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Original Research Article

A rare case of low-grade appendiceal mucinous neoplasm with serosal involvement: a case report

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ABSTRACT

Low-grade appendiceal mucinous neoplasm (LAMN) is a malignant lesion of the appendix with an unknown etiology. It is a rare occurrence and incidental finding with only a few cases documented in literature. We present the case of a 55-year-old male with a 5-day history of diffuse abdominal pain, which then localized to the right lower quadrant (RLQ). Abdominal imaging revealed a soft tissue mass in and around the appendix, eventually leading to the diagnosis of a mucinous tumor. Exploratory laparotomy revealed a mucinous tumor of the appendix with rupture. Appendectomy was performed and the specimen along with the mucinous material was submitted for histology. Histopathological examination confirmed the diagnosis of LAMN with serosal perforation. Postoperatively the patient is stable and doing well on follow up. The patient was referred to oncology for further management. Although, LAMN is rare, there is a lack of standardization of treatment, with the need for development to prevent the latter complication of the ill-fated outcome, pseudomyxoma peritonei (PMP).

Keywords: Appendix, Low-grade appendiceal mucinous neoplasm, Case report, Pseudomyxoma peritonei

INTRODUCTION

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy of the appendix and is discovered in less than 2% of appendectomy specimens.[1] Patients can present with varying symptoms, but most commonly patients are asymptomatic. LAMN usually presents incidentally; however, some patients present with RLQ abdominal pain or a mass in the iliac fossa.[2] There are also multiple classification types for LAMNs: mucinous adenocarcinoma, neuroendocrine carcinoma, colonic-type, and goblet cell adenocarcinoma.[3] Majority of cases present as a mucinous adenocarcinoma, with gross examination demonstrating a mucin-filled appendix.[4,5] LAMN can also present as hyalinized, thin, fibrotic, or calcified wall of the appendix.[5] LAMNs are correlated with secondary conditions such as herniations, dissections, diverticula, and rupture.[5] Pseudomyxoma peritonei (PMP), is a life-threatening complication of LAMN, which presents with seeding of mucin into the peritoneum.[5] However, in our case the patient's condition was diagnosed early enough before PMP could develop. The case we present is unique due to the abundant amount of mucin identified in the intraperitoneal periappendiceal space from a serosal perforation.

METHODS

A 55-year-old Caucasian male with a past surgical history of a ventral supraumbilical hernia repair 2-1/2 weeks prior presented to the hospital with a 5-day history of diffuse abdominal pain. The pain increased in severity and localized to the RLQ for the last 2 days. The patient did not present with a history of diarrhea, fever, or chills. He had significant RLQ tenderness with some muscle guarding, but no signs of generalized peritonitis. There was a palpable mass appreciated on examination of the RLQ with deep palpation. Labs demonstrated mild elevation of the white blood cell (WBC) count at 12,000. The rest of the lab results were normal, regarding normal

enzymes, hemoglobin, and hematocrit. Computed tomography (CT) scan of the abdomen and pelvis with intravenous (IV) contrast demonstrated a mucocele or mucinous tumor localized in the appendix, without perforation or free intraperitoneal air demonstrated in Figure 1.

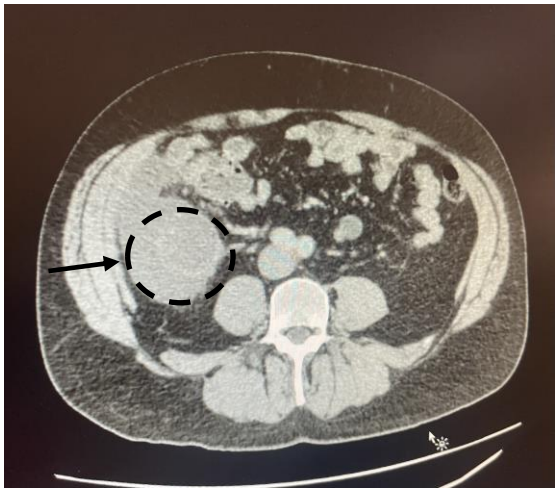

Figure 1: CT abdomen and pelvis with contrast. Axial view showing a ring enhanced mucocele or mucinous tumor localized in the appendix.

Surgery was recommended as the standard of care, the patient agreed, and an exploratory laparotomy was done. An appendectomy was performed during the exploratory laparotomy, revealing a mucinous tumor with rupture. About 100 ml of mucinous material was identified in the periappendiceal area around the cecum shown in Figure 2, which was then aspirated and submitted for further review by histology.

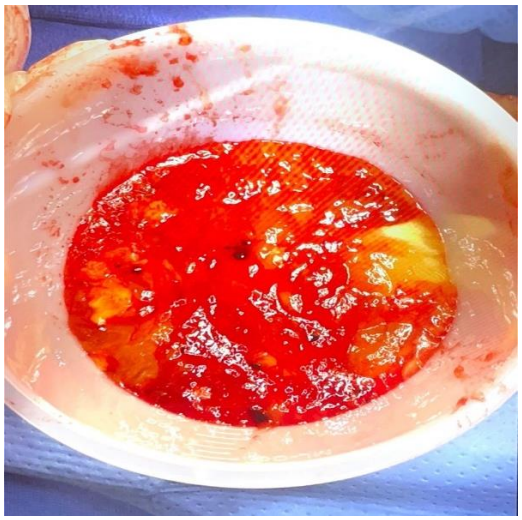

Figure 2: 100 ml of mucinous material found in the intraperitoneal periappendiceal tissue.

Identifying if the specimen was malignant vs benign could not be ascertained during the procedure, but the excision of the periappendiceal mass and appendix itself with the mucocele formation was submitted for histology of frozen section. Gross examination of the appendix demonstrated features consistent with mucocele of the appendix shown in Figure 3 and 4.

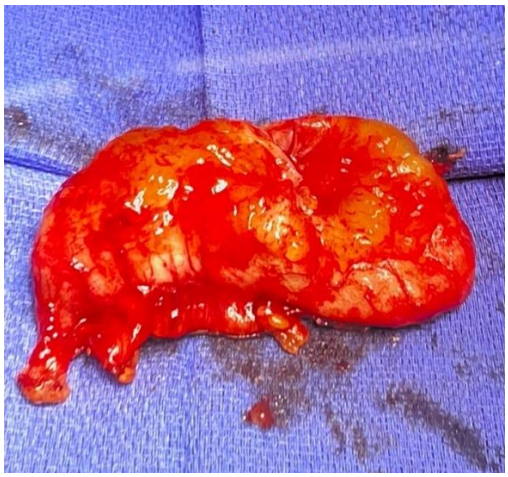

Figure 3: Gross appendiceal specimen.

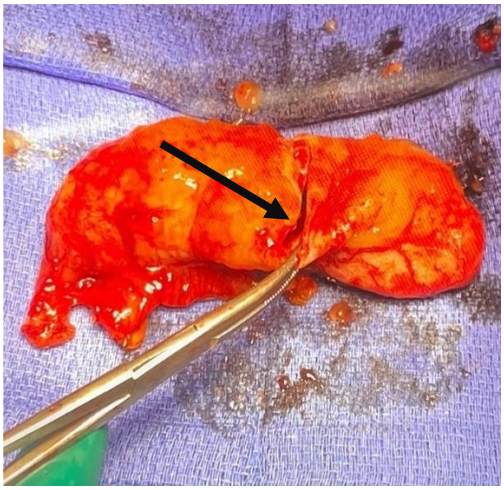

Figure 4: Gross appendiceal specimen, with an area of rupture identified.

Gross examination of the specimen reveals a vermiform appendix embedded within a pale-yellow, slightly hemorrhagic mucinous material. The appendix measures 13.8 cm in length with variation in diameter from 0.8 cm to 3 cm. The mucinous material collected measures 17.9 x 9.3 x 2.9 cm. Some clear adherent mucin measuring 8.1 x 2.7 cm covered a portion of the serosa. There was an area of rupture identified on the serosa measuring 1.9 x 1.3 cm. The defect near the tip measures 0.4 x 0.1 cm. The mesoappendix measures 5.4 x 1.2 x 0.9 cm. Specimen pathology revealed LAMN with serosal perforation by extravasated mucin into the intraperitoneal periappendiceal space. There was abundant mucin production with low-grade surface epithelial dysplasia, but no evidence of high-grade dysplasia or invasive carcinoma identified. There were neoplastic cells

identified floating in intramural dissecting mucin within the muscularis propria adjacent to the site of wall perforation, however they were not definitively identified outside of the appendix. The proximal appendiceal margin and mesoappendiceal margin were positive for acellular mucin.

Histopathology confirmed the diagnosis of LAMN with serosal involvement. Post-operatively the patient was stable and had no surgical complications. The patient was referred to oncology for further management.

DISCUSSION

The occurrence of a LAMN is rare, and often incidentally found. LAMNs occur more commonly in the sixth decade of life for men.[1] LAMN can present with obstruction or RLQ abdominal pain, but patients usually present asymptomatically.[1,2] This neoplasm is commonly misdiagnosed as an appendicitis or an adnexal mass.[2] CT findings ordinarily demonstrate cystic dilation inside the appendiceal lumen with concurrent wall calcifications and appendiceal wall thickening.[2] On gross examination, fibrosis and hyalinization are present in the walls of appendix as well as swelling of the appendix from the accumulation of mucin, which was observed in our case.[5] The larger the LAMN, generally larger than six cm, the higher occurrence of malignancy, perforation, and PMP.[5] LAMNs that are smaller than two cm are usually benign or retention mucoceles.[5] Patients that present with localized LAMN have a five-year survival rate of 95%.[5] However, the five-year survival rate for PMP is 25%.[5]

The main goal in managing LAMN is to prevent the development of PMP.[2] The most common approach for treatment of LAMN is an appendectomy; however, if the malignancy infiltrates into the submucosa or if lymph node metastasis is present, a right hemicolectomy is the treatment of choice.[3] Although our case demonstrated mucin production with low grade surface epithelial dysplasia as well as neoplastic cells within the muscularis propria, there was no evidence of high-grade dysplasia or invasive carcinoma. Lymph node metastasis is unlikely, occurring in only 4.2% of patients; however, when it occurs it requires aggressive treatment.[5] The main complication of LAMN is PMP, which develops from peritoneal seeding in patients with a mucinous adenoma.[5] Patients with PMP are treated with an appendectomy, however, hyperthermic intraperitoneal chemotherapy and cytoreductive surgery are other aggressive approaches of treatment that can be used as well.[6] Therefore, accurate diagnosis of mucocele of the appendix through physical examination and imaging is pertinent in deciding the choice of operation.[7]

The current method of surveillance of patients with LAMN is imaging every six months following an appendectomy for two years.[5] Patients that are considered high risk, including those with infiltration of the submucosa or lymph node metastasis should follow up for five years after diagnosis.[5] Our patient was referred to an oncologist and will require close follow up due to their high risk. Further interventions need to be made since the treatment and surveillance of LAMN is inconsistent.

CONCLUSION

In conclusion, the lack of standardization and surveillance of treatment methods of LAMN enforces physicians to make case by case decisions for the patient's quality of care. In addition, the various methods used to diagnose, monitor, and treat LAMN are indeterminate to prevent and identify such an occurrence. This case demonstrates the need to develop appropriate ways to measure, recognize, and treat LAMN to prevent the latter complication of PMP.

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