Case Report

Low-Grade Appendiceal Mucinous Neoplasm: A Rare Case Masquerading as Acute Appendicitis

Ayesha Khatoon, Shakti Kumar Yadav, Sompal Singh, Namrata Sarin, Sonam Kumar Pruthi

Department of Pathology, North Delhi Municipal Corporation Medical College and Hindu Rao Hospital, Delhi, India

Abstract

Appendiceal mucinous neoplasms are an extremely rare group of neoplasms and can range from mucocele to invasive adenocarcinoma. Low-grade appendiceal mucinous neoplasm (LAMN) is an extremely rare form of mucinous neoplasm affecting the appendix. In the majority of patients, it presents with acute abdomen, nausea, vomiting, and change in bowel habits. Pseudomyxoma peritonei is the most dreaded complication. We present a case report of a 35-year-old female who presented to the surgical emergency department of our hospital with complaints of acute abdomen. On examination, a lump was felt in the right iliac fossa, due to which a clinical diagnosis of acute appendicitis with perforation was suspected. However, histopathological examination revealed low-grade appendiceal mucinous neoplasm. Therefore, LAMN should be considered as one of the differentials in patients, who present with acute abdomen and right iliac fossa mass.

Keywords: *Acute abdomen, acute appendicitis, low-grade appendiceal mucinous neoplasm*

Introduction

Primary mucinous neoplasms of the appendix range from simple mucocele to invasive adenocarcinoma.[1] Mucocele of appendix represents an obstructive dilatation of the lumen due to abnormal accumulation of mucus either due to a retention cyst, mucosal hyperplasia, adenoma, and adenocarcinoma. Low-grade appendiceal mucinous neoplasm (LAMN) is rare appendiceal neoplasm, most commonly presents as cystic dilatation of mucocele of the appendix.[2]

On appendicectomy specimen, the incidence rate of appendiceal neoplasms ranges from 0.2% to 0.3%.[3] About 23%–50% of patients are completely asymptomatic.[4] In most of the occasions, these tumors are incidentally detected during surgeries performed for other cause.[5]

LAMN is rare and accounts for <1% of all gastrointestinal malignancies.[6,7] Surgical treatment includes appendicectomy with the removal of mesoappendix or right hemicolectomy.[2] LAMN can also present with diverticula, herniations, rupture, intestinal obstruction, and gastrointestinal bleeding. However, pseudomyxoma peritonei is the most dreaded complication that occurs due to the seeding of the mucin into the peritoneal cavity.[8] Approximately 20% of patients with appendiceal mucinous neoplasm develop pseudomyxoma peritonei.[6]

We present a case of LAMN, which was clinically diagnosed as cecal carcinoma.

Case Report

A 35-year-old female with no significant past medical history presented to the surgical emergency department with right iliac fossa pain, which was radiating to the epigastrium. The pain was associated with nausea and vomiting. On per abdominal examination, a tender mass was felt in the right iliac fossa. Based on the examination findings, a clinical diagnosis of acute appendicitis with perforation and periappendicitis was suspected. An appendectomy was performed, and the specimen was sent to the histopathology department.

On gross examination, the appendix measured 5 cm in length, the lumen was dilated (diameter 3.5 cm), and the cut surface demonstrated a cystic cavity measuring 3 cm × 3 cm, arising from the appendix, extending up to the resected margin. Wall thickness ranged from 0.1 to 0.3 cm. The whole appendix was sectioned and processed.

How to cite this article: Khatoon A, Yadav SK, Singh S, Sarin N, Pruthi SK. Low-grade appendiceal mucinous neoplasm: A rare case masquerading as acute appendicitis. Clin Cancer Investig J 2020;9:165-7.

Submitted: 31-Mar-2019
Revised: 06-Jun-2020
Accepted: 20-Jun-2020
Published: 14-Aug-2020

Address for correspondence:
Dr. Sonam Kumar Pruthi, North Delhi Municipal Corporation Medical College, Hindu Rao Hospital, Delhi - 110 007, India.
E-mail: sonam_manipal@yahoo.com

Access this article online

Website: www.ccij-online.org

DOI: 10.4103/ccij.ccij_47_20

Quick Response Code:

Sections from the appendix showed a cystic cavity lined by columnar cells with basally placed nuclei. The villous and flattened proliferation of the mucinous epithelial cells containing mucin was evident in the sections from appendix [Figure 1]. Nuclear stratification and mild dysplastic changes were noted focally in the lining epithelium [Figure 2].

Mucicarmine and immunohistochemical marker, i.e., CK 20 were applied. Tumor cells demonstrated mucicarmine and CK-20 positivity [Figures 3 and 4].

Based on the histomorphological and immunohistochemical interpretation, a diagnosis of LAMN was made.

Discussion

Primary mucinous appendiceal neoplasms are rare and account for <1% of all the gastrointestinal malignancies.[6,7] Incidence ranges from 0.2% to 0.3%.[3,4] LAMN is more frequent in women and is seen in patients older than 50 years of age.

Common clinical presentation includes lower right quadrant pain, nausea vomiting, change in bowel habits, weight loss, and intestinal obstruction.[9]

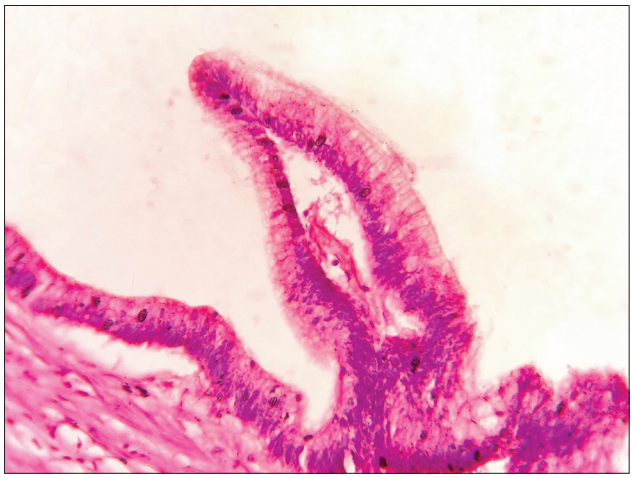

Figure 1: Cystic cavity lined by columnar epithelium showing villous architecture (H and E, ×100)

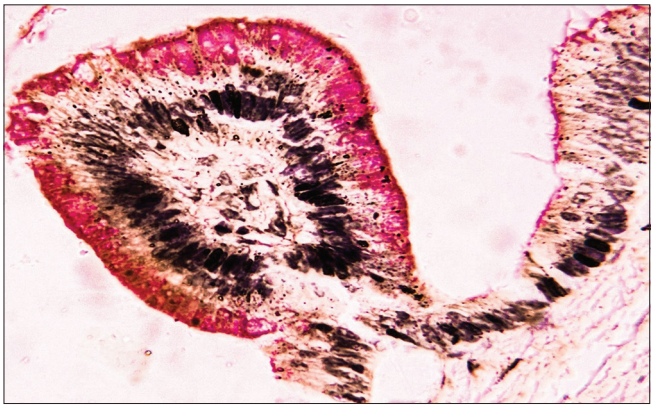

Figure 3: Tumor cells demonstrating mucicarmine positivity (diaminobenzidine, ×400)

Computed tomography (CT) scan is the most commonly used radiological investigation for preoperative diagnosis. LAMN can also present as an incidental finding during surgery. In female patients, these neoplasms can be misdiagnosed as right adnexal masses on radiological studies.[10-12] LAMN has a higher prevalence in women than men (4:1), with a mean age of diagnosis being 54 years.[13-16] Complications of LAMN include intussusception, ureteral obstruction, volvulus, small bowel obstruction, and pseudomyxoma peritonei. Elevated levels of carcinoembryonic antigen and carbohydrate antigen 19-9 act as useful markers for the diagnoses of LAMN.

Right hemicolectomy is the treatment of choice in patients with infiltration of submucosa or lymph node metastasis. In the absence of invasion or lymph node metastasis, appendicectomy with the removal of mesoappendix should be performed.[2]

On gross examination, the appendix appears to be swollen due to the accumulation of mucin with hyalinization and fibrosis of the appendicular wall. LAMN can be classified

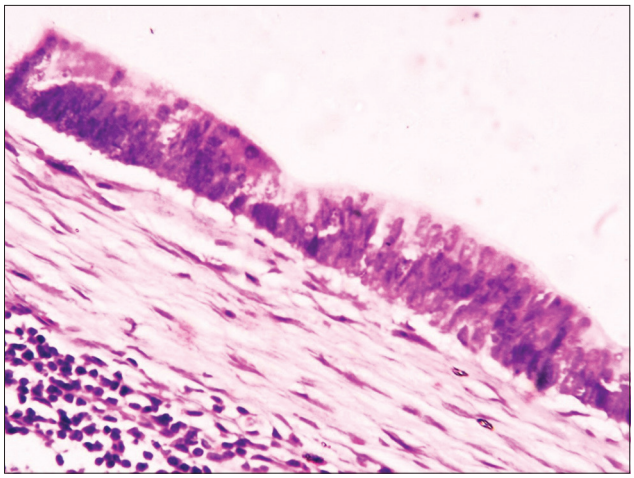

Figure 2: Foci of nuclear stratification and dysplastic changes noted in the lining epithelium (H and E, ×400)

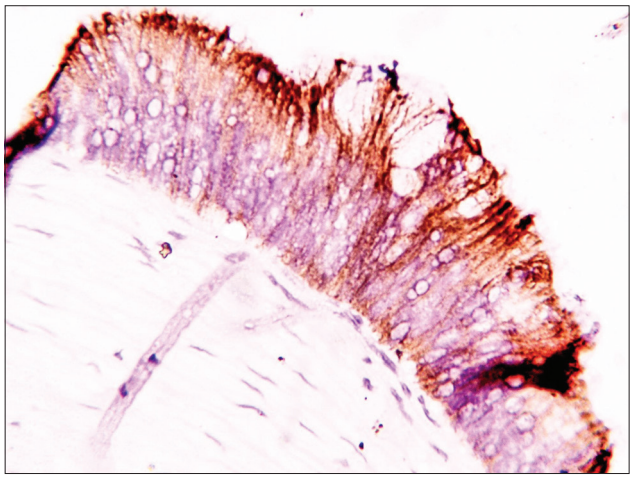

Figure 4: Tumor cells demonstrating CK 20 positivity (diaminobenzidine, ×400)

as villous or flat with atrophied lymphoid tissue and involves the appendix circumferentially.

LAMN <2 cm is rarely malignant as compared to masses larger than 6 cm, which can show malignant cells and an association with pseudomyxoma peritonei.

A comprehensive immunohistochemical panel of CK 20, MUC-2, and β-catenin is useful for the confirmation of the diagnosis.[6] In our case report, tumor cells demonstrated CK 20 positivity.

Careful pathological evaluation is required for determining the risk of malignancy and recurrence.[6]

LAMNs localized to the appendix and follow an indolent course; however, as soon as the neoplastic epithelium escapes the appendix, there is a significant increase in morbidity and mortality.

LAMN associated with pseudomyxoma peritonei can be diagnosed with the help of imaging modalities such as ultrasonography, CT scan, and magnetic resonance imaging and can be confirmed on the histopathological examination which will demonstrate epithelial cells and mucin in the peritoneum.[17] The 5-year survival rate of pseudomyxoma peritonei is 25%.[18]

Close follow-up is recommended for all LAMN patients for 5–10 years. The 5-year survival rate for localized LAMN is 95%.[19]

In the present case, right hemicolectomy was performed. The patient is under close follow-up for the past 6 months and is asymptomatic to date.

LAMN though rare can create a diagnostic dilemma due to its nonspecific clinical findings, and thus clinicians and pathologist should be aware of its varied clinical and morphological spectrum. Imaging studies and colonoscopy can be helpful to rule out other causes of acute abdomen. However, accurate diagnosis can only be made on histopathologic examination of the excised specimen. Iatrogenic rupture of the appendix must be prevented during surgery to minimize the risk of pseudomyxoma peritonei.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Vavinskaya V, Baumgartner JM, Ko A, Saenz CC, Valasek MA. Low-grade appendiceal mucinous neoplasm involving the endometrium and presenting with mucinous vaginal discharge. Case Rep Obstet Gynecol 2016;2016:1-4.
2. Mohanty AK, Anand G, Nemec J, Alnajjar A, Albarrak J. Low grade appendiceal mucinous neoplasm- rare neoplasm: A case report. J Cancer Prev Curr Res 2018;9:77-9.
3. Bartlett C, Manoharan M, Jackson A. Mucocele of the appendix – A diagnostic dilemma: A case report. J Med Case Rep 2007;1:183.
4. Soweid AM, Clarkston WK, Andrus CH, Janney CG. Diagnosis and management of appendiceal mucoceles. Dig Dis 1998;16:183-6.
5. Bradley RF, Stewart JH 4th, Russell GB, Levine EA, Geisinger KR. Pseudomyxoma peritonei of appendiceal origin: A clinicopathologic analysis of 101 patients uniformly treated at a single institution, with literature review. Am J Surg Pathol 2006;30:551-9.
6. Ramaswamy V. Pathology of mucinous appendiceal tumors and pseudomyxoma peritonei. Indian J Surg Oncol 2016;7:258-67.
7. Padmanaban V, Morano WF, Gleeson E, Aggarwal A, Mapow BL, Stein DE, *et al*. Incidentally discovered low-grade appendiceal mucinous neoplasm: A precursor to pseudomyxoma peritonei. Clin Case Rep 2016;4:1112-6.
8. Carr NJ, Sobin LH. Unusual tumors of the appendix and pseudomyxoma peritonei. Semin Diagn Pathol 1996;13:314-25.
9. Korkolis DP, Apostolaki K, Plataniotis GD, Tzorbatzoglou J, Karaitianos IG, Vassilopoulos PP. Mucocele of the appendiceal stump due to benign mucinous cystadenoma. Anticancer Res 2006;26:635-8.
10. Scaffa C, Di Bella O, Tartaglia E, Rotondi M, Lup F, Messalli EM. Surgical approach to appendiceal mucocele mimicking an adnexal complex mass: Case report. Eur J Gynaecol Oncol 2007;28:503-5.
11. Gortchev G, Tomov S, Dimitrov D, Nanev V, Betova T. Appendiceal mucocele presenting as a right adnexal mass: A case report. Obstet Gynecol Int 2010. pii: 281053.
12. Dragoumis K, Mikos T, Zafrakas M, Assimakopoulos E, Venizelos I, Demertzidis H, *et al*. Mucocele of the vermiform appendix with sonographic appearance of an adnexal mass. Gynecol Obstet Invest 2005;59:162-4.
13. Kalu E, Croucher C. Appendiceal mucocele: A rare differential diagnosis of a cystic right adnexal mass. Arch Gynecol Obstet 2005;271:86-8.
14. Madwed D, Mindelzun R, Jeffrey RB Jr., Mucocele of the appendix: Imaging findings. AJR Am J Roentgenol 1992;159:69-72.
15. Dachman AH, Lichtenstein JE, Friedman AC. Mucocele of the appendix and Pseu- domyxoma peritonei. AJR Am J Roentgenol 1985;144:923-9.
16. Jeshil RS, Vipul P. Ruptured giant mucocele of the appendix with Pseudomyxoma peritonei. Appl Radiol 2012;42:29-30.
17. Misdraji J, Young RH. Primary epithelial neoplasms and other epithelial lesions of the appendix (excluding carcinoid tumors). Semin Diagn Pathol 2004;21:120-33.
18. Akagi I, Yokoi K, Shimanuki K, Satake S, Takeda K, Shimizu T, *et al*. Giant appendiceal mucocele: Report of a case. J Nippon Med Sch 2014;81:110-3.
19. Gonzalez HH, Herard K, Mijares MC. A rare case of low-grade appendiceal mucinous neoplasm: A case report. Cureus 2019;11:e3980.