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Research Article

A RARE CASE OF LOW-GRADE APPENDICEAL MUCINOUS NEOPLASM: A CASE REPORT

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A B S T R A C T

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy with symptoms varying depending on the clinical manifestations. The most worrisome complication of this particular neoplasm is seeding of mucin into the adjacent peritoneum leading to pseudomyxoma peritonei (PMP). We present an unusual case of a 51-year-old female found to have LAMN status post elective right hemicolectomy.

INTRODUCTION

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy accounting for 1% of gastrointestinal neoplasms and is found in less than 0.3% of appendectomy specimens [1-2]. LAMNs are diverse and can be classified as colonic-type, mucinous adenocarcinoma, goblet cell adenocarcinoma, or neuroendocrine carcinoma [3]. Mucinous adenocarcinoma accounts for the majority of cases according to the literature [2]. This malignancy is commonly an incidental finding during operative exploration and is often diagnosed late.Gross examination of LAMN may be unremarkable or may appear as a mucin-filled, cystically dilated tissue. The appendix wall may appear thin, fibrotic, hyalinized, or calcified with a smooth or granular appearance [4]. Similar to polyps found in the colon, LAMN can be classified as villous or flat with atrophied lymphoid tissue. Neoplastic tissue growth occurs in a "pushing" invasion pattern wherein no tumor budding or single-cell invasion is noted [4]. LAMNs are associated with diverticula, herniations, dissections, and rupture [4]. The most feared complication is seeding of mucin into the adjacent peritoneum, leading to pseudomyxoma peritonei (PMP), associated with a high rate of mortality [1-2]. Seeding into the peritoneum occurs in the late stages of the disease. Our case was unique as CECT abdomen showed mucocele of appendix intussuscepting in to caecum with no evidence of periappendiceal acellular mucinous deposits, bowel obstruction or perforation.

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Case Report

A 51 year old woman was brought to emergency with complaints of right sided abdominal painsince 5 days. There were no associated complaints of vomiting/fever/diarrhea/constipation. There were no significant findings on examination of the abdomen. USG abdomen was suggestive of cystic lesion in right iliac fossa ?appendicular mucocele with ileoileal intussusception. Laboratory investigations: Hb-14.2gm%, TLC-11100/cumm, Total protein-7.2gm/dl and albumin-3.7gm/dl, S. creatinine-0.70mg/dl. CECT abdomen showed mucocele of appendix intussuscepting pinto caecum with no evidence of bowel obstruction. On lower midline exploratory laparotomy, inflamed appendix and caecum were seen with base of the appendix dilated to approx. 5cm and intussuscepting into the caecum. Resection of 10cm of terminal ileum, caecum, appendix and 10cm of proximal ascending colon with ileo-ascending end to side anastomosis was performed. The resected specimen was sent for histopathological examination which on ulceration and flattening of the lining epithelium at places showing nuclear stupid and stratification.The dysplastic lining epithelium along with pools of mucin are seen percolating within muscularis propia in focal areas suggestive of low grade appendicealmucinous neoplasm. Postoperatively patient recovered well and was discharged in stable condition on 8th postoperative day.

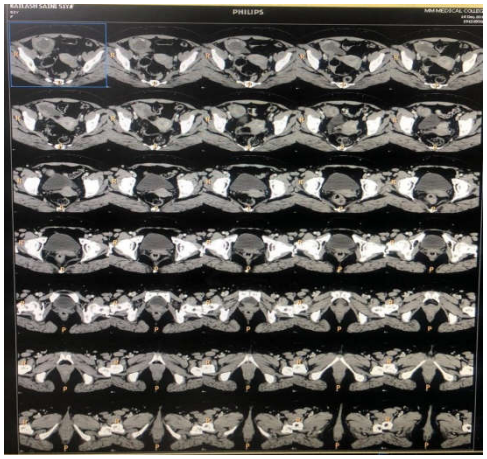

Figure 1 CECT abdomen (coronal sections)

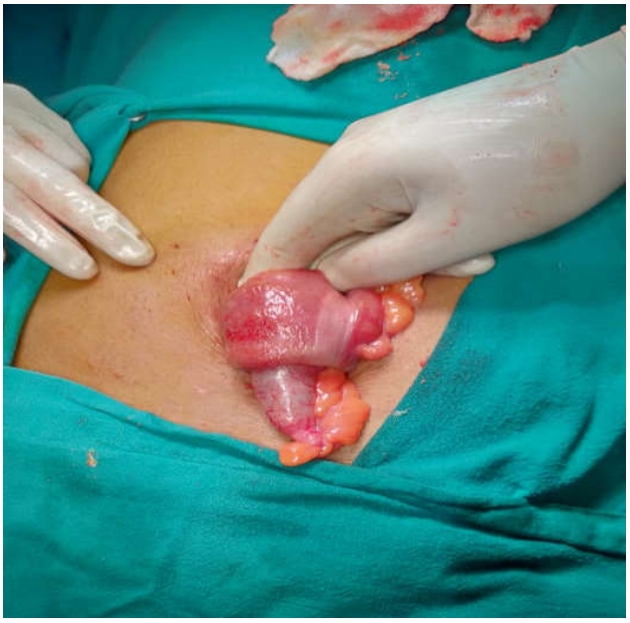

Figure 2 Intraop picture showing dilated appendix intussuscepting into the caecum

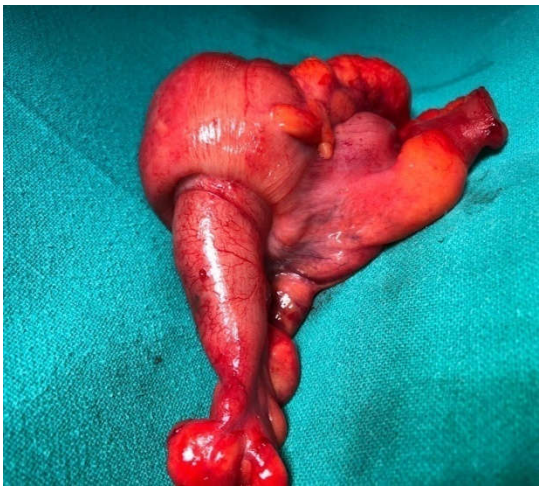

Figure 3 Picture of resected 10cm of terminal ileum, caecum, appendix and 10cm of proximal ascending colon.

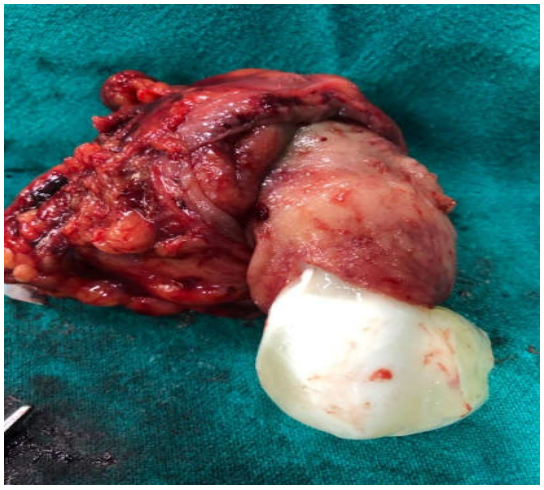

Figure 4 Picture of mucoid material seen extruding from the cutopen cystic swelling at the base of appendix.

DISCUSSION

LAMNs are rare adenomas localized in the appendix or the surrounding appendiceal mucosa wall. These neoplasms are more commonly diagnosed in men, particularly in the sixth decade of life. Patients with LAMN can present with abdominal pain, intussusception, and obstruction. However, LAMNs are often incidentally found in asymptomatic patients. Complications of LAMN include intussusception, ureteral obstruction, volvulus, small bowel obstruction (SBO), rupture, and PMP [1-2].Often, this malignancy is misdiagnosed as acute appendicitis, retroperitoneal tumors in the right iliac fossa, or an adnexal mass [2]. Imaging modalities for diagnosis include ultrasound (US) and CT, with CT as the most commonly used radiographic interpretation for preoperative diagnosis. The common abdominal CT findings include cystic dilation within the appendiceal lumen with wall calcifications and irregular appendiceal wall thickening. Grossly, specimens of LAMN include hyalinization and fibrosis of the appendiceal wall with a grossly swollen appendix secondary to mucinous accumulation [1-2,4]. LAMNs less than two centimeters (cm) are rarely malignant and are classified as benign simple or retention mucoceles. Masses larger than 6 cm present with a higher risk of malignant cells, a higher risk of appendiceal perforation, and development of PMP [2]. 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 [4]. Mucinous, colonic, and goblet cells are also often identified within LAMN [5]. Elevated CEA, Ca 19-9, and Ca-125 may be detected in 56.1-67.1% of patients with LAMN [6]. These tumor markers can also be used for the surveillance of peritoneal malignancy following surgical or medical intervention. There is also a 35% risk of a concurrent GI malignancy in patients with LAMN [5]. 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 [2]. The practice of right hemicolectomy in the absence of lymph node metastasis has been replaced, with an appendectomy only approach used for the treatment of benign appendiceal mucoceles. Upon discovery of infiltration of malignancy into submucosa or with the presence of lymph node metastasis, right hemicolectomy with or without omentectomy may be required. Our patient

underwent a laparoscopic procedure that allows magnification of the surgical field and rapid patient recovery. The risk of peritoneal seeding increases with the removal of specimens through the port site but can decrease the risk of seeding overall as reported by Fujuni *et al*. [7]. Lymph node metastasis is a rare occurrence in only 4.2% of patients but would require an aggressive treatment [8].

PMP is a complication of mucinous LAMN that can develop from peritoneal seeding in 20% of patients with a mucinous adenoma. It can be diagnosed using various modalities such as ultrasonography, CT scan, and magnetic resonance imaging depicting the presence of gelatinous mucinous nodules in the peritoneal cavity [1]. However, these imaging modalities have only been shown to identify up to 29% of adenomas prior to surgical intervention [6]. Histopathology of PMP depicts epithelial cells and mucin in the peritoneum [4]. Further advances in biomarkers and molecular genetics demonstrate CDX2, MUC-2, CK 20, β- catenin, CEA, CA 19-9, and KRAS mutations identified in hopes of improving early identification [1]. The five-year survival rate for PMP is 25% [9]. Aggressive treatments are required for PMP including appendectomy, as the appendix is the source of malignant cells in 95% of cases [1]. Aggressive strategies also include cytoreductive surgery and hyperthermic intraperitoneal chemotherapy [6].

Surveillance of patients with LAMN incorporates radiographic imaging every six months post appendectomy/right hemicolectomy for two years for adequate monitoring of tumor recurrence and complications associated with PMP [10]. Accurate pathological assessment and classification of LAMN are important to assess for malignancy risk, seeding, recurrence, and patient prognosis [1]. 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. Additional surveillance and treatment studies are needed, but until then, the treatment for LAMNs will remain inconsistent due to a lack of standardized interventions based on diagnostic criteria. Close follow-up was recommended for our patient, due to increased risk of LAMN with acellular mucin deposits outside appendix developing recurrence or PMP. 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%.l

CONCLUSIONS

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 modality to prevent recurrence, seeding, and later development of PMP.

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