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A RARE PRESENTATION OF LOW-GRADE APPENDICEAL MUCINOUS NEOPLASM IN A ROUTINE APPENDICECTOMY SPECIMEN

Pathology

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ABSTRACT

Low grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy which is mostly discovered incidentally and can present with a variety of symptoms depending on the clinical manifestations. Due to an extensive accumulation of copious gelatinous material in the appendiceal lumen, the appendix becomes cystically dilated. It can be detected on 0.7 to 1.7% of all appendicectomies. The most concerning side effect of this neoplasm is mucin seeding, which might result in pseudomyxoma peritonei (PMP). Herein we report a 42-year-old female with lower abdominal pain associated with nausea and regurgitation for 1 week. A clinical diagnosis of acute appendicitis was made and laparoscopic appendicectomy was performed. Histopathological study of the appendix revealed a low grade appendiceal mucinous neoplasm.

KEYWORDS

Appendix, mucinous neoplasm, pseudomyxoma peritonei (PMP).

INTRODUCTION:

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy that is detected in 0.7 to 1.7% of all appendicectomies.[1] Histologically, LAMNs are characterized by mucinous epithelium with low-grade cytologic atypia and the absence of any infiltrative growth pattern.[2] LAMNs restricted to the appendiceal lumen do not exhibit obvious malignant characteristics but have the ability to proliferate malignantly outside of the appendix, leading to the formation of pseudomyxoma peritonei, a potentially fatal consequence with a 45% 10- year survival rate.[3] However, the preoperative diagnosis of appendiceal mucinous neoplasm is difficult because of the non-specific clinical manifestations of the disease. We present a case that was thought to be acute appendicitis prior to surgery but was diagnosed histopathologically as LAMN.

Case Report:

A 42-year-old female presented with a complaint of lower abdominal pain with right iliac fossa tenderness for 1 week. A clinical diagnosis of acute appendicitis was made. On ultrasound abdomen probe tenderness was elicited in the right iliac fossa, a peristaltic blind ending tubular, non-compressible measuring approximately 14mm showing peripheral vascularity was noted in the right iliac fossa with edematous wall and the diagnosis was given as features likely suggestive of acute appendicitis. Laparoscopic appendicectomy was done.

We received an appendicectomy specimen measuring 12cm in length with attached peri-appendiceal fat in our Pathology laboratory. On gross examination of the external surface of the appendix, the tip appears bulbous and thickened. (Figure 1) On cut surface, the tip of the appendix was obscured by thick whitish firm area measuring 2x1cm. The entire appendix was embedded and studied.

Histopathological examination of the sections taken from the appendix show mostly ulcerated and denuded mucosa with muscle bundles infiltrated by islands of mucin pools and mucinous material (Figure 2 a and b) with no stratification of the epithelium or loss of polarity except for few cystically dilated glands filled with mucin.(Figure 2c) The histologic type according to the College of American Pathologists protocol (Version: 5.1.0.0) was reported as low-grade appendiceal mucinous neoplasm(LAMN) G1, well differentiated with acellular mucin invading visceral peritoneum(serosa) and mesenteric region focally with pT category as pT4a i.e., tumor invades through visceral peritoneum, including the acellular mucin or mucinous epithelium involving the serosa of the appendix or serosa of the mesoappendix according to pTNM classification(AJCC 9th version). The patient recovered uneventfully post-surgery.

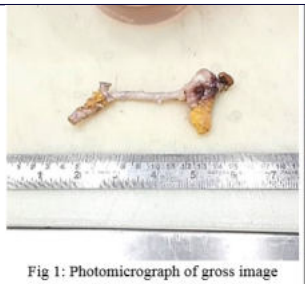

Fig 1: Photomicrograph of gross image showing bulbous and thickened appendix.

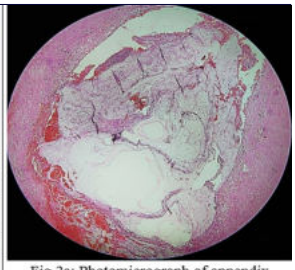

Fig 2a: Photomicrograph of appendix showing ulcerated and denuded mucosa filled with mucin (H&E, 10X).

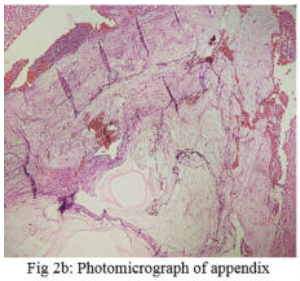

Fig 2b: Photomicrograph of appendix showing ulcerated and denuded mucosa filled with mucin (H&E, 40X).

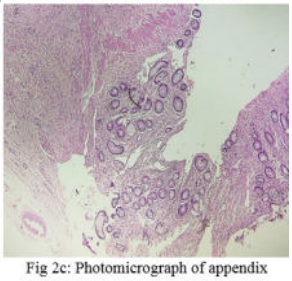

Fig 2c: Photomicrograph of appendix showing cystically dilated glands filled with mucin (H&E, 40X).

DISCUSSION:

One of the most frequent procedures in urgent abdominal surgery is appendicectomy for acute appendicitis. Appendiceal neoplasms usually do not exhibit any particular symptoms before surgery; instead, they are found during histopathological examination.[4] According to the 2019 WHO classification of tumors of the digestive system, appendiceal mucinous neoplasms are divided into serrated lesions, hyperplastic polyps, LAMNs, high-grade appendiceal mucinous neoplasms (HAMNs), and mucinous adenocarcinomas that present with or without appendiceal perforation.[5]

LAMN is a rare condition with a wide range of symptoms and clinical presentations.[6] The appendix becomes cystically dilated because of the substantial buildup of gelatinous material in the appendiceal lumen. It can cause a variety of problems, such as intussusception, volvulus, small bowel obstruction, and rupture. [7]

Grossly, the appendix in LAMN may be unremarkable or dilated due to abnormal mucus buildup and, in some cases, perforation with mucin extrusion. The wall might be considerably thicker and partially attenuated.[8] LAMNs are tumors that exhibit low-grade cytological atypia and minimal architectural complexity and may demonstrate “pushing invasion” with dissection of acellular mucin through the appendiceal wall.[7,9] LAMNs were classified as mucinous neoplasms with low-grade cytology and any combination of the following by the Peritoneal Surface Oncology Group International (PSOGI): loss of the muscularis mucosae, submucosal fibrosis, "pushing" or diverticular-like growth into the wall, dissecting acellular mucin, flattening, or undulating epithelial growth, and mucin or neoplastic cells outside the appendix.[2] According to the American Joint Committee on Cancer (AJCC), LAMNs are categorized as pTis (LAMN) when they are limited to the muscularis propria (and fall into prognostic stage group 0). They are defined as pT3 when acellular mucin or neoplastic mucinous epithelium extends to the sub-serosal soft tissue and as pT4a when they reach the serosa or mesoappendix, respectively. Furthermore, peritoneal deposits of neoplastic mucinous epithelium that contain well-differentiated (G1) epithelium and belong to the same overall prognosis group (stage IVA) are defined as metastatic disease (pM1a) rather than intraperitoneal dissemination of acellular mucin.[2]

Acellular or cellular mucinous deposits throughout the peritoneal cavity may result in pseudomyxoma peritonei (PMP), which is seen in approximately 20% of cases[10], which is a potentially fatal consequence with a 45% 10-year survival rate.[3]

Treatment for all mucinous neoplasms is dependent on histology and stage, and the only curative option is surgical excision. A complete right hemicolectomy with lymphadenectomy is carried out in case of diffuse peritoneal mucinous neoplasms.[7] Follow-up should be continued for the first five years following a diagnosis of LAMN in patients who have a high risk of disease development, as indicated by the presence of lymph node metastases or infiltration of malignancy into the submucosa. Overall, the 3-year survival rate is 91% to 100%, while the 5-year survival is 79% to 86%. [6,7]

CONCLUSION:

The rising number of LAMN instances being documented raises awareness of the condition and advances knowledge of it. Our case is an example to highlight the fact that simple appendicectomy specimen can turn out to be a mucinous neoplasm. Here, the histopathological report plays an important role in definitive treatment and prognosis. Choosing the best surgical or medical treatment option and avoiding PMP recurrence, seeding, and development may be aided by a timely, accurate diagnosis.

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