

Case Report

Mucinous Appendiceal Neoplasia: Case Report and Literature Review

Rafael Francisco Alves Silva^{1*}, Luiz Carlos Araújo de Sousa¹, Alexandre Rodrigues Alves², Vitor Breves de Paiva², Lívia Zaiden Carvalho Martins de Sá², Marcos Alexandre Lourenço Almeida¹, Gabriel França Costa¹, Marcelo Alencar da Fonsêca³ and Adriano Pamplona Torres⁴

¹Graduate Student, Medical School, University Center of Brasília (UniCEUB), Brasília, Brazil

²General Surgeon Resident, Regional Hospital of Asa Norte (HRAN), Brasília, Brazil

³General Surgeon Resident, Regional Hospital of Gama (HRG), Brasília, Brazil

⁴General Surgeon, Regional Hospital of Asa Norte (HRAN), Brasília, Brazil

Abstract

Introduction: Primary appendiceal tumors are rare occurrences, found in less than 2% of appendectomies. Aim of this study is to report a case of acute appendicitis caused by Low Grade Mucinous Neoplasia (LAMN) and to review histopathological classification and treatment in the literature.

Case report: 75 years-old, female, complaint of pain in the right iliac fossa for 15 days, associated with nausea and hyporexia for 1 day. At physical examination, diffuse pain at palpation of right hemi-abdome. Hemogram showed 15.200 leukocytes, with 4% of rods. Ultrasonography of abdomen without alterations and computed tomography suggestive of appendicitis. During exploratory laparotomy appendix was enlarged, hyperemic with thick walls and hyperemic cecum with signs of ischemia, suggesting a neoplastic process. Opted for right colectomy with primary anastomosis. Patient progresses well with discharge in the 10th postoperative. Histopathological analysis of low grade mucinous neoplasm in the appendix.

***Corresponding author:** Rafael Francisco Alves Silva, Graduate Student, Medical School, University Center of Brasília, UniCEUB, Brasília, Brazil, Tel: +55 61999232232; E-mail: rafalves97@gmail.com

Citation: Silva RFA, Souza LCA, Alves AR, De Paiva VB, Torres AP, et al. (2019) Mucinous Appendiceal Neoplasia: Case Report and Literature Review. J Surg Curr Trend Innov 3: 012

Received: February 13, 2019; **Accepted:** February 15, 2019; **Published:** February 28, 2019

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Discussion/conclusion: The classification of appendiceal tumors is controversial; LAMN is a submucosal restricted epithelial tumor with low degree of cellular atypia. Treatment is based on removal of the primary site of injury by appendectomy or colectomy.

Introduction

Primary appendiceal tumors are rare occurrences, found in less than 2% of appendectomies [1]. The mucinous appendix neoplasia accounts for 0.2-0.7% of the cases [2]. Diagnosis is usually performed in the sixth decade of life [3]. Clinical manifestations range from absence of symptoms, acute appendicitis, non-spastic abdominal pain and palpable abdominal tumor [2]. The surgical treatment can be performed appendectomy or hemicolectomy, according to the extent of the lesion [4]. The aim of this study is to report a case of acute appendicitis caused by Low Grade Mucinous Neoplasia (LAMN) and to review histopathological classification and treatment in the literature.

Case Report

75 years-old, female, hypertensive and diabetic. He was admitted in the Hospital, complaint of pain in the right iliac fossa for 15 days, associated with nausea and hyporexia for 1 day. Deny vomiting, diarrhea and urinary symptoms. Physical examination showed (HR: 88bpm, SatO₂: 98%) semi globous abdomen, abdominal auscultation without alterations, with diffuse pain on right hemi-abdome palpation. Hemogram with 15,200 leukocytes, 4% of rods.

Ultrasonography of total abdomen performed on the day of care without alterations. Computed tomography showed a distended, thickened appendix with adjacent adipose densification, with no associated perforation or collection signs, suggesting a diagnosis of appendicitis (Figure 1). An exploratory laparotomy was indicated for possible appendectomy.

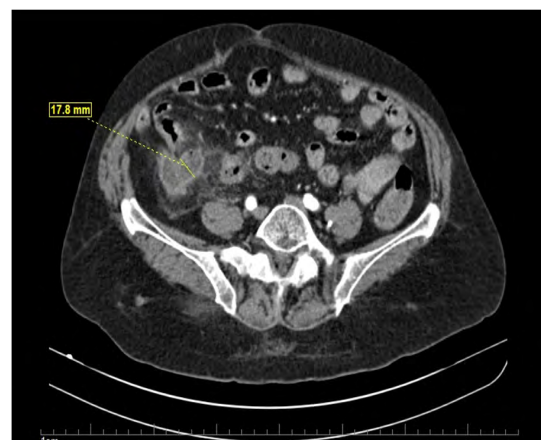


Figure 1: Computed Tomography (CT) of sagittal incidence evidencing a distended appendix, thickened with adjacent adipose densification.

During the intra operative period, the cecal appendix was observed, blocked by the large omentum. The appendix was enlarged, hyperemic with thick walls and hyperemic cecum with signs of ischemia, suggesting a neoplastic process (Figure 2). Absence of extravasation of fecal or mucinous contents in the peritoneal cavity, without lymph node enlargement or peritoneal or hepatic implants. Right colectomy was chosen, with primary anastomosis. Patient progressed uneventfully, being discharged on the 10th postoperative day. On the 17th postoperative day, he returned to the hospital with a complaint of surgical wound infection, opted for conservative treatment with antibiotic therapy and wound cleaning.



Figure 2: Aspect of the appendix during the intraoperative.

Histopathological analysis of the surgical specimen presented cecal appendix with 5.0x3.0cm. Presence of low grade, well differentiated appendiceal mucinous neoplasia with extension in every organ, with tumor free margin and lymph nodes without involvement (Figure 3).

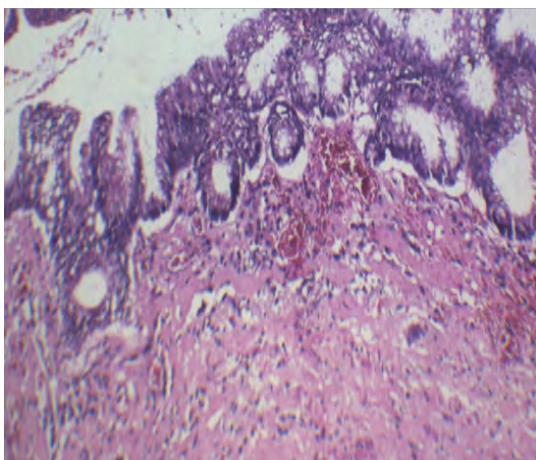


Figure 3: Histopathological analysis of the appendix, H&E stain. Presented dysplastic epithelium, sometimes venous, sometimes tubular, with a presence of mucin in the cytoplasm of the cell.

Discussion

The classification of appendix tumors is controversial. Mucin-producing tumors have epithelial origin, according to WHO classification of 2010, mucin-producing neoplasms were divided into 3 groups: Adenomas, LAMN and adenocarcinoma of the appendix [1].

Currently, by the 2016 Modified Delphi Consensus Protocol the mucinous neoplasms are classified as: LAMN, high grade mucinous neoplasia, mucinous adenocarcinoma with or without signet cell [5].

LAMN and high-grade mucinous neoplasia are lesions restricted to the submucosa and are differentiated by the level of cellular atypia, although these lesions are not malignant, they may present extra appendicular implantation. Mucinous adenocarcinoma is a malignant lesion, which has its own muscular invasion [6]. It is believed that LAMN is the initial neoplastic process, with an intermediate stage marked by high grade atypia followed by mucinous adenocarcinoma. The presence of ring cells is indicative of a worse prognosis [7].

The main complication of LMNA is the rupture of the appendix causing pseudomyxoma peritonei, a clinical condition characterized by peritoneal neoplastic implantation with accumulation of ascites and fatal outcome [1,2,4,7,8].

LMNA treatment is based on removal of the primary lesions by appendectomy [7]. Colectomy is indicated when the tumor is larger than 2cm in size or involvement of the pericecal region or signs of malignancy [4]. In case of pseudomyxoma peritonei, surgical cytoreduction and hyperthermic intraperitoneal chemotherapy are always indicated [4,7,8].

Conclusion

The LMNA of appendix is a rare disease, usually the diagnosis is made in the intraoperative appendectomy. The histopathological classifications are controversial and the treatment varied, so the surgeon and the pathologist must be attentive to the patient's best conduct and follow-up.

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