

Low-Grade Appendiceal Mucinous Neoplasia: A Rare Case Report

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ABSTRACT

Low-grade appendiceal mucinous neoplasms (LAMN) are rarely seen. They are incidentally detected during an appendectomy for appendicitis. Treatment is appendectomy in low-grade appendiceal mucinous neoplasms. Those having adverse features based on the appendix, lymph node metastasis, and tumour size greater than 2 cm, appendectomy with hemicolectomy is done. We present a rare case report of appendiceal neoplasm diagnosed with low-grade appendiceal mucinous neoplasm (LAMN) managed by appendectomy with right hemicolectomy.

Keywords: Appendectomy, hemicolectomy, mucinous neoplasms of appendix.

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I. INTRODUCTION

Mucinous neoplasms of the appendix comprise about 0.4–1% of all gastrointestinal malignancies. It is a rare malignancy, but an important diagnostic entity [1]-[3].

This disease is extremely easy to misdiagnose, and missed diagnosis is common. Most cases do not manifest any typical clinical symptoms, and Low-grade appendiceal mucinous neoplasms (LAMN) are often found incidentally in physical examinations or other operations [4].

We report a case of low-grade appendiceal mucinous neoplasm (LAMN) in a 70-year-old male. Appendectomy with right hemicolectomy was done with a preoperative clinical diagnosis of mucocele. Histopathology was suggestive of low-grade mucinous appendiceal neoplasm. Our case represents the importance of developing a high index of suspicion of appendiceal malignancy and the aggressive strategy adopted helped in preventing recurrence.

II. CASE SUMMARY

A 70-year-old male diabetic gentleman presented to emergency with right lower abdominal pain for 2 days which was recurrent for the last 3 months. The pain was non-radiating, with no exacerbations or remissions and no aggravating or relieving factors. On examination, he has right iliac fossa tenderness with no abdominal distension or guarding. On abdominal ultrasound (Fig. 1) a blind loop-like structure within the right iliac fossa was about 1.16cm in diameter. An appendix with a lateral hypoechoic focal mass lesion measuring about 2.3 x 2.8 cm. CT scan abdomen (Fig. 2) shows the appendix is seen arising from the inferior aspect

of the cecum with the base obstructed by a large non-enhancing cystic lesion measuring about 3.7x3.8x2 cm with no solid component and not invading the cecum. Appendix distended with fluid content with 12 mm calibre and unclear surroundings fat plane (picture suggestive of acute appendicitis with obstructed base).

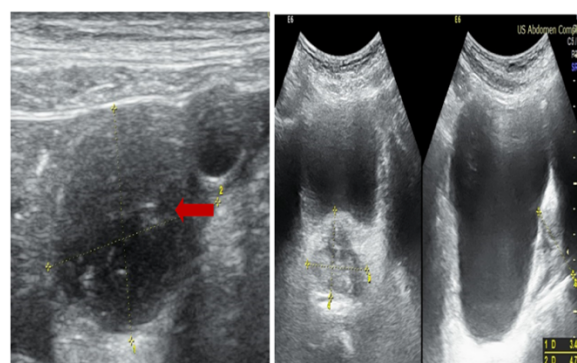


Fig. 1. Abdominal Ultrasound shows mucocele appendix in right iliac fossa.

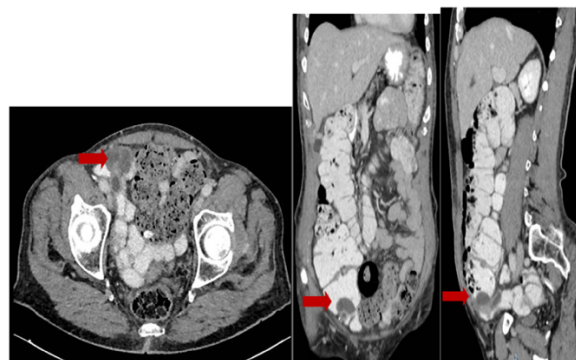


Fig. 2. CT Abdomen and Pelvis with IV and oral contrast.

On exploratory laparotomy, operative findings were cystic swelling inside the cecum at the appendiceal base with a distended appendix with no signs of appendiceal perforation. Mild amount of serous peritoneal fluid in the pelvis and right paracolic gutter (sampled for bacteriology and cytology). No lymphadenopathy, no hepatic metastasis, no additional lesions, and no intraperitoneal mucous (Fig. 3).

Limited right hemicolectomy was done and ileocolic isoperistaltic side-to-side anastomosis (Fig. 4). Biopsy of the specimen suggests low grade appendiceal mucinous neoplasia (LAMN) stage Tis with free margins.

Follow-up colonoscopy at 3 months and CT of abdomen & pelvis at 6 months revealed no residual and recurrent disease.

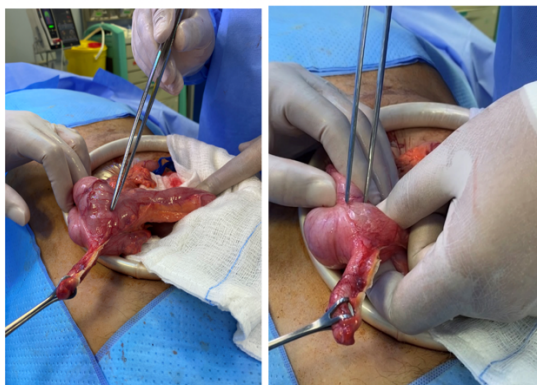


Fig. 3. Intraoperative view of appendiceal mucinous neoplasia.



Fig. 4. Limited rt hemicolectomy and ileocolic isoperistaltic side to side anastomosis.

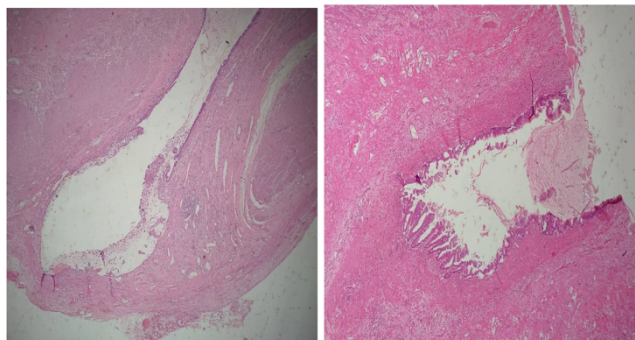


Fig. 5. LAMN composed of flat proliferation of mucinous epithelial cells resting in fibrotic submucosa with incomplete obliteration of lamina propria and disruption of muscular mucosae with intraluminal mucin. tongues of epithelium and mucin dissecting through but confined to muscularis propria. No evidence of extra appendiceal mucin or neoplastic epithelium.

III. DISCUSSION

Mucinous neoplasms make up approximately 70% of the epithelial tumours of the appendix [1], [2]. They are more common in females and present in the 6th decade. They are divided into four types: adenoma, low-grade appendiceal mucinous neoplasm (LAMN), high-grade appendiceal mucinous neoplasm (HAMN), and mucinous adenocarcinoma [5], [6].

Pain in the abdomen suggestive of appendicitis is the most common clinical presentation. Pain and abdominal distention secondary to mucin accumulation suggest advanced disease. Serious complications associated with appendiceal mucinous neoplasm include intestine obstruction, intestine perforation, rupture of mucocele, and pseudomyxoma peritonei [1]-[3]. Elevation of tumour markers CA-19.9, CEA, and CA-125 is found in 60–65% of mucinous tumours of the appendix [7]. Mucocele of the appendix on ultrasound is the first sign seen. CT scan of the abdomen is needed for confirmation and staging of disease. CT scan typically shows cystic dilation of the lumen of the appendix and irregular thickening of its wall [8].

On histopathological examination presence of mucin is diagnostic along with atypical glandular and epithelial cells. These tumours are positive for CK20 (100%), MUC5AC (86%), and DPC4 (100%) and are usually negative for CK7 (71%) [9]. The treatment plan is made according to primary tumour characteristics, grade, stage of disease, and peritoneal disease [10].

The surgical approach (open versus laparoscopic) is debated in various studies. Appendectomy is treatment in localized low-grade localized adenoma with care to avoid rupture and spillage of tumour cells and mucin [11], [12]. Five-year and 15-year survival rates for localized low-grade appendiceal mucinous neoplasm are 95% and 59%, respectively [6], [11].

Laparoscopic surgical exploration helps in better visualization with the caveat of increased chances of rupture of the appendix which may lead to pseudomyxoma peritonei [13].

Appendectomy and right hemicolectomy with lymph node dissection is done in advanced tumours like high-grade appendiceal mucinous neoplasm (HAMN), and mucinous adenocarcinoma [11]-[13]. Reference [12] proposed the criteria for right hemicolectomy in patients with high-risk features including poorly differentiated tumour, high mitotic activity, involvement of the base of the appendix, lymph node metastasis, and tumour size greater than 2 cm. There is controversy and a lack of clear guidelines for the management of cases of low-grade appendiceal mucinous neoplasm with positive surgical margins after appendectomy. Simple cecostomy versus right hemicolectomy versus observation may be the treatment options for the early stage low-grade appendiceal mucinous neoplasm with positive cut margins. Reference [14] described those patients with positive margins when treated with caecal resection had no recurrence.

Patients with pseudomyxoma peritonei are treated with appendectomy, cytoreductive surgery, and hyperthermic intraperitoneal chemotherapy [6], [11]. Patients with adverse histopathologic features high-grade poorly differentiated tumours or lymph node metastasis or perforation during surgery were treated with the chemotherapeutic agent

fluorouracil [6], [11]. Capecitabine and mitomycin C is advised for recurrent advanced stage inoperable patients with a response rate of 38% to 50% and a median overall survival of 26 months [6], [11]. Appendiceal mucinous neoplasms should be followed for the monitoring of recurrence and second malignancy [6], [11]. Follow-up comprises six monthly tumour markers e.g., CA19-9, CEA, and CA-125, and a CECT scan for up to 2 years [6], [11]. Thereafter yearly tumour markers and a CECT scan abdomen are recommended.

Reference [15], advocated an aggressive strategy that involves peritoneal and visceral resection and is also known as cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (CRS + HIPEC).

IV. CONCLUSION

Diagnosis and treatment of LAMN if timely done, excellent treatment results can be achieved. An aggressive strategy helps decrease recurrence and long-term survival in patients with LAMN.

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