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 tumours. It is evoked on the basis of appendicular mass syndrome and ultrasound-CT imaging. The most feared complication is perforation leading to peritoneal pseudomyxoma. The recommended treatment is always surgical, sometimes combined with intraperitoneal chemotherapy. In this paper 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 revealed an appendicular mucocele. He underwent a right hemicolectomy with transverse ileo anastomosis via laparotomy. Postoperative management was uncomplicated. The pathological anatomy of the surgical specimen revealed 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 due to accumulation of mucus as result of obstruction of its lumen, either mechanical in the case of ligature or stercolith, or locoregional in the case of benign tumours: endometriosis, villous adenoma villeux or malignant: carcinoma, carcinoid tumour. (1).

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

Preoperative diagnosis, which is generally difficult due to non- specific clinical symptoms, is nowadays facilitated by medical imaging. The biology is not specific. Ultrasound and computed tomography (CT) have specific features that allow the suspiscion of an appendicular mucocele, the assessment of its benign or malignant nature, and initiate appropriate surgical management (16).

Surgical management which can be performed by laparotomy or laparoscopy can range from simple appendectomy for benign tumors to right hemicolectomy for cancers. (15)

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

2. CASE PRESENTATION

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

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 elevated 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.

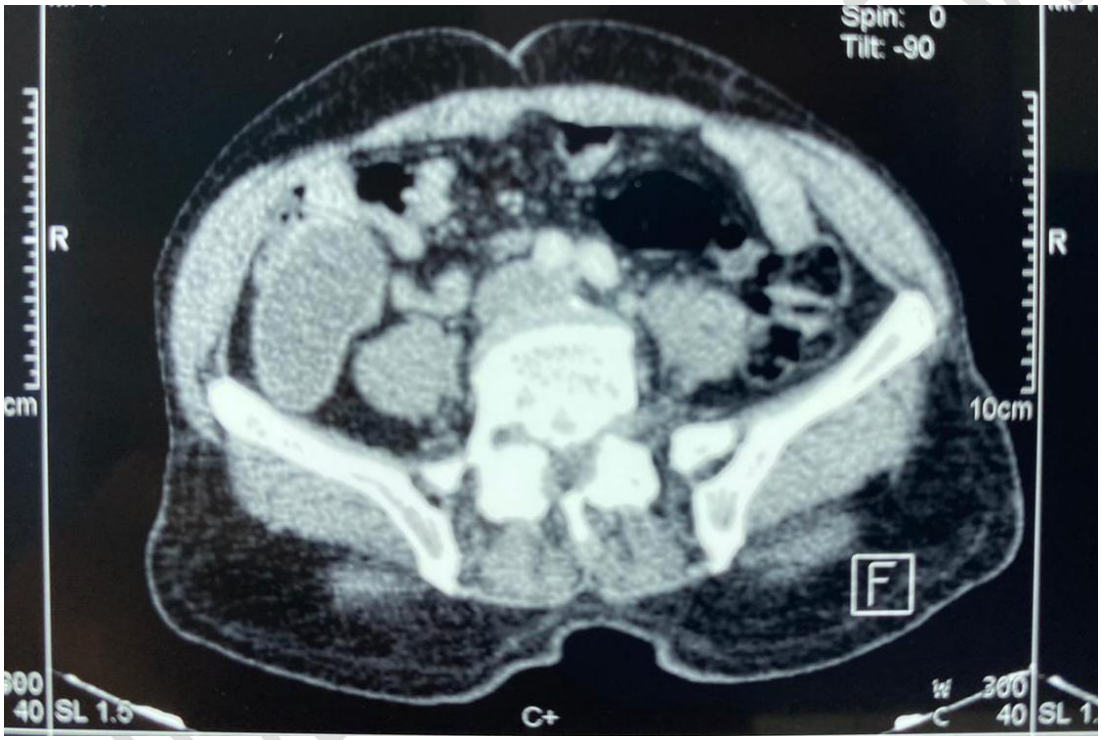

Fig 1: Abdominal CT in axial section after IV injection of the contrast product

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

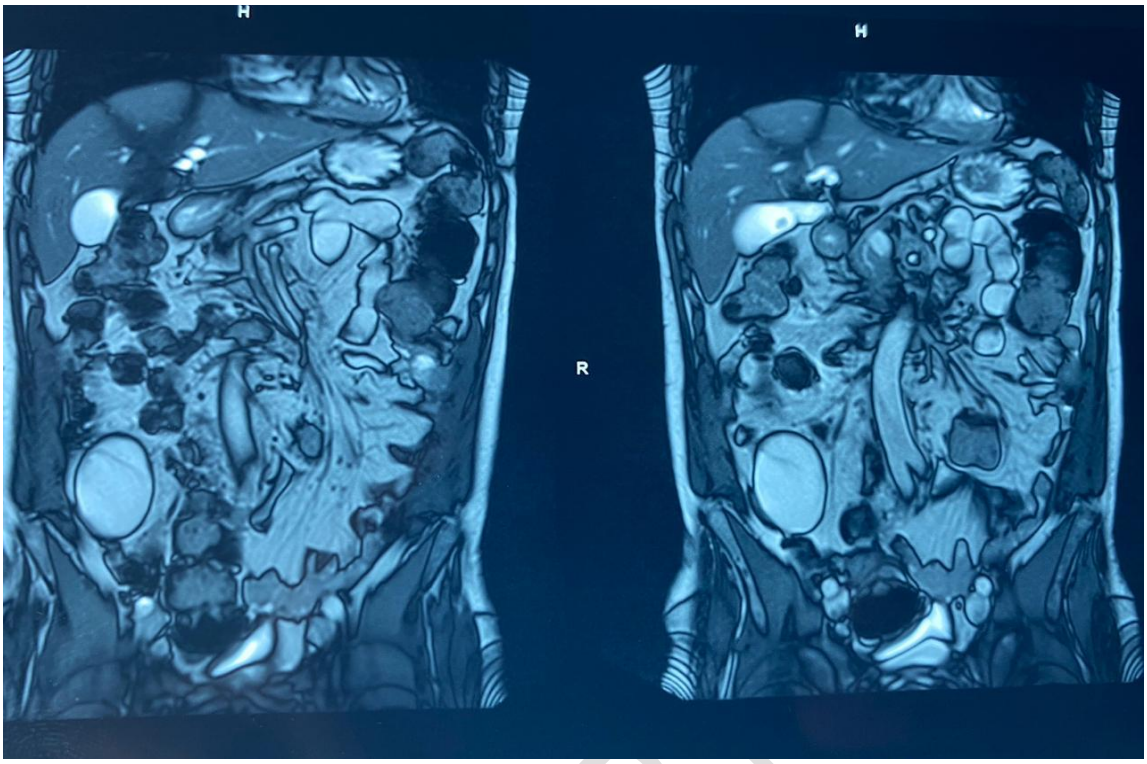

Fig 2: Abdominal MRI in longitudinal section

The patient was scheduled for surgery. 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 with mechanical ileotransverse anastomosis was performed without perforation of the tumor mass

Postoperative management was simple. Pathological anatomy revealed a cystic formation with the lining replaced by a flattened epithelioma with a clear cytoplasmic mucosecretory wall. The lumen was interspersed 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.

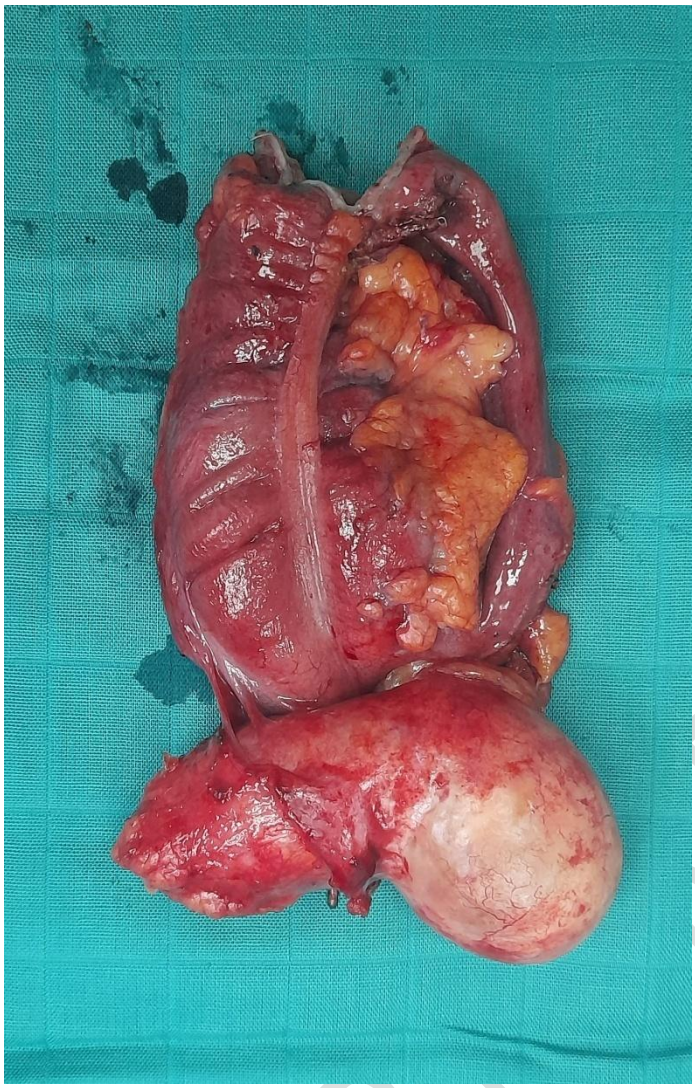

Fig 3: Right hemicolectomy surgical specimen

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 average age at diagnosis is 59.6 years (2). Our patient, a male was older. The sex ratio varies widely from series to series, with female predominance (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 (17). However, approximately 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).

Imaging 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). Certain characteristics like diameter larger than 1.5 cm, “onion skin” appearance, nodular enhancing of wall on ultrasonography are suggestive of appendicular mucocele (11) .The colonoscopy can reveal a mount-like elevation of the appendix orifice, also known as the ‘volcano sign’ (3) and also used for the diagnosis of synchronous or metachronous colon cancer when present; no preoperative colonoscopy was done in our case.

The most effective imaging test is the abdomino-pelvic CT scan with iodine contrast injection. The presence of larger lesion, soft tissue thickening, wall irregularity, egg cell calcifications with normal wall thickness on CT are suggestive of neoplastic nature of lesion while the presence of peri-appendiceal inflammation or abscess is suggestive of appendicitis and is not usually found in appendiceal mucinous lesions (11). On abdomino-pelvic magnetic resonance imaging (MRI), the mucocele in our observation 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 colorectal adenocarcinoma 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). 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). The choice between an open approach and laparoscopy is controversial. In terms of management, there is a lack of standardized treatment approach for appendicular mucocele without initial apparent metastasis (6). Both open surgery and laparoscopic surgery have yielded positive results in the literature (3-20). Laparoscopy has been used in many recent cases (17-18-19); dissection has been meticulous, with care taking care to avoid spillage of contents leading to the formation of a peritoneal pseudomyxoma. Laparotomy has generally been preferred for safe handling and removal of the surgical specimen, but laparoscopic surgery has recently become increasingly popular, offering the advantages of good exposure and assessment of the entire abdominal cavity as well as faster recovery by avoiding a large incision and a better cosmetic result (18).

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, in particular the carcinoembryonic antigen, are non-specific but allow postoperative follow-up of mucinous disease. Similarly, MRI is only useful to assess the extent of peritoneal pseudomyxoma. Treatment is surgical. Laparotomy is the preferred approach to avoid tumour effusion, which worsens prognosis, but laparoscopic approaches are increasingly being used.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

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