

Case Report

Low-Grade Appendiceal Mucinous Neoplasm and Acute Appendicitis. Clinical Case Report

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Summary

Low-grade appendiceal mucinous neoplasm corresponds to a rare histopathological lesion of the vermiform appendix and is considered even rarer when it manifests as an acute appendicitis. A clinical case of a 37-year-old female patient, with no clinical or surgical history, no allergies, is presented, who went to the emergency service by acute appendicitis chart and after surgery an appendicular mucocele was obtained, this is a generic terminology that has been the subject of controversy for its concept, histopathological classification and postoperative clinical evolution. Also discussed is open versus laparoscopic handling, several authors exclusively recommend open handling, because in laparoscopic management they report an increased risk of accidental opening of the surgical part with the subsequent possibility of developing Pseudomyxoma Peritonei. Some series of cases report laparoscopy as a treatment considering the dimensions of the appendix and the expertise of the surgeon. Clinical follow-up is very important considering possible condition to neighboring organs, disease progression and other possible complications; it is done at 6, 12 months and 5 years; with analysis of complementary laboratory, imaging and endoscopic examinations. According to this result is determined a medical or surgical treatment.

Keywords: Appendicular Mucocele; Low-Grade Appendiceal Mucinous Neoplasm; Pseudomyxoma Peritonei

Introduction

Low-grade appendicular mucinous neoplasm (LAM) corresponds to a rare pathological entity, estimated only between 0.1 and 0.3% of histopathological reports of appendectomy [1,2]. LAMN are among some of the benign and malignant causes of the term appendicular mucocele (AM) which is solely descriptive and is the source of a controversy in the medical literature. The AM was first recognized as a pathological entity by Carl Freiherr von Rokytansky in 1842 and in 1876 Faren began the use as medical term [3,4].

The proper use of terminology has been the reason for several scientific meetings to reach a consensus of clinical and histopathological concepts in order to make a correct diagnosis and / or surgical in addition to short, medium and long term follow-up according to the results of the analysis of complementary laboratory, imaging and endoscopic examinations. In 2012 in the city of Berlin

all these parameters were analyzed, for the correct nomenclature of mucinous neoplasms of the appendix, adenocarcinomas and Pseudomyxoma peritonei (PMP); was held at the World Congress of the Group of International Oncology of the Peritonei Surface (PSOGI). Here a large number of pathologists, oncologists and surgeons experts on the subject were referred; they came to the modified *Delphi Consensus*, which recognizes LAMN and high-grade appendiceal mucinous neoplasm (HAMN), within mucinous neoplasms and appendix adenocarcinomas [5]. The incidence occurs mainly considering sex, age, in women within the fifth and sixth decade of life [6]. Cases have also been reported at younger ages and in males [3,7].

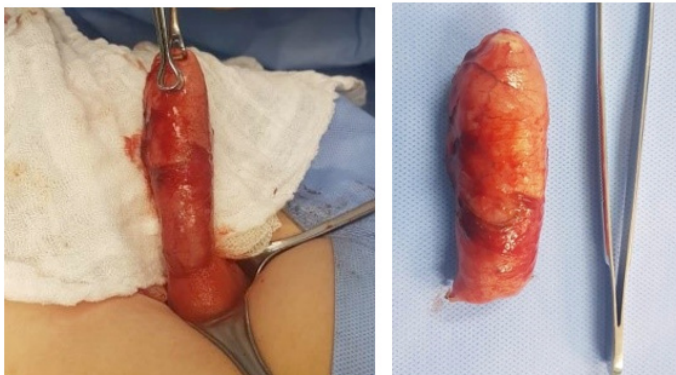
Clinical manifestations of LAMN are nonspecific, may be asymptomatic or manifest as chronic pain and/or palpable mass in the right lower quadrant of the abdomen along with symptoms and gastrointestinal signs of neoplastic consideration. In most cases it does not manifest and is accidentally found in imaging exams and endoscopy as a mass of apparent origin in blind or right ovary in women and even in surgical findings [8-10]. They report some series of cases that manifestation is less common as acute appendicitis

[2]. The treatment of choice is surgical, only appendicectomy is recommended in case of LAMN that does not exceed the serosa and in the case of PMP when there is spontaneous or accidental opening of AM the first treatment of choice is cytoreducing surgery/ intraperitoneal hyperthermal chemotherapy (HIPEC) [11].

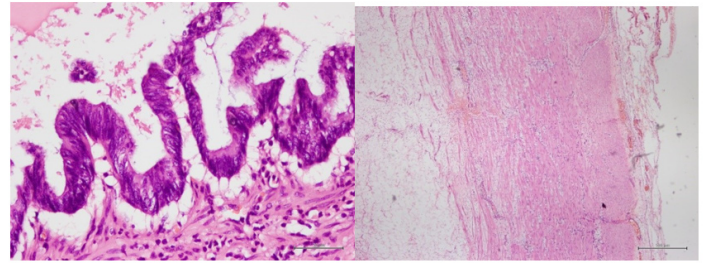
Right hemicolectomy, a appendectomy with partial blind resection or ileal resection are reported as unnecessary in LAMN without PMP and also in those cases reporting the result of histopathology do not exceed the serosa; most authors recommends surgical resolution openly. The follow-up should be every 6 months, then each year, until 5 years due to cancer and PMP risk, is done with the help of laboratory and imaging tests; especially computed tomography (CT), and colonoscopy with biopsy [6,12,13].

Case Report

We present a patient 37-year-old female patient, with no clinical, surgical or allergy history, she came through an abdominal pain picture of approximately 13 hours of evolution, colic type, located in right iliac fossa (RIF), irradiated to flank and hypochondrium of ipsilateral, without any accompanying symptoms. On the physical examination was a tense, painful abdomen in RIF, signs of Mc Burney, Blumberg and Rovsing positive. Hematic biometrics showed leukocytosis with neutrophilia, it was decided to perform emergency surgery on suspicion of AA. Open appendectomy was performed through an incision of Mc Burney, finding a subcecal and subserosa appendix measuring approximately 7 x 2.3 cm (Figures 1,2); in addition to moderately inflamed inflammatory fluid and the blind tumefact at the appendicular base. Post-surgical developments were favorable and were discharged to the third day of hospitalization with control at 8 days and 3 months with the results of histopathology that I conclude in LAMN and AA (Figures 3,4).



Figures 1-2: Surgical part showing mucinous fluid buildup.



Figures 3, 4: Microscopy of histopathological diagnosis showing ulcerated and possibly cylindrical epithelium stratified with hyperchromatic nuclei in cigars, towards light is observed eosinophilic amorphous material (mucin) hypertrophy of the muscle layer and polymorphonuclear infiltrate that compromises all layers to the serosa.

Discussion

We present a case corresponding to an unusual diagnosis within the epidemiological, clinical and histopathological parameters mentioned above, once the case was assessed in the emergency the diagnostic impression of AA was reached performing a surgical procedure and finding as a transoperative finding an AM, which considering several series of cases is not in the mean of the reported age [6]. In addition, the correct open surgical management recommended the greatest amount of medical literature available, although there are certain indications for possible laparoscopic management such as those found accidentally, less than 2cm and not associated with PMP [4]. Our patient only performed appendectomy which is recommended as a treatment in LAMN [6,14]. There is adequate post-surgical evolution with recommended follow-up, laboratory tests such as carcinoembryonic antigen (CEA), CA-19 9 and CA-125 should be performed, also a CT with contrast for total assessment of the abdominal and pelvic cavity, colonoscopy is recommended in some cases with biopsy up to 5 years after appendectomy [11]. Our patient is under follow-up with the negative results of these tests.

Conclusions

LAMN is a very rare entity that must be known for surgical and postoperative management to provide adequate treatment and monitoring of evolution, can be accidentally diagnosed during an imaging examination or in emergency or scheduled surgery.

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