

Non-resolving Appendicular Lump can be an Appendiceal Mucinous Neoplasm: A Case Report.

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Abstract

Low grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy which are mostly incidentally diagnosed after appendectomy. Unspecific ways of presentation makes it more difficult to diagnose it pre-operatively. We present here a case of 35 years old male who presented clinically as appendicular lump not resolving completely; another unusual way of its presentation.

Keywords: Low grade appendiceal mucinous neoplasm, LAMN, Pseudomyxoma peritonei, PMP, appendicular lump, acute appendicitis, HIPEC, Non-resolving appendicular lump.

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I. INTRODUCTION

Low-grade appendiceal mucinous neoplasm (LAMN) is a rare malignancy accounting for 1% of gastrointestinal neoplasm and is found in <0.3% of appendectomy specimens^[1]. Majority of the cases are incidental findings of appendectomy specimen done for acute appendicitis or appendicular lump. LAMNs are diverse, poorly understood and are classified as colonic type, mucinous adenocarcinoma, goblet cell adenocarcinoma or neuroendocrine carcinoma^[2]. 65% of malignant appendiceal tumors are of neuroendocrine origin^[3]. Although there are pathological classifications, surgical resection is the accepted treatment modality for each histological type to prevent the development of pseudomyxoma peritonei (PMP) that is associated with a high mortality rate^[4-6]. LAMNs are associated with diverticula, herniations, dissections, and rupture^[7]. Seeding into the peritoneum occurs in the late stage of the disease which causes pseudomyxoma peritonei (PMP), most feared complication of this neoplasm.

Here we will present a case of LAMN who presented initially as non-resolving appendicular lump; yet another way of AMNs atypical presentation.

II. PRESENTATION OF THE CASE

Case is a 35 years old male who presented to Surgery OPD for pain in right lower abdomen (on & off) for last 02 months and burning sensation of epigastrium for last 02 weeks. There was no other associated symptoms except for nausea and loss of appetite. He had history of similar pain 02 months ago, for which he was admitted and treated in the line of appendicular lump.

On examination, vitals were unremarkable.

Per-abdominal examination findings were, soft with no distension and a vague lump of approximately 05 x 04 cm in right iliac fossa, which was tender on palpation with local guarding and rebound tenderness.

Blood investigations were unremarkable.

Transabdominal ultrasonography was suggestive of appendicular abscess with perforation & a left renal calculus.

Chest/Abdominal X-Rays were negative for pneumoperitoneum.

He was admitted in male surgery ward and Ochsner-scherren regime initiated. Gradually he showed improvement in terms of settling pain and decreased lump size after 10 days of therapy. Vague lump later turned into discrete lump of approximately 02 x 02 cm which was palpable per abdominally and non-tender.

Repeat transabdominal ultrasonography showed 08 mm appendicular diameter.

Open appendectomy under spinal anaesthesia was planned and executed. Intraoperative findings were a firm lump of 02 x 02 cm in middle one-third of appendix and appendicular tip adhesion with omentum. Other findings were unremarkable. A 02 cm margin of mesoappendix was also excised along with appendix. Intraoperative findings were suggestive of appendicular fecolith.

Histopathological examination of the specimen revealed Low-grade Appendiceal Mucinous Neoplasm (LAMN),
pTNM: pT_{4a}N₀M₀(Stage IIB, AJCC 8th Edition).

Histological grade was G1 and well differentiated. No lympho-vascular and peri-neuron invasion. Proximal margin was free of tumor with a distance of 2.7 cm from LAMN and mesenteric margin was also free of tumor with a distance of 1.8 cm from LAMN. Additional finding was a diverticula at distal appendix with greatest dimension of 0.5 cm.

Looking at his intraoperative and histopathological findings, we considered that simple appendectomy alone would suffice his treatment.

Postoperatively he recovered well and was discharged on 04th post-operative day.

He came for follow up after 03 weeks of discharge. He was doing well without any signs of complications till then. He was suggested for regular follow ups.

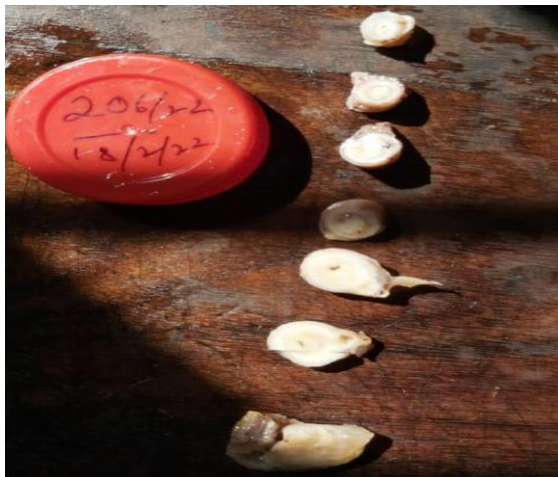


Figure 1.

Cut sections of appendix specimen.



Figure 2.

Appendiceal mucosa lined by multi-layered dysplastic epithelium which are arranged in indulating pattern, few in villiform pattern and some with flattened epithelium. The submucosa shows marked fibrosis. Acellular mucin is seen in appendiceal lumen (blue arrow), mucosa and also seen dissecting into the appendiceal wall.

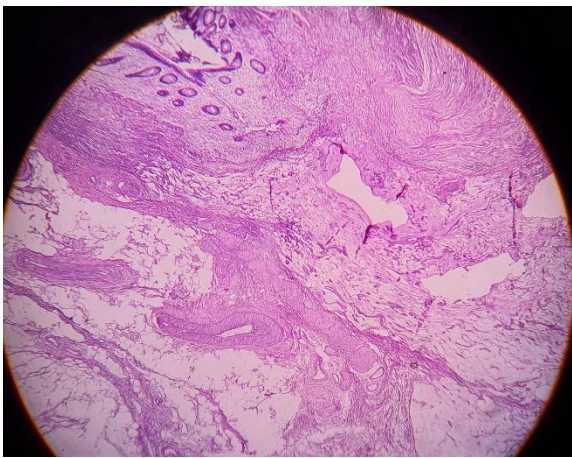


Figure 3.

Acellular mucin dissecting into appendiceal wall reaching upto the serosa with neovascularization and inflammatory infiltrates surrounding it.

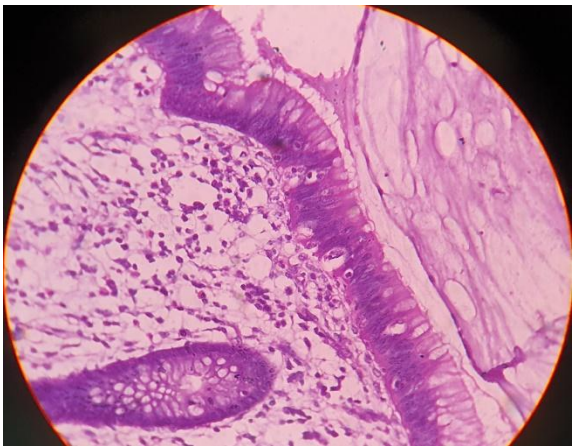


Figure 4.

Dysplastic tumor cells are tall columnar with basally located hyperchromatic nuclei with inconspicuous nucleoli and have abundant apical mucin filled clear cytoplasm. Submucosa shows marked fibrosis.

III. DISCUSSION

LAMNs are rare gastrointestinal malignancy which are often encountered clinically with misdiagnosis of acute appendicitis. Right iliac fossa pain is almost always a suspicion for acute appendicitis in a clinician's mind and in addition, unspecific way of LAMNs presentation makes it more difficult to diagnose pre-operatively. Complications of LAMNs are intussusception, ureteral obstruction, volvulus, intestinal obstruction, rupture, and PMP^[1].

Elevated tumor markers viz. CEA, Ca 19-9, and Ca-125 may be detected in 56.1-67.1% of patients with LAMN^[8].

Ultrasonography, CT scan and MRI have only been shown to identify up to 29% of adenomas prior to surgical intervention^[9].

LAMNs less than two centimeters (cm) are rarely malignant and are classified as benign simple or retention mucocoeles. Masses larger than 6 cm present with a higher risk of malignant cells, a higher risk of appendiceal perforation, and development of PMP^[10].

Surgical resection is the accepted treatment modality for each histological type to prevent the development of pseudomyxomaperitonei (PMP) but with discrepancies in extent of resection. A simple appendectomy should suffice in case of intact mucocoele away from appendicular base with no lymph node involvement but in cases of adjacent organ seeding, CRS with HIPEC is recommended^[11]. There are also controversies regarding better method of resection (laparoscopic versus laparotomy), adjuvant chemotherapy, choice of investigation and follow up duration. Care must always be taken to avoid seeding of malignant cells into peritoneum to avoid pseudomyxomaperitonei (PMP).

We feel that laparoscopic is a better method than open appendectomy in the sense that the prior has advantage of better intraabdominal view than the latter to look for any other suspicious findings.

We also feel that taking peritoneal lavage fluid for analysis would definitely add to advantage and help decide post-operative requirement of hyperthermic intraperitoneal chemotherapy (HIPEC).

IV. CONCLUSION

Due to rare nature of AMNs we still lack definite treatment protocol. Pre-operative diagnosis is difficult due to its diverse presentation and overwhelming cases of appendicitis in emergency room. First thing that comes in a clinician's mind of right iliac fossa pain is "Acute Appendicitis". AMNs should always be in differential diagnosis in cases with atypical presentations like in our case; an appendicular lump which refuses to resolve completely.

INFORMED WRITTEN CONSENT:

Informed written consent has been obtained from the patient and ready for reference whenever sought.

CONSENT TO PARTICIPATE

Consent has been obtained from the patient for participation.

CONSENT FOR PUBLICATION

Patient has been informed & consent taken for publication.

AVAILABILITY OF DATA & MATERIAL

Data & materials are available in the MRD section of hospital which is available for reference with permissions from the officer in charge of concerned section.

CODE AVAILABILITY

Not applicable.

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COMPETING INTEREST

Author(s) declares that they have no competing interests.

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