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Case Report

Appendiceal Mucocele: A Case Report

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Abstract

Appendiceal mucocele is a rare pathology, which poses a double problem by its potential malignancy and the risk of peritoneal pseudomyxoma in case of perforation. We report here the case of a 57 year old patient operated for anal fistula in whom the clinical examination revealed a mass of the right iliac fossa. Ultrasound showed a cystic pericaecal mass. The patient underwent a laparoscopic appendectomy removing the caecal base and the appendicular operative specimen measured 130 mm in length and 64 mm in diameter. Pathological analysis of the specimen confirmed the diagnosis of appendiceal mucocele without malignant cells. The postoperative course was simple and the patient was discharged on the sixth postoperative day. The aim of this study was to show the diagnostic and therapeutic features of this condition in our context.

Keywords: Mucocele, tumor, appendectomy.

INTRODUCTION

Appendiceal mucocele or appendiceal mucosecretory tumor is defined as a cystic dilatation of the lumen of the appendix as a result of intraluminal accumulation of translucent, gelatinous, mucinous secretions, which may involve either the entire organ or a segment, usually distal.

Mucinous distension of the appendicular lumen may be of tumoral or non-tumoral origin, benign or malignant [1, 11]. According to the literature, it represents 0.15 to 0.6% of appendectomy specimens [2, 3].

According to the modern classification, there are 4 histological types:

Retention cyst, mucinous hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma [4, 5].

The clinical presentation of the disease is not specific. It is often asymptomatic. In about 50% of cases, it is discovered accidentally during radiological and endoscopic examinations or during surgery. Clinical symptoms may include right iliac fossa pain, a palpable abdominal mass, nausea, vomiting, weight loss, gastrointestinal bleeding, or signs of intestinal invagination [6-10].

Preoperatively, it is essential to recognize an appendiceal mucocele, in order to adapt the surgical procedure. Imaging currently plays an important role in the diagnosis, but the definitive diagnosis is based on the histological study, which must be systematic for all appendectomy specimens. Treatment ranges from simple appendectomy in benign forms to right hemicolectomy for cancer in malignant mucocele [11].

Appendiceal mucocele poses the double problem of its possible malignancy and the risk of gelatinous disease of the peritoneum (pseudomyxoma peritoneum) in case of perforation.

PATIENT AND OBSERVATION

We present the case of Mr. M.L, 57 years old, married and father of 3 children with a history of anal fistula operated 26 years ago.

He was admitted to our hepato-gastro-enterology department for a mass in the right Iliac fossa.

The history of his illness goes back to 6 months after his admission date with the fortuitous discovery during auto palpation of a mass in the right iliac fossa which was progressively increasing in volume and causing a sensation of discomfort.

No transit disorder, no externalized digestive bleeding, no fever, no alteration of general condition.

The clinical examination revealed a well-limited 4-5 cm deep, hard, indolent mass in the iliac fossa.

On the paraclinical level:
The biological work-up did not reveal an inflammatory syndrome:
C-reactive protein at 5 and leukocytes at 7310 /UL.

In addition, a congenital factor VII deficiency was discovered fortuitously in the presence of a low prothrombin level after elimination of other causes: no hepatocellular insufficiency, no cholestasis, and no signs of disseminated intravenous coagulation or use of anticoagulants.

Patient was treated with a FFP protocol with good evolution.

Abdominal ultrasound showed a mass in the right Iliac fossa with fluid content in intimate contact with the cecal wall.

Abdominal CT scan with injection showed: Cystic formation (23 HU) of the right iliac fossa.

Thickened wall which is enhanced after injection of contrast medium and continues with the appendix the appendix measuring: 64* 61*70 mm (AP *T* H) suggesting an appendiceal mucocele appendicular.

Total colonoscopy: a submucosal mass at the level of the caecum, yellowish, with a mucinous consistency of about 25 cm.

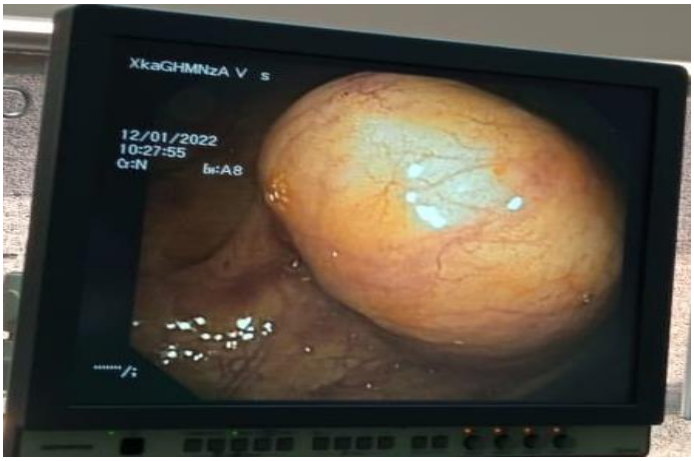

Figure 1: Colonoscopic aspect of an appendiceal mucocel

Our patient was transferred to the visceral surgery department where he underwent a laparoscopic appendectomy removing the caecal base.

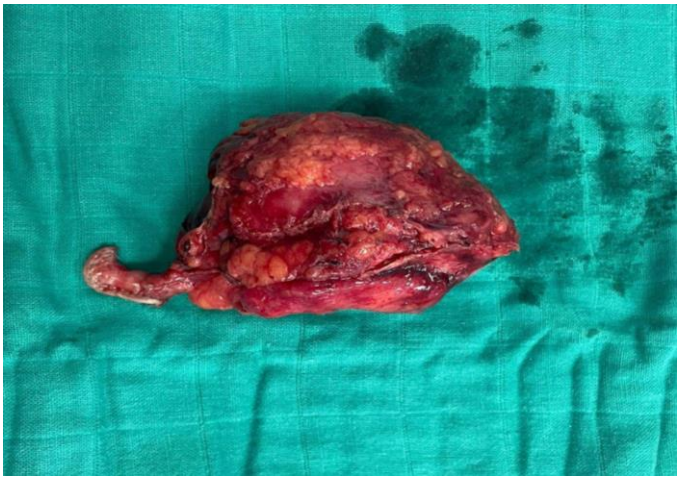

Figure 2: Surgical specimen (front view, dimensions: 13x6x5.5 cm)

Anatomopathological analysis of the surgical specimen confirmed the diagnosis of appendiceal mucocele without malignant cells, of the mucinous cystadenoma type with low grade dysplasia.

The postoperative course was simple and the patient was discharged on the sixth postoperative day. He was reviewed at six months and at one year; no obvious signs of locoregional recurrence were noted.

1-year follow-up CT scan: No abdominal-pelvic mass syndrome.

DISCUSSION

First described by Rokitansky in 1842 and named by Feren in 1876 [12, 13], appendiceal mucocele is an uncommon condition accounting for 0.15 to 0.6% of appendectomies [2-12, 14], this incident varies with series reported in the literature. It is due to dilatation of the appendicular lumen following accumulation of intraluminal mucinous secretions.

It is found preferentially in adults with an average age between 50 and 60 years as we noted in our observation.

As for the sex, it is a man in our observation but the sexe ratio varies according to the series. Moreieurs studies show the nette predominance of females in this affection [15-17], with a sexe ratio of four females to one male while others found an equale distribution between the both sexes [18] and others owhere a predominance of males was reported [19, 20].

Positive diagnosis is a essential step in the treatment of appendiceal mucocele as de it derives the choice of treatment eand the prognosis. However, it was rarely done en preoperatively given the clinical et even paraclinical non- specificity of times de this condition.

Two clinical situations are the most frequently rencontrated: the painful forms simulating a simple or complicated acute appendicite, generally leading to surgical intervention from the outset et the pseudotumorales forms where the lesion presents as a mobile or fixed mass of the right iliac fossa or as an abdomino-pelvic. The latter are more accessible to a preoperative diagnosis based on clinical and especially radiological arguments as in the case of our patient.

However, asymptomatic forms discovered fortuitously during laparotomy done for another pathologie [21] or a radiological examen [22] or endoscopic [22] or forms complicated by peritoneal pseudomyxoma, intestinal occlusion, appendicular volvulus, invagination in the coecum, or digestive bleeding may exist.

Imaging plays a fundamental role in the preoperative diagnosis of appendiceal mucocele. The most effective imaging examination is the abdominal-pelvic CT scan with injection of iodinated contrast medium at portal time [23-26].

Typical mucocele appears as a base caecale, rounded en bienlimited, thin-walled masse with finee parietale calcifications; the density en scanner varies, from liquidienne to tissulaire.

A stercolithe sometimes visible to base de appendice. The wall of the mucocele may be thickenede, irregulare, with contrast-enhancing nodule orienting to a cystadenocarcinome; however, there existe no radiological signe to affirm or exclude with certainty the malignancy of the underlying appendicular tumor.

Our patient, the abdomino-pelvic CT scan showed: a cystic formation (23 HU) of the right iliac fossa, well limited and round, with a thickened wall which is enhanced after contrast injection which continues with the appendix measuring: 64*61*70 mm (AP *T* H) suggesting an appendiceal mucocele.

The treatment of appendicular mucocele is based on surgery alone or associated with intraperitoneal chemohyperthermia in case of peritoneal gelatinous disease [27, 28]. However, this surgery must meet certain imperative criteria which are complete removal of the appendix, passing through a healthy zone at the base and the absence of intraoperative appendicular trauma which could lead to dissemination of mucus and epithelial cells in the peritoneum. Surgery was the only treatment performed in our patient and the postoperative course was simple.

In our case, the histology of the appendectomy specimen confirmed the diagnosis of appendicular mucinous cystadenoma as low grade dysplasia.

CONCLUSION

Appendicular mucocele is a rare pathology with a varied and non-specific clinical presentation. Medical imaging based on ultrasound and scannographic exploration usually allows the diagnosis to be evoked and specific surgical management to be proposed. Anatomopathological analysis of the surgical specimen is systematic to confirm the diagnosis and to look for signs of malignancy.

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