PROGRESS IN MEDICAL SCIENCES, 2021
VOL 5, NO. 2, PAGE 1 – 3
DOI: doi.org/10.47363/PMS/2021(5)162

CASE REPORT

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Misdiagnosis of Low-Grade Appendiceal Mucinous Neoplasm: A Case Report

Best Alexander*, Ananthan Mathura, Dhillon Sirjana and Tiesenga Frederick

Community First Medical Center, Dept. of Surgery, Chicago, USA

ARTICLE HISTORY

Received October 15, 2021
Accepted October 22, 2021
Published October 26, 2021

Introduction

Low-grade Appendiceal Mucinous Neoplasms (LAMNs) are rare tumors dismissed on Computed Tomography as preoperative diagnoses of acute appendicitis. Only 0.2%–0.3% of appendectomy specimens comprise mucinous neoplasms according to a population-based study by Smeenk et al [1]. AMNs function as 0.4%–1% of all gastrointestinal malignancies in the U.S., approximately decoding 1,500 new cases annually [1]. Elderly patients have an increased risk of postoperative complications. Earlier studies show higher risk of complications due to more comorbidities in the elderly population [2]. In this case, a 74-year-old African American male presented with typical findings of appendiceal colic confirmed on Computed Tomography. The patient underwent a Laparoscopic Appendectomy and the results are discussed in this case report.

Case Report

HH is a 74-year old African American male who presented for surgical consultation on an abnormal CT scan completed one-year prior. The patient also admitted to experiencing chronic, right lower quadrant pain (RLQ) for over a year, extending posteriorly with frequent constipation. Preoperative evaluation did not report any signs of fever, night sweats, significant weight gain or loss, hematemesis, diarrhea, hematochezia or melena. His past medical history was significant only for carcinoma of the prostate. His surgical history consisted of a prostatectomy. His family history was negative for any gastrointestinal cancer or polyps. His physical exam revealed a healthy African American male within normal vital sign limits. Abdominal exam revealed no significant pannus. Bowel sounds were heard across all four quadrants. His abdomen was soft and without tenderness. He showed no evidence of guarding, rebounding, or distention. The diagnosis was made to be appendiceal colic. After a review of records, seeing the patient and documenting medical records, the recommendation was made for laparoscopic appendectomy/possible open.

The patient's radiology report revealed an abutting, inferior aspect marked as a 6 x 5.5 x 12cm post-contrast 12 hounsfield units cystic tube; noted as possibly markedly distended appendix (See Figures 1 and 2). The cystic tubule wall was not thickened. There was no gross obstructive dilatation of the small bowel or colon. No free fluid or pneumoperitoneum was seen.

Figure 1: CT scan of the abdomen/pelvis in coronal view showing a markedly enlarged appendix measuring 12-cm in length in the right lower quadrant without contrast

Figure 2: CT scan of the abdomen/pelvis in sagittal view showing a markedly enlarged appendix without contrast

Contact Best Alexander ✉ bestalexander6@gmail.com Community First Medical Center, Dept. of Surgery, Chicago, USA.

The patient was admitted to the hospital for a scheduled LA procedure. The intraoperative findings were consistent with the diagnosis; however, microscopy determined the specimen to be a low-grade appendiceal mucinous neoplasm. The wall of the neoplasm had a thickness measuring up to 2mm. Mucosal surface was smooth. Serosal surface was smooth, slightly rough in some areas. The lumen had tan, thick mucinous material found upon sectioning. No perforation was seen. The patient took the surgery well and was discharged the next day. Follow up was done one week after the procedure with no complications to note.

Discussion

Low-grade Appendiceal Mucinous Neoplasms (LAMNs) arise from dysplastic mucin-producing cells in the mucosa of the appendix. Abundant mucin is produced and characteristically exhibits expansile growth with a "pushing" border, which may cause loss of the muscular components of the wall and mural fibrosis [3]. LAMN does not invade but mucin may push on the appendix and spill out with a risk of pseudomyxoma peritonei (PMP); one of the most worrisome complications.

LAMNs are capable of traversing the four layers of tissue – mucosa, submucosa, muscularis propria, and serosa. Tumor extension may spread to adjacent organs especially if cellular mucin (contains tumor cells) is determined. The 8th edition American Joint Committee on Cancer (AJCC) staging manual for staging both LAMNs current guideline used. Three different grades separate appendiceal mucinous tumors. Grade 1 tumors are considered LAMN as they resemble normal tissue [4].

This is in contrast to appendicitis, likely due to luminal obstruction and is classified as complicated or non-complicated. Causes are multifactorial, ranging from fecaliths, lymphoid hyperplasia to cancer. Once obstruction has occurred, the appendix will secrete mucus until intraluminal pressure reaches 85 cm of water [5]. This will result in edema and distention of the appendix thus increasing the risk for vascular compression and venous stasis. Most common bacterias are gram-negative, mainly Escherichia coli (present in 76 % of cases), followed by Enteroccocus (30 %), Bacteroides (24 %) and Pseudomonas (20%) [6]. These bacteria contribute in releasing toxins as a result of venous stasis. Bacteria proliferate and may extend into the appendiceal wall increasing risk of ischemia and peritonitis. The mucosa undergoes erosion and ulceration as bacteria gain momentum into the muscularis and serosa. Overtime, vascular compromise increases the risk for necrosis and perforation. Perforation generally takes 24-36 hours to occur [3].

Often, LAMN is misdiagnosed as acute appendicitis, retroperitoneal tumors in the right iliac fossa, or an adnexal mass [1]. Imaging modalities for diagnosis include ultrasound (US) and CT, with CT as the most commonly used radiographic interpretation for preoperative diagnosis. Ironically, CT has also been proven to inaccurately exclude neoplastic etiologies that may be secondary to acute appendicitis [4]. In the current case, the radiologist could not adequately distinguish between the two. The Journal of Trauma and Acute Care Surgery, conducted a study with a total of 5,224 patients who underwent appendectomy [7]. Of the total participants, 73% underwent preoperative CT [7]. Interpretation of the scan only resulted in a positive diagnosis of noninvasive tumor in 16% of the cases eligible [7].

The common abdominal CT findings include cystic dilation within the appendiceal lumen with wall calcifications and irregular appendiceal wall thickening as demonstrated in our case [2]. Grossly, specimens of LAMN include hyalinization and fibrosis of the appendiceal wall with a grossly swollen appendix secondary to mucinous accumulation. LAMNs less than two centimeters (cm) are rarely malignant and are classified as benign simple or retention mucoceles (as in our case). Masses larger than 6 cm present with a higher risk of malignant cells, a higher risk of appendiceal perforation, and development of PMP. Histological evidence of LAMN includes atypical glandular cells and epithelial cells with “pushing invasion” of malignant cells creeping into the appendiceal wall with possible diverticular formation [2]. Mucinous, colonic, and goblet cells are also often identified within LAMN. Elevated CEA, CA 19-9, and CA-125 may be detected in 56.1-67.2% of patients with LAMN [8]. These tumor markers can also be used for the surveillance of peritoneal malignancy following surgical or medical intervention [2]. There is also a 35% risk of a concurrent GI malignancy in patients with LAMN [1].

Controversy remains on the best surgical approach (laparoscopic vs open), adjuvant therapy, follow-up duration, and imaging technique. The goal of management of LAMN includes the prevention of rupture, seeding, and development of PMP. Complications of undiagnosed LAMNs include intussusception, urethral obstruction, volvulus, small bowel obstruction, rupture and PMP [8].

Surveillance of patients with LAMN incorporates radiographic imaging every six months post appendectomy for two years for adequate monitoring of tumor recurrence and complications associated with PMP [1]. Accurate pathological assessment and classification of LAMN are important to assess for malignancy risk, seeding, recurrence, and patient prognosis [3]. For patients with a high risk of disease progression, follow-up should continue for the first five years after diagnosis of LAMN. High-risk patients include those with evidence of infiltration of malignancy into submucosa or with the presence of lymph node metastasis [2]. Follow-up should continue for five to 10 years with physical exams, annual CT, and monitoring of tumor markers. The five-year survival rate for localized LAMN is 95% [8].

This case highlights the need for more research in how LAMNs are diagnosed. Due to the rarity of this particular case, LAMN should always be considered when cystic lesions of the right lower quadrant are discovered [5]. Fortunately for our patient, the LAMN was intact, confirming benign etiology, making ileocecal resection deemed excessive. Others, however, are not as lucky. With concern for possible malignancy, patients undergo right hemicolectomy as confirmed in various published case reports as well as reviews in the world literature. Follow up with an oncologist is recommended to ensure malignancy has been resected appropriately.

Conclusion

The 74 year old African American male presented with RLQ pain and was misdiagnosed on CT. Overall, further studies are needed for a more definitive method of diagnosis, treatment, and monitoring of LAMN. Diagnosis to date varies by imaging modality, the tumor markers utilized, and classification of disease. There remains a lack of standardization for post-treatment surveillance lengths and methods. This case presents the importance of developing a

high index of suspicion regarding the development of appendiceal malignancies and choosing the appropriate surgical or medical treatment; due to the nature of this patient's appendix, it was determined to undergo extension of the surgical incision rather than removal laparoscopically. This case raises awareness of an atypical physical exam. It is important for clinicians to be aware of such anatomical variants and the use of open vs. laparoscopic appendectomy as both an exploratory and therapeutic technique.

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