatory processes, cystadenomas or cysta denocarcinomas [3]. Besides these causes, other tumor lesions in the appendix or cecum may present as mucocele [4]. Its main complication is pseudomyxoma peritonei.

Case Study

A 50 years old female came to Out Patient Department of Department of General surgery, with 4 months history of vague pain abdomen, more localized to right lower abdomen, associated with generalized weakness, nausea and decreased appetite from last 4 months, no history of surgeries in the past. Patient reported mild right tenderness in right iliac fossa on palpation. She was afebrile. On further evaluation, laboratory investigations showed leucocytosis with neutrophilia. Abdominal ultrasonography showed encapsulated cystic lesion in the lower quadrant of the abdomen with a liquid content of variable echogenicity-?Appendicular abscess / Mucocele appendix. Abdominal CECT was done which showed well circumscribed low attenuating tubular mass contiguous with the base of the caecum showing thin curvy linear mural calcifications with few low attenuating areas along the surface of the lesion f/s/o Mucocele of Appendix. The patient was taken to operation theater. Under all aseptic precautions, under spinal anaesthesia, laparotomy was done with Lanz incision. Mucocele of size 6 cm × 5 cm was identified at the base of the caecum (fig 1). Mesoappendix along with appendicular artery was ligated and cut. Appendix identified and cut (fig 3). Penrose drain placed after achieving hemostasis. Histopathological report showed recurrent appendicitis with mucin flecks in the luminal surface of the appendix. After 6 months of surgery patient is doing well with no postoperative complications.

Fig 1 – intraoperative picture of appendiceal mucocele

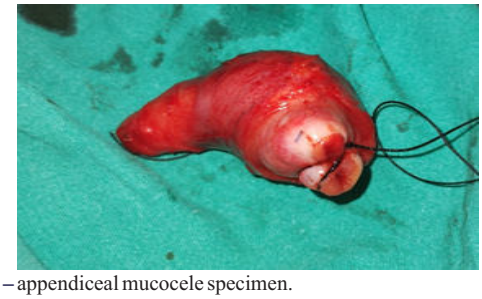

Fig 2 – appendiceal mucocele specimen.

DISCUSSION

The mucocele of the appendix - 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. While some articles confirm its prevalence among women [5,6], others demonstrate a higher incidence among men [7,8]. Mucocele in the appendix may be classified according to the histological characteristics of lumen obstruction [9]

- Simple mucocele (inflammatory, obstructive or retention cyst) - 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.
- The mucinous cystadenoma is an appendix neoplasm with dysplastic epithelium similar to colon adenomatous polyps.
- The mucinous cystadenocarcinoma presents high grade cellular dysplasia and stromal invasion, besides muscularis mucosae.
- In both types described, the mucus material contains epithelial adenoma cells with low or high grade of dysplasia. The rupture of the appendix may lead to the dissemination of the epithelium that produces mucins in the abdominal cavity, causing mucinous ascites or pseudomyxoma peritonei. The clinical flow of the disease does not have a specific picture. It often flows asympto matically. In about 50% of the 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. 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 [10,11]. 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 [12–15]. 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. By colonoscopy an elevation of the appendiceal orifice is seen and a yellowish mucous discharge would be visible from this orifice. 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. 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.

An algorithm for the selection of the type of surgery has been furnished by Dhage-Ivatury and Sugarbaker [16]. 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 [16]. In our case mucocele appendix was not perforated without any involvement of lymph nodes, so appendicectomy was performed.

Treatment of pseudomyxoma peritonei is variable, both due to the rarity of the disease and to its frequently slow-growing nature [17]. Current treatment strategies range from watchful waiting

to extensive cytoreductive surgery alone or with hyper thermic intraoperative peritoneal chemotherapy (HIPEC) or early postoperative intraperitoneal chemotherapy (EPIC) [18]. Based on the Sugarbaker peritonectomy procedure, a recent study showed that cytoreductive surgery with intraperitoneal hyper thermic perfusion permitted complete tumor removal, confirming the efficacy of this combined treatment in terms of improved long-term survival and better regional control of the disease [19]. However, other studies support that fluorouracil-based adjuvant systemic chemotherapy should be the standard of care for patients with PMP of appendiceal origin [20]. In situations where surgery is not immediately required, patients can be monitored via CT scans, tumor markers, laboratory tests, and physical symptoms, to determine when, and if, surgery is warranted. Since the risk of developing an adenocarcinoma of the colon is 6 times greater in patients with a mucocele than in the general population, colonic surveillance is warranted in these cases [21]. The prognosis of patients with pseudomyxoma peritonei was very poor, with limited life expectancy and no chances of healing. The cytoreduction associated with hyperthermic intraperitoneal chemotherapy has reached survival rates in five years of 50% to 96%, in selected cases, when peritoneal cytoreduction is complete and there are no distant metastases [22].

CONCLUSIONS

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 open surgery should be favored against laparoscopic surgery.

REFERENCES:

[1] C.F. Rokitansky, A Manual of Pathological Anatomy, Vol. 2, Blancard and Lea, Philadelphia, 1855, pp. 89, English translation of the Vienna edition (1842).
[2] R. Woodruff, J. McDonald, Benign and malignant cystic tumors of the appendix, Surg. Gynecol. Obstet. 71 (1940) 751–755.
[3] E. Higa, J. Rosai, C.A. Pizzimbono, L. Wise, Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendyx. A re-evalutation of appendiceal mucocele, Cancer 32 (6) (1973) 1525–1541.
[4] D.K. Driman, D.E. Melega, G.A. Vilos, E.A. Plewes, Mucocele of the appendix secondary to endometriosis. Report of two cases, one with localized psedomyxoma peritonei, Am. J. Clin. Pathol. 113 (6) (2000) 860–864.
[5] J. Misdraji, R.K. Yantiss, F.M. Graeme-Cook, U.J. Balis, R.H. Young, Appendiceal mucinous neoplasms. A clinicopathologic analysis of 107 cases, Am. J. Surg. Pathol. 27 (8) (2003) 1089–1103 [Links].
[6] L. Stocchi, B.G. Wolff, D.R. Larson, J.R. Harrington, Surgical treatment of appendiceal mucocele, Arch. Surg. 138 (2003) 585–590.
[7] J. Ruiz-Tovar, D.G. Teruel, V.M. Castineiras, A.S. Dehesa, P.L. Quindós, E.M.Molina, Mucocele of the appendix, World J. Surg. 31 (3) (2007) 542–548.
[8] S.H. Kim, H.K. Lim, Mucocele of the appendix: ultrasonographic and CT findings, Abdom. Imaging 23 (3) (1998) 292–296.
[9] E. Higa, J. Rosai, C.A. Pizzimbono, L. Wise, Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendyx. A re-evalutation of appendiceal mucocele, Cancer 32 (6) (1973) 1525–1541.
[10] P.H. Sugarbaker, Appendiceal epithelial neoplasms and pseudomyxoma peritonei, a distinct clinical entity with distinct treatments, in: K.J. Bland, M.W. Büchler, et al., General Surgery. Principles and International Practice, Springer, London-Limited, 2009, pp. 885–893.
[11] S. Dhage-Ivatury, P.H. Sugarbaker, Update on the surgical approach to mucocele of the appendix, J. Am. Coll. Surg. 202 (4) (2006) 680–684.
[12] W.C. Lien, S.P. Huang, C.L. Chi, K.L. Liu, M.T. Lin, T.I. Lai, et al., Appendiceal outer diameter as an indicator for differentiating appendiceal mucocele from appendicitis, Am. J. Emerg. Med. 24 (7) (2006) 801–805.
[13] G. Francica, G. Lapiccirella, C. Giardibello, et al., Giant mucocele of the appendix: clinical and imaging finding in 3 cases, J. Ultrasound Med. 25 (5) (2006) 643–648.
[14] B.A. Birnbaum, S.R. Wilson, Appendicitis at the millennium, Radiology 215 (2) (2000) 337–348.
[15] K. Sasaki, H. Ishida, et al., Appendiceal mucocele: sonographic findings, Abdom. Imaging 28 (1) (2003) 15–18.
[16] S. Dhage-Ivatury, P.H. Sugarbaker, Update on the surgical approach to mucocele of the appendix, J. Am. Coll. Surg. 202 (4) (2006) 680–684.
[17] R. Buell-Gutbrod, K. Gwin, Pathologic diagnosis, origin, and natural history of pseudomyxoma peritonei, Am. Soc. Clin. Oncol. Educ. Book (2013) 221–225.
[18] S. Dhage-Ivatury, P.H. Sugarbaker, Update on the surgical approach to mucocele of the appendix, J. Am. Coll. Surg. 202 (2006) 680–684.
[19] M. Deraco, D. Baratti, M.G. Inglese, et al., Peritonectomy and intraperitoneal hyperthermic perfusion (IPHP): a strategy that has confirmed its efficacy in pseudomyxoma peritonei, Ann. Surg. Oncol. 11 (2004) 393–398.
[20] C.F. Chen, C.J. Huang, W.Y. Kang, J.S. Hsieh, Experience with adjuvant chemotherapy for pseudomyxoma peritonei secondary to mucinous adenocarcinoma of the appendix with oxaliplatin/ fluorouracil/leucovorin (FOLFOX4), World J. Surg. Oncol. 6 (2008)118.
[21] M.P. Federle, V.S. Anne, Mucocele of the appendix, in: M.P. Federle (Ed.), Diagnostic Imaging, Amirsys, Abdomen Salt Lake City, UT, 2004, pp. 26–27.
[22] V.J. Verwaal, F.A.N. Zoetmulder, Follow-up of patients treated by cytoreduction and chemotherapy for peritoneal carcinomatosis of colorectal origin, Eur. J. Surg. Oncol. 30 (3) (2004) 280–285.
[23] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating consensus surgical case report (SCARE) guidelines, Int. J. Surg. (60) (2018) 132–136.