Case report

Appendicular mucocele, a case report

ABSTRACT

Appendicular mucocele is a rare condition in which the appendix is dilated, either mechanically or due to benign or malignant tumors. Diagnosis is evoked by an appendicular mass syndrome and imagery based on ultrasound-CT. The most feared complication is perforation, resulting in peritoneal pseudomyxoma. Recommended treatment is always surgical, sometimes combined with intraperitoneal chemotherapy. In this work, we report the case of an 81-year-old patient admitted for abdominal pain with a mass syndrome in the right iliac fossa, whose exploration evoked an appendicular mucocele. He underwent a right hemicolectomy with transverse ileo anastomosis by laparotomy. Postoperative management was straightforward. Pathological anatomy of the surgical specimen indicated a mucinous cystadenoma.

***Keywords**: mucocele; appendix; abdominal pain; cystadenoma; haemicolectomy*

1. INTRODUCTION

Appendiceal mucocele, a rare but potentially dangerous condition worldwide, corresponds to dilatation of the appendix through accumulation of mucus secondary to obstruction of its lumen, either mechanical in the case of ligature or stercolith, or locoregional in the case of benign tumors: endometriosis, villous adenoma villeux or malignant: carcinoma, carcinoid tumor. (1).

The most dreaded complication is perforation, resulting in pseudomyxoma peritonei, formerly known as gelatinous disease of the peritoneum.

Preoperative diagnosis, which is generally difficult, is nowadays facilitated by medical imaging, notably ultrasound and CT scans, which enable the diagnosis to be evoked and appropriate surgical management to be proposed. (16)

Anatomopathological examination of the surgical specimen remains the definitive diagnosis, and should be carried out systematically.

Surgical management can range from simple appendectomy for benign tumors to right hemicolectomy for cancers. (15)

2. CASE REPORT

We report the case of an 81-year-old patient with no medical or surgical history, who was referred to our department for management of right iliac fossa pain.

On clinical examination, the patient was in good general condition, neurologically, hemodynamically and respiratorily stable. Physical examination revealed a normal-volume abdomen with a palpable mass painful on deep palpation in the right iliac fossa and right flank. There were no associated urinary signs. Rectal examination was normal.

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Biologically, the patient presented neutropenia with leukocytes at 3000 elements/mm3. He also had normochromic normocytic anemia with hemoglobin at 9.5g/dl. Platelets were normal at 159,000. Prothrombin rate was 78%. C-reactive protein (CRP) was 40.30mg/l. Ionogram and renal function were normal. Carcinoembryonic antigen **(**CEA) was increased to 12.72 ng/ml.

Radiological features were suggestive of an appendicular mucocele. Abdominal ultrasound and CT scan revealed a well-limited oblong formation connected to the caecal fundus, with a thin wall, homogeneous fluid content and calcification measuring 88 x 39 mm.

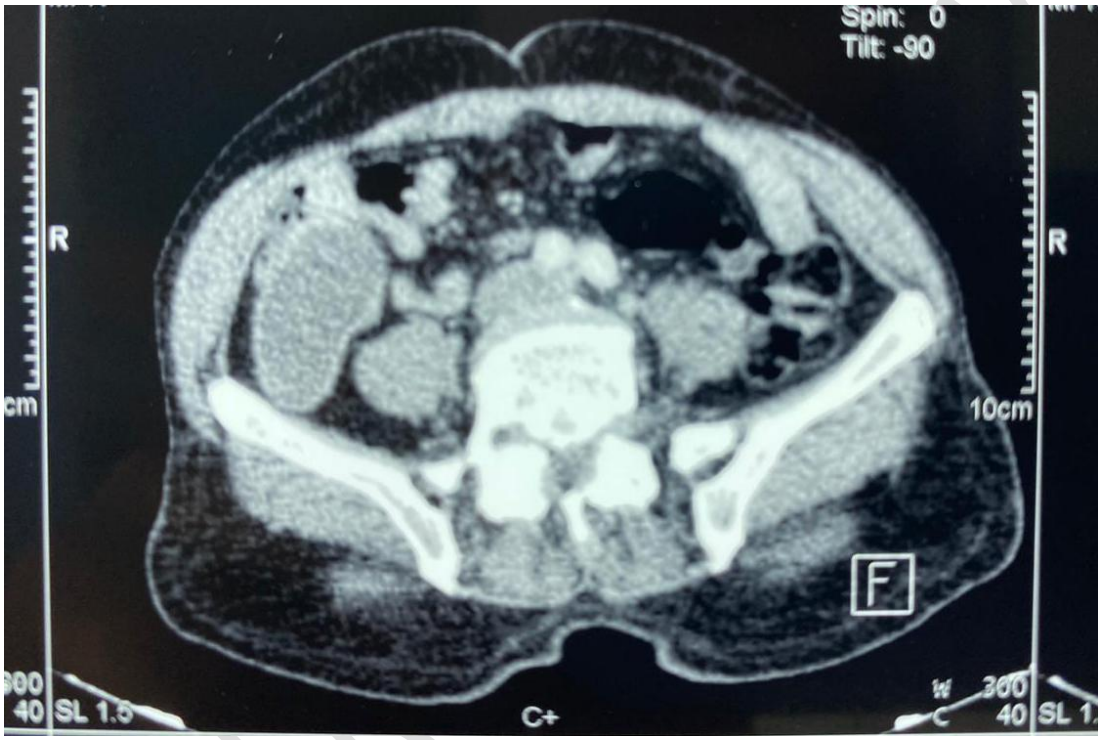

Fig 1: Abdominal CT in axial section after IV injection of the contrast product

MRI revealed a formation corresponding to an oblong structure with a large vertical axis projecting from the right iliac fossa, with a wall similar to that of the digestive tract, and a liquid content with a homogeneous T2 hypersignal.

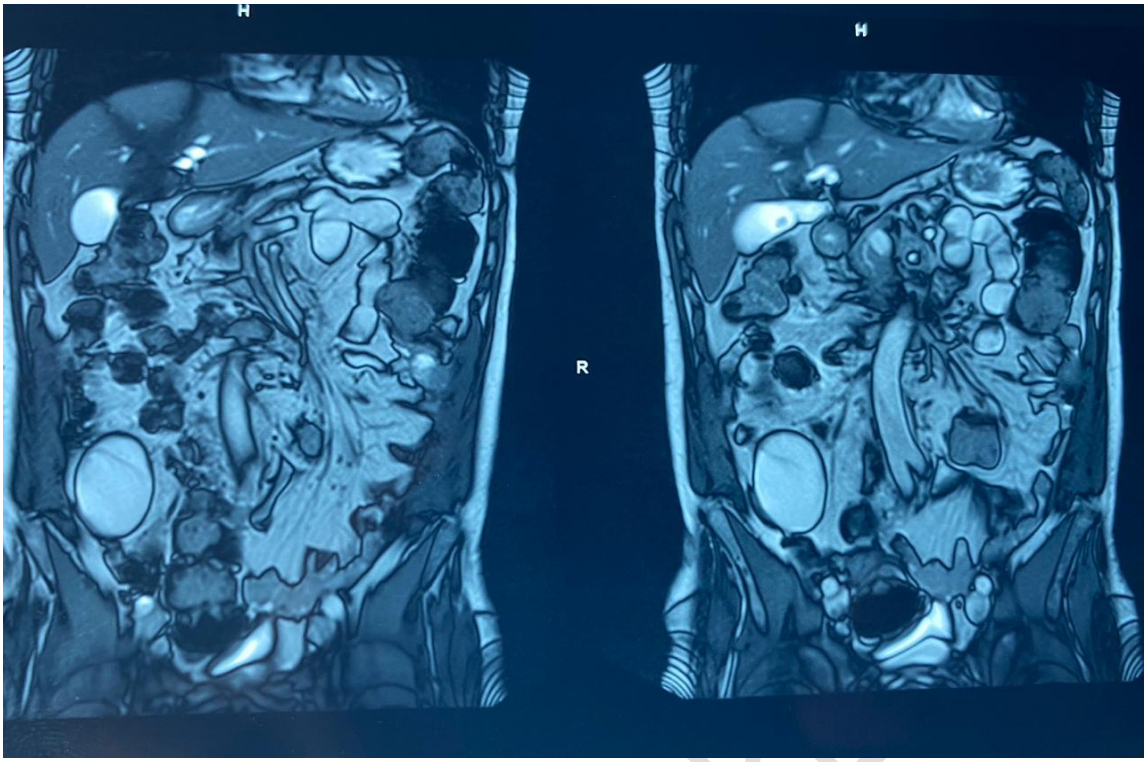

Fig 2: Abdominal MRI in longitudinal section

The patient was scheduled and then operated on. The approach was the median subumbilical route under general anaesthesia. Intra-abdominal exploration revealed a clean abdominal cavity, without ascites or peritoneal carcinosis. The mass attached to the cecum was renitent. The patient received an intraoperative transfusion. A right hemicolectomy was performed with mechanical ileotransverse anastomosis, without perforation of the tumor mass

Postoperative management was simple. Pathological anatomy revealed a cystic formation whose lining was replaced by a flattened epithelioma with a clear cytoplasmic mucosecretory wall. The lumen was occupied by a mucoid substance punctuated by inflammatory elements. This was a low-grade appendicular mucinous neoplasia in relation to a mucinous cystadenoma with healthy resection margins without effraction.

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Fig 3: Right hemicolectomy surgical specimen

3. DISCUSSION

First described by Rokitansky in 1842 and named by Feren in 1876, appendicular mucocele is an uncommon condition accounting for 0.15 to 0.6% of appendectomies (1).

The mean age of diagnosis is 59.6 years (2). Our patient, a man was older. The sex ratio varies widely from one series to another, with either female predominance (3-4), male predominance (5) or equal distributions (6). The clinical presentation of the disease is not typical. Like our patient, the most frequent presentation is pain in the right iliac fossa, similar to that of acute appendicitis, or a palpable abdominal mass associated with nausea or vomiting. However, around 25% of patients are asymptomatic and discovered incidentally (7).

Tumour markers such as carcinoembryonic antigen, Carbohydrate antigen 19-9 (CA19-9) and Cancer antigen 125 (CA-125) are non-specific, but should be measured and systematically repeated to monitor disease progression, as available evidence suggests that their elevated levels correlate with advanced tumour stage in the majority of patients (12).

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Imagery plays a fundamental role in the preoperative diagnosis of appendicular mucocele. An unprepared abdominal X-ray, not performed in our case, would reveal fine arciform calcifications opposite the appendicular cavity in half of all cases (8).

The most effective imaging test is the abdomino-pelvic CT scan with iodine contrast injection. On abdomino-pelvic magnetic resonance imaging (MRI), the mucocele presented as a cystic pericaecal lesion in T1 hyposignal, frank T2 hypersignal with parietal contrast after injection in our observation. This aspect of the pathology has been described by authors in the literature (8). In patients with mucocele, the risk of developing adenocarcinoma of the colon is six times higher than in the general population (9). Also for women, the diagnosis of a mucocele requires a search for the association with an ovarian mucinous tumour. MRI is only useful for assessing the extent of peritoneal pseudomyxoma following mucocele perforation (14).

Surgery was the treatment carried out in our patient, and the post-operative course was uncomplicated. The treatment of appendiceal mucocele is based on surgery alone or combined with intraperitoneal hyperthermic chemotherapy in case of peritoneal gelatinous disease (10, 11). As in our patient's case, excision must be performed without invading the tumour wall, otherwise it could result in a peritoneal pseudomyxoma with a worse prognosis (13). Laparotomy is therefore recommended for safe handling and extraction of the surgical piece.

4. CONCLUSION

Appendicular mucoceles are rare. They should be suspected in the presence of an atypical appendicular syndrome or a mass in the right iliac fossa. This symptomatology requires ultrasound and CT scans for early diagnosis. Tumor markers, notably the carcinoembryonic antigen, are non-specific but enable postoperative follow-up of mucinous disease. Similarly, MRI is only useful for assessing the extent of peritoneal pseudomyxoma. Treatment is surgical, with laparotomy as the preferred approach to avoid any tumour effraction, which darkens the prognosis.

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REFERENCES

1. 1. Fairise A, Barbary C, Derelle AL, Tissier S, Granger P, Marchal F et al. Appendicular mucocele and peritoneal pseudomyxoma. JRadiol.2008; 89(6): 751-62.
2. 2. Shaib WL, Goodman M, Chen Z, et al. Incidence and survival of appendixal mucinous neoplasms: a SEER analysis. Am J Clin Oncol 2017;40:569-73.
3. 3. Aho AJ. (1973). Benign and malignant mucocele of the appendix: histologic types and prognosis. Acta Shir Scand, 139, 392–400.
4. 4. Scotté, M, Laquerrire A, Riff Y, Majerus B, Manouvrier B, and Leblanc I. (1994). Appendicular mucoceles: pathophysiology and therapeutic implications. Journal of Surgery (Paris. 1908), 131(6-7), 303-312.
5. 5. Mary A. (2008). Appendectomy by laparotomy for appendicitis. EMC. Elsevier Masson SAS, Paris.
6. 6. Wolff M, and 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), 2511-22.

7. 7. Demetrashvili Z, Chkhaidze M, Khutsishvili K, Topchishvili G, Javakhishvili T, Pipia I, Qerqadze V. Mucocele of the Appendix: Case Report and Review of Literature. International Surgery. 2012;97(3):266-269.doi:10.9738/CC139.1.

8. 8. Rouchaud A, Glas L, Gayet M, Bellin MF.Mucinous cystadenoma of the appendix. Journal of Diagnostic and Interventional Radiology 2014; 95 (1): 113-116

9. 9. Thomas HL, MD, Felter FD. Mucocele of the appendix. Baylor University Medical Center Proceedings 2014; 27 (1): 33-34

10. 10. Goéré D, Dartigues P, Elias D. Treatment of appendicular mucoceles. Hépato Gastro2011; 18: 581-588

11. 11. Ingo H, Michael M, Thomas R. Appendiceal Mucinous Cystadenoma. RadioGraphics 2008;28: 1524-1527

12. 12. Carmignani CP, Hampton R, Sugarbaker CE, Chang D, Sugarbaker PH. Utility of CEA and CA 19-9 tumor markers in diagnosis and prognostic assessment of mucinous epithelial cancers of the appendix. J Surg Oncol 2004;87:162-6

13. 13. Zanati F. Appendicular mucocele. J Chir. 2007; 144(7):146. PubMed | Google Scholar

14. 14. Derelle AL, Tissier S, Granger P, Barbary C, Rousseau A, Laurent V, et al. Early diagnosis of pseudomyxoma peritonei localized to perforated appendixal mucocele: Imaging and pathologic aspects. J Radiol 2007;88:289-95

15. 15. Moujahid M, Ali A, Achour A, Janati MI. Appendicular mucocele: about ten cases. African Journal of Cancer. 2010;2(2):107–111.

16. 16. Lorenzon L, De Dominicis C, Virgilio E, Balducci G. The appropriate management of an appendix mucocele. BMJ Case Rep 2015; doi:10.1136/bcr-2014-209045

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