Low-Grade Appendiceal Mucinous Neoplasm (LAMN) in a 46-Year-Old Male

Western Reserve Health Education
A Steward Teaching Affiliate

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Introduction

Neoplasms of the appendix are very rare with an incidence of around 1%. These malignancies are usually asymptomatic, with a majority of cases being discovered incidentally in patients only presenting with signs and symptoms of appendicitis. The Peritoneal Surface Oncology Group International (PSOGI), Chicago Consensus Working Group (CCWG), and the American Joint Committee on Cancer (AJCC) have established diagnostic guidelines, diagnostic criteria, and multidisciplinary management recommendations for appendiceal neoplasms and peritoneal surface malignancies. Among these diagnostic guidelines include criteria for low-grade appendiceal mucinous neoplasm (LAMN). This case report describes a case of LAMN to highlight the current options and complications involved in the surgical and medical management of LAMN.

Case Report

46-year-old male with a past medical history of HTN, obesity, and coronary artery disease presented to the ED with gradually worsening right lower quadrant pain for 2 days. He has a surgical history of cholecystectomy and a vasectomy, and a family medical history significant for appendiceal cancer in his maternal grandfather. Physical exam showed moderate RLQ tenderness without rebound, guarding, or rigidity. He was afebrile and without leukocytosis and non-significant lab findings.. CT A/P with contrast showed 8.0 x 5.5 cm partially calcified lesion in the lumen of the appendix with surrounding fat stranding, without free air or periappendiceal abscesses. Diagnostic colonoscopy showed inflammatory process and mild purulence from the appendiceal orifice. Diagnostic laparoscopy showed adherent mass to the right lateral wall of the RLQ. Blunt dissection revealed mucinous-like material that was extruding from the mass. The suspicion for a mucinous neoplasm of the appendix was present and the case was converted to an exploratory laparotomy. Inside, there was exudative material from the 8 cm mass in the RLQ, which was extracted and sent for pathology. As the suspicion for neoplasm was higher, right hemicolectomy was decided. The liver and the remaining colon revealed no metachronous lesions. There appeared to be no peritoneal metastases or deposits noted within the omentum, but due to its significant size and redundancy, a partial omentectomy was performed. The retroperitoneum was noted to be adherent and scarred at the point of the attachment to the appendix. An enterocolostomy was created with proper hemostasis and reapproximation of the mesentery.

Pathology report indicated low-grade appendiceal mucinous neoplasm (LAMN) involving a large portion of the appendix. There was also a periappendiceal cystic mass containing acellular mucin and calcifying mucopurulent material. Mucin was negative for neoplastic cells. Twenty-two regional lymph nodes were negative for metastatic carcinoma. No neoplastic cells were observed in the right colon segment or terminal ileum. Pathologic staging is pT (LAMN) and pN0 according to the AJCC staging system. After the surgery, further treatment with HIPEC/debulking therapy for possible peritoneal invasion was discussed with the patient.

Discussion

Clinical presentation & diagnostic criteria for an appendiceal neoplasm

Most mucinous neoplasms present with acute appendicitis-like symptoms, diagnosed incidentally postoperatively.[2] According to the AJCC 8th edition and PSOGI guidelines, LAMN is defined as a mucinous neoplasm with low-grade cytology and any of the following: loss of lamina propria, muscularis mucosa, submucosal fibrosis, diverticulum like growth into the wall, dissection of acellular mucin into wall, or mucin or neoplastic cells outside of the appendix.[3]

Surgical management

The current treatment option for a LAMN is an appendectomy with biopsy of the specimen.[1] Surgical removal is indicated to prevent overflow of cellular and acellular mucin in the peritoneum which could lead to pseudomyxoma peritonei. A right hemicolectomy should be considered to clear the tumor margin to prevent recurrence in case of involvement of the peri-appendiceal area, tumor size of 2cm or larger, high grade histology, tumor invasion through the muscularis propria, or lymph node metastases.[7]

Complications

The CCWG reports that a surgery may be curative if the patient is a low grade appendiceal neoplasm. The peritoneum is the most common site of metastasis of appendiceal neoplasms, depending on the histologic characteristics and perforation.[10] Pseudomyxoma peritonei is due to a neoplastic mucus secreting cell in the peritoneal cavity, most commonly due to a rupture of an appendiceal neoplasm or mucocele. Patients may present with increased abdominal girth, ascites, bowel obstruction.[13]

Cytoreductive surgery/HIPEC Chemotherapy

Once a complete cytoreductive surgery is achieved, chemotherapy may begin if the tumor size is less than 2.5mm.[12] Hyperthermic intraperitoneal chemotherapy (HIPEC) delivers chemotherapy diluted in saline solution and infused at high temperatures around 40ºC to an open or closed abdomen for one hour to 90 minutes.[12] Current intraperitoneal chemotherapy options include mitomycin, mitomycin plus doxorubicin, 5FU, and oxaliplatin or cisplatin.[1,10]

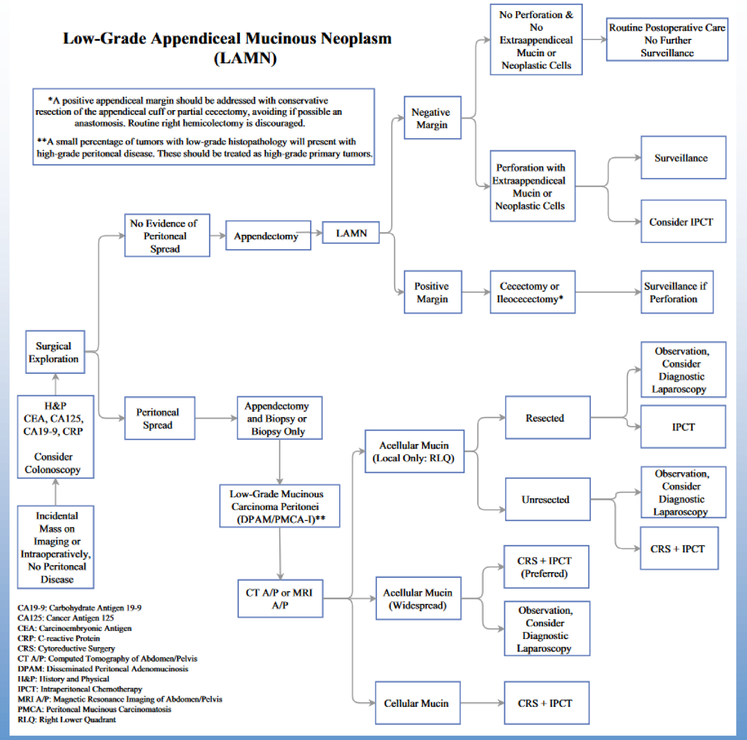


Fig. 1 Low-grade appendiceal mucinous neoplasm management pathway by The Chicago Consensus Working Group.[10] © American Cancer Society and Society of Surgical Oncology 2020.

Conclusion

Appendiceal tumors are rare, however are increasing in overall incidence in the US.[1] Current surgical and medical management include an appendectomy, right hemicolectomy, cytoreductive surgery, and HIPEC chemotherapy, with the goal of preventing any further complications. Overall, the recognition and the identification of LAMN in its early stages and distinguishing it from a benign appendicitis is critical in management and treatment of patients for overall better outcomes.

References

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