Case Report

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Appendiceal mucinous cystadenoma: a case report and review of the literature

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ABSTRACT

Appendicular mucocele is a low incidence neoplasm with a frequency of 2 cases per 10,000 laparotomies. It is found in 0.2 to 0.3% of all appendectomies, with a higher incidence in women after the fifth or sixth decade of life. It is considered a benign lesion, although in some occasions, approximately 20% of cases it is associated with neoplasms of other intra-abdominal organs. Authors present the case of a 88-year-old patient with clinical findings consistent with acute appendicitis resolved with a simple appendicectomy based on the macroscopic findings during surgery.

Keywords: Appendicitis, Mucocele, Mucinous cystadenoma, Cystadenocarcinoma

INTRODUCTION

Appendicular mucocele is a low incidence neoplasm; it is a benign lesion that macroscopically is described as a dilation of the appendicular lumen due to accumulation of mucinous secretion, generally secondary to an obstruction. The term was proposed by Karl Freiherr von Rokitansky in 1842 who described the transformation of the appendix into a mucus-filled sac, regardless of etiology. 2

Authors described a case of an 88 year old female who presented with symptoms of acute appendicitis with a mucinous cystadenoma of the appendix as a surgical and histopathological finding.

CASE REPORT

Authors present the case of an 88-year-old female with the following medical conditions: chronic history of atrial fibrillation, chronic obstructive pulmonary disease,

systemic arterial hypertension. She presented to the emergency department of our hospital with generalized colic abdominal pain of 3 days of evolution as well as nausea, no vomiting, no fever and no diarrhoea.

Her vital signs on admission were heart rate of 90 beats per minute, 20 breaths per minute, temperature of 36.7 °C and blood pressure of 130/80 mm Hg. Through directed physical examination, the abdomen was found to be soft, depressible, and painful in the right iliac fossa, positive appendicular maneuvers, negative rebound, decreased peristalsis and no peritoneal irritation.

An ultrasound was performed with the following findings: avascular, aperistaltic loop, 12 mm thick, with striation of the pericecal fat.

Laboratory results revealed leukocytes 13×109/L with 88% granulocytes and hemoglobin (Hb) 12.0 gm%. Given the clinical and paraclinical findings consistent with acute appendicitis, emergency surgery was performed.

Appendicectomy was performed and the following surgical findings were found: 10×2 cm retrocecal appendix, dilated in its middle third, healthy base, rubbery consistency, pearly coloration. (Figure 1 and 2).

Histopathological report was: mucinous cystadenoma, acute appendicitis and clean surgical edges.



Figure 1: Pearl-colored appendix.



Figure 2: Appendix dilated in its middle third, with mucinous content.

The patient remained in hospital 3 days without complications, with follow-up by surgical oncology for 3 months.

DISCUSSION

The clinical presentation of appendicular mucinous cystadenoma is not very specific in terms of symptoms, since it can present as an acute appendicitis, abdominal mass, cecal tumor, and even with gynecological symptoms.³ The most common clinical manifestation is pain in the right iliac fossa that simulates acute appendicitis.⁴ Likewise, it is described in the literature that a mass in the right lower quadrant can be palpated in up to 50% of cases on abdominal physical examination.⁵

Complementary studies such as laboratory analysis and imaging tests can be carried out, in ultrasonography studies a hypoechoic lesion can be observed, in the tomography the images are hypodense and homogeneous described as "onion layers".⁶ While in a colonoscopy a

central hole is observed through which mucus comes out, being called the "volcano sign".⁷

Because preoperative diagnosis is difficult, the mucocele is generally intervened with a diagnosis of acute appendicitis and in most cases it is an incidental finding in surgery.⁸

The definitive diagnosis is made through a pathological study of the surgical piece.⁹

Histopathologically, appendiceal mucocele can be divided into 4 pathological types: simple mucocele, mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma. 10

In simple mucocele and in mucosal hyperplasia there is no epithelial atypia, in mucinous cystadenoma a certain degree of atypia has been observed; however, due to the benign nature of the aforementioned variants, the performance of a simple appendectomy is usually curative. On the other hand, cystadenocarcinoma is characterized by the presence of stromal invasion by malignant glands, the presence of mucus and mucus-secreting cells in the peritoneal cavity, so in this case it is usually necessary to perform a right hemicolectomy.

The treatment is totally surgical and can range from appendectomy with free margins for those patients with small lesions without rupture and that do not have a neoplastic appearance.¹³ In cases where a macroscopic lesion with a neoplastic appearance is present, an appendectomy and ileo-cecus appendicular lymphadenectomy will be performed with an intraoperative pathological study.14 If the appendicular stump is affected, a cecal resection will have to be performed and if the lymphadenectomy is positive, the surgery should be extended to a right hemicolectomy plus adjuvant chemotherapy.¹⁵

Filho et al proposed that in appendix tumors as long as the histology is favorable, the base is not involved, the tumor is smaller than 2 cm and the lymph nodes are negative, it is safe to perform a simple appendectomy, however, if any of the conditions described above are not met, they propose to perform a right hemicolectomy.¹⁶

CONCLUSION

Due to the clinical rarity of this condition, there are no official guidelines for the management of mucocele, so nowadays it has been managed according to the personal experience of the surgeon, as well as the case reports described in the literature. The surgery varies from appendectomy to a right hemicolectomy, which depends on several factors: the size of the tumor, its location within the appendix, involvement of the cecum and ileum, presence of collections of mucus, lymph nodes present, and the histology, all of them are considered as the determining factors. For this reason, surgeons must be

prepared for a major surgery such as right hemicolectomy when considering the diagnosis of a mucocele. In this case we performed a simple appendicectomy based on the macroscopic findings during surgery, the subsequent histopathological study was compatible with benign mucinous cystadenoma.

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