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Case Report

Appendicular Mucocele Complicated by Peritoneal Pseudomyxoma

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Abstract

Appendiceal mucocele (AM) or Appendiceal mucinous neoplasms is a rare but potentially dangerous condition. In the latter, a spontaneous or iatrogenic rupture of the mucocele can lead to mucinous intraperitoneal ascites, a syndrome known as pseudomyxoma peritonei. We present the case of a 76-year-old female patient who reported right iliac fossa pain with RIF tenderness on clinical examination, and underwent an abdominal CT scan showing an appendiceal mucocele associated with a small peritoneal effusion suspecting a ruptured mucocele, related to peritoneal pseudomyxoma. The patient underwent appendectomy with peritoneal lavage and aspiration of all mucin found. Anatomopathological analysis confirmed the diagnosis of appendiceal mucocele (a low-grade appendiceal mucinous neoplasm).

Keywords: Appendicular mucocele, pseudomyxoma peritonei, appendectomy.

INTRODUCTION

Appendiceal mucocele is defined as cystic dilatation of the appendiceal lumen as a result of intraluminal accumulation of translucent, gelatinous, mucinous secretions, which may affect either the entire organ or a segment, most often distal [1].

The size of the mucocele is highly variable, ranging from a few centimeters to very large masses (40 cm); It is said to be “giant” when it reaches large sizes [2].

Since its first description, this pathological entity has piqued the interest of several authors due to its rarity, its nonspecific clinical presentation, the difficulty of its preoperative diagnosis (AM is histologically discovered postoperatively in nearly 70% of cases) [3], the absence of histological uniqueness, its controversial etiopathogenesis and its particular complications.

It poses the dual problem of its possible malignancy and the risk of gelatinous disease of the peritoneum (peritoneal pseudomyxoma) in the event of perforation [3].

CASE REPORT

Mrs K.F, 76 years old, who has a past medical history of arterial hypertension treated by losartan and amlodipine, a dyslipidemia treated by fenofibrate. And an hepatitis C virus (HCV) with a sustained virological response. She had also a past Surgical history of Cholecystectomy 1 year ago and a Recurrent umbilical hernia

The history of the disease dates back to 1 year before her admission to our department, with an onset abdominal pain in the right iliac fossa, without fever, transit disorders or externalized hemorrhage. The patient's general condition was marked by asthenia and weight loss.

Clinical examination revealed tenderness in the right iliac fossa (RIF)

- Biological workup showed no inflammatory syndrome (protein c reactivation: 6 and leukocytes: 4200).
- Abdominal pelvic ultrasound revealed a cystic pericaecal mass.
- An abdominal CT scan was therefore requested for a better characterization and showed hydric distension of the appendix, measuring 20 mm with a thin wall, containing parietal calcifications, associated with a thin layer of adjacent effusion, free peritoneal effusion of low perihepatic and pelvic level suspecting a ruptured mucocele.

The patient underwent colonoscopy to look for a synchronous tumor, which was incomplete, preventing exploration of the cecum due to the presence of solid stools.

On the basis of these clinical and paraclinical examinations, the diagnosis was Appendicular mucocele complicated by peritoneal pseudomyxoma

Our patient was transferred to the visceral surgery department, where she underwent a laparoscopic appendectomy with peritoneal lavage and aspiration of all mucin found.

Pathological analysis of the surgical specimen (appendix) showed a low-grade appendiceal mucinous neoplasm, and cytoreduction analysis confirmed the presence of a peritoneal pseudomyxoma (acellular mucin, WHO 2019) with no invasive elements.

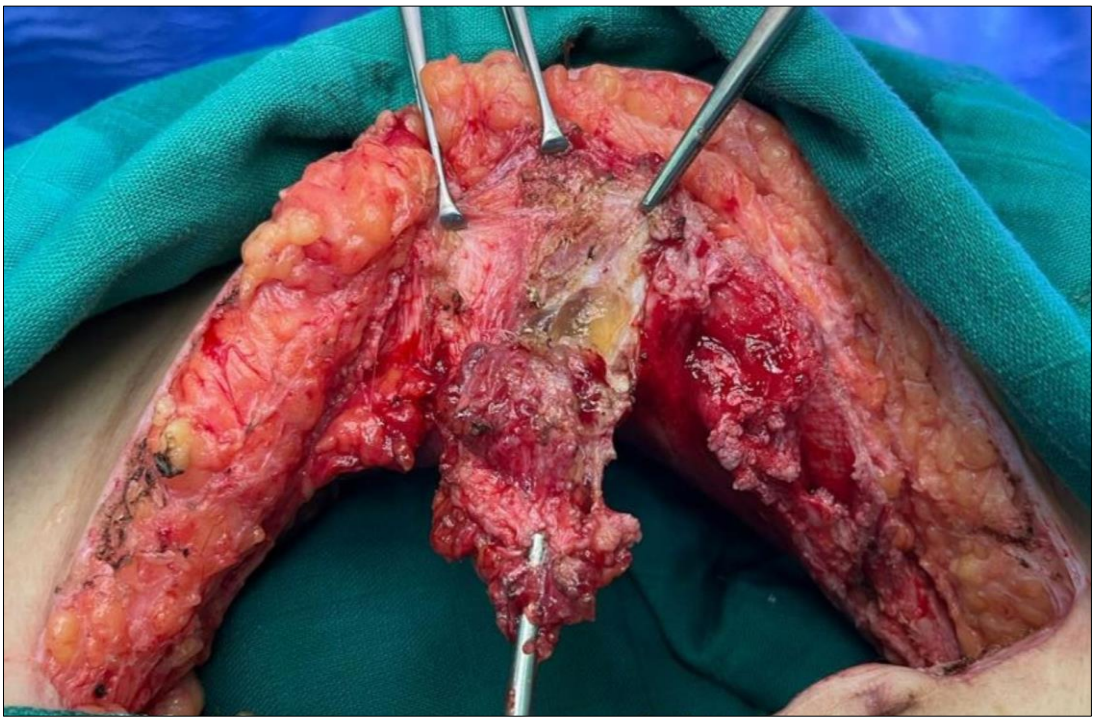

Figure 1: Swollen appendix perforated in the apex and containing mucin

DISCUSSION

AM is a rare pathology (0.2 to 0.3% of appendectomy specimens), histologically discovered postoperatively in almost 70% of cases [3, 4], and frequently reported in female patients over 50 years of age, as in our patient's case [5].

From an anatomopathological standpoint, four types of histological lesions can be distinguished [1-6].

Retention cyst: resulting from obstruction. There is accumulation of mucinous debris in de lumière with increase of the endoluminal pressure resulting in epithelial hyperplasia

Epithelial villous hyperplasia
Mucinous cystadenoma
Mucinous cystadenocarcinoma

Appendiceal mucocele has a highly variable and nonspecific clinical presentation and in 23 to 50% of cases the lesion is discovered peroperatively. The patients could also be asymptomatic [7, 8].

The most frequently encountered presentations are: appendicitis-like painful forms and pseudotumoral forms.

1) asymptomatic Form

Approximately 23% to 50% of patients with appendiceal mucocele are asymptomatic [9]. The lesion is then discovered by chance during imaging (CT scan, ultrasound....), colonoscopy [10], or even intraoperatively or postoperatively by anatomopathological study of the resection specimens [11].

2) Painful Pseudoappendiceal Forms

It's the most frequent form and amounts to 48% of the cases (WESSER and EDELMANN) [12].

It is reported that up to 50% of patients with appendiceal mucocele may have symptoms compatible with typical acute appendicitis [13].

3) Pseudotumoral Forms

The lesion presents as a non-painful mass, of variable sizes, elongated, firm and mobile [14, 15], or as an abdomino-pelvic mass. These forms are more likely to be diagnosed preoperatively on the basis of clinical and, above all, radiological evidence.

4) Complicated Forms

In some cases, appendiceal mucocele is revealed by complications, including the following:

Peritoneal pseudomyxoma (PMP) 10% to 15% of appendiceal mucoceles progress to PMP [16], due to intraperitoneal rupture of a mucinous tumour. The fundamental histological feature is the presence of extracellular mucin in the peritoneal cavity, which may be associated with more or less well-differentiated mucinous epithelial cells.

Complications such as appendicular volvulus [17, 18], rectorrhagia, melena [19, 20] or urological complications due to a mass effect on the right ureter may also reveal AD [7].

Ultrasound-CT imaging is of vital importance for preoperative diagnosis. The typical mucocele appears on both ultrasound and CT as a caecal-based, rounded, well-limited, thin-walled mass with fine parietal calcifications; density on CT is variable, from fluid to tissue. The wall of the mucocele may be thickened and irregular, with contrast-enhancing nodules, suggesting a cystadenocarcinoma; however, there is no radiological sign to confirm or exclude with certainty the malignancy of the underlying appendiceal tumor [22].

Complications secondary to mucocele can also be visualized on a CT scan or ultrasound: intussusception, torsion, ureteral compression.

Colonoscopy is a non-specific examination that confirms the integrity of the caecal mucosa and allows us to look for any related colonic tumors [3].

It can show a submucosal mound in the cecum, if the appendicular orifice is seen in the center of this mound. This sign is known as the "volcano sign" [1].

Preoperatively, whether it is during the clinical or paraclinical phase, numerous differential diagnosis of appendiceal mucocele should be taken into consideration. Chez la femme il faudra éliminer: A simple cyst or an ovarian tumor [23], notably because the latter can be associated with a mucocèle. Hydrosalpinx [24], which can be confused with mucocele on ultrasound.

Treatment of appendiceal mucocele is always surgical, ranging from simple appendectomy in benign forms to right hemicolectomy in malignant mucocele.

Benign mucoceles require conventional appendectomy (Mac Burney or laparoscopy), with the usual surgical asepsis measures to prevent peritonitis [21]. Simple appendectomy seems sufficient for several authors, sometimes combined with resection of the caecal fundus, to ensure complete removal of the lesion. Some authors suggest that an extemporaneous examination has to be carried out if malignancy is suspected [25].

Treatment of pseudomyxoma peritoneal pseudomyxoma has 3 essential aims

- Evacuate ascites.
- Investigate and treat the initial tumor(s) and peritoneal lesions.
- Prevent recurrence and complications whenever possible.

There are essentially two main types of surgical management for PMP: multiple surgical debulking and cytoreductive surgery (CCR) with perioperative intraperitoneal chemotherapy: hyperthermic intraperitoneal chemotherapy (CHIP) with or without immediate postoperative intraperitoneal chemotherapy (IPIC).

In our case, the patiente was treated by cytoreduction surgery without CHIP.
The evolution of these conditions is unpredictable. It depends on the nature and quality of the surgical procedure.

CONCLUSION

When a right iliac fossa mass or appendicular syndrome is discovered or persists, appendicular mucocele should be considered, even though it is rare. Preoperative diagnosis is possible and important, requiring ultrasound and/or CT scan to alert the surgeon to the risk of rupture during surgery, and to avoid pseudomyxoma of the peritoneum. Systematic histological study of all appendectomy specimens should be the rule. Management consists of simple appendectomy in the majority of cases, but a right hemicolectomy should be performed systematically in cases where signs of local malignancy are evident or confirmed by pathological analysis of the surgical specimen.

REFERENCES

1. Moujahid, M., Ali, A. A., Achour, A., & Janati, M. I. (2010). Mucocčle appendiculaire: ā propos de dix cas. *Journal africain du cancer/African Journal of Cancer*, *2*(2), 107-111.
2. Francica, G., Lapiccirella, G., Giardiello, C., Scarano, F., Angelone, G., De Marino, F., & Molese, V. (2006). Giant mucocele of the appendix: clinical and imaging findings in 3 cases. *Journal of ultrasound in medicine*, *25*(5), 643-648.

3. Souei-Mhiri, M., Tlili-Graies, K., Derbel, F., Hmissa, S., Dahmen, Y., & Jeddi, M. (2001). Mucocele of the appendix. Retrospective study of 10 cases. *Journal de Radiologie*, *82*(4), 463-468.
4. Matsui, H., Igarashi, N., Okamura, A., Itano, O., Koyama, Y., & Miyakita, M. (2007). Laparoscopy-assisted resection of an appendiceal mucinous cystadenoma. *Tokai J Exp Clin Med*, *32*(4), 140-143.
5. Touzani, M. A., Regragui, S., & Slaoui, A. (2019). Retroperitoneal mucocele of the appendix: diagnosis and management. *Urol Nephrol Open Access J*, *7*(5), 96-99.
6. Abreu Filho, J. G. D., & Lira, E. F. D. (2011). Mucocele of the appendix: appendectomy or colectomy?. *Journal of Coloproctology (Rio de Janeiro)*, *31*, 276-284.
7. Haritopoulos, K. N., Brown, D. C., Lewis, P., Mansour, F., Eltayar, A. R., Labruzzo, C., & Hakim, N. S. (2001). Appendiceal mucocoele: a case report and review of the literature. *International surgery*, *86*(4), 259-262.
8. Soweid, A. M., Clarkston, W. K., Andrus, C. H., & Janney, C. G. (1998). Diagnosis and management of appendiceal mucoceles. *Digestive diseases*, *16*(3), 183-186.
9. Scotté, M., Laquerrire, A., Riff, Y., Majerus, B., Manouvrier, B., & Leblanc, I. (1994). Les mucocèles appendiculaires: physiopathologie et implications thérapeutiques. *Journal de chirurgie (Paris. 1908)*, *131*(6-7), 303-312.
10. Gillion, JF, Franco, D., Chapuis, O., Serpeau, D., Convard, JP, Jullès, MC, ... & Berthelot, G. (2009). Appendicular mucoceles, pseudomyxomas peritonei and mucosecreting tumors of the ileocecal junction: contribution of imaging in the therapeutic strategy and the choice of laparoscopic or classical approach. *Journal of Surgery*, *146* (2), 150-166.
11. Wakunga, E., Mukuku, O., Bugeme, M., Tshiband, M., Kipili, A., Mobambo, P., ... & Wakunga, W. (2014). Mucocèle appendiculaire: à propos d'un cas observé à Lubumbashi. *Pan African Medical Journal*, *18*(1).
12. Wesser, D. R., & Edelman, S. (1961). Experiences with mucoceles of the appendix. *Annals of Surgery*, *153*(2), 272.
13. Bennett, G. L., Tanpitukpongse, T. P., Macari, M., Cho, K. C., & Babb, J. S. (2009). CT diagnosis of mucocele of the appendix in patients with acute appendicitis. *American Journal of Roentgenology*, *192*(3), W103-W110.
14. Wolff, M., & Ahmed, N. (1976). Epithelial neoplasms of the vermiform appendix (exclusive of carcinoid). II. Cystadenomas, papillary adenomas, and adenomatous polyps of the appendix. *Cancer*, *37*(5), 2511-2522.
15. Soweid, A. M., Clarkston, W. K., Andrus, C. H., & Janney, C. G. (1998). Diagnosis and management of appendiceal mucoceles. *Digestive diseases*, *16*(3), 183-186.
16. García Lozano, A., Vázquez Tarrago, A., Castro García, C., Richart Aznar, J., Gómez Abril, S., & Martínez Abad, M. (2010). Mucocele apendicular: presentación de 31 casos. *Cir. Esp.(Ed. impr.)*, 108-112.
17. Chan, K. P. (1965). Volvulus Complicating Mucocele of the Appendix. *Br J Surg*, *52*, 713-4.
18. Akers, D. L., Hendrickson, M. F., Markowitz, I. P., & Kerstein, M. D. (1988). Volvulus of an appendiceal mucocele presenting as a small bowel obstruction. *J La State Med Soc Off Organ La State Med Soc*, *140*(11), 29.
19. Dachman, A. H., Lichtenstein, J. E., & Friedman, A. C. (1985). Mucocele of the appendix and pseudomyxoma peritonei. *American journal of roentgenology*, *144*(5), 923-929.
20. Corder, A. P., Masters, A., & Heald, R. J. (1990). Sigmoid invasion as a late complication of mucinous cystadenoma of the appendix: report of a case. *Diseases of the colon & rectum*, *33*, 619-620.
21. Weber, G., Teriitehau, C., Goudard, Y., De Saint-Blancard, P., Minvielle, F., Chavihot, C., ... & Baccialone, J. (2009). Appendicular mucocele. *Radiology Papers*, *49* (1), 40-44.
22. Fairise, A., Barbary, C., Derelle, A. L., Tissier, S., Granger, P., Marchal, F., ... & Régent, D. (2008). Mucocele of the appendix and pseudomyxoma peritonei. *JOURNAL DE RADIOLOGIE-PARIS-*, *89*(6), 751.
23. Higa, E., Rosai, J., Pizzimbono, C. A., & Wise, L. (1973). Mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma of the appendix. A re-evaluation of appendiceal "mucocele". *Cancer*, *32*(6), 1525-1541.
24. Creuze, N., Savoye-Collet, C., Lemoine, F., Tapon, E., Ribeiro, C., & Thiebot, J. (2008). Mucocèle sur moignon appendiculaire. *Journal de radiologie (Paris)*, *89*(1), 57-59.
25. Macek, D., Jafri, Z. H., & Madrazo, L. (1992). Ultrasound Case of the Day. *Radio Graphics*, *12*, 1247-9.