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APPENDICEAL MUCINOUS NEOPLASM : INDOLENT BUT NOT INNOCENT – A CASE REPORT

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ABSTRACT
Appendiceal mucinous neoplasms are a rare group of epithelial tumours accounting for <1% of gastrointestinal neoplasms and typically affecting middle aged to older adults with no stronger gender predilection. It is characterized by appendix lumen dilatation, caused due to the accumulation of mucus with varying degrees of cytological atypia. It is commonly observed as incidental finding during surgical interventions for acute appendicitis or other abdominal procedures. This is a case report of a 30-year-old-female who presented with recurrent right-sided abdominal pain. The pain was associated with nausea. Computed tomography findings showed a cystic lesion in the right iliac fossa, suggestive of appendiceal mucinous neoplasm, which was managed by laparoscopic appendectomy. Attributed to generalized clinical presentation, the diagnosis of appendiceal mucinous neoplasm can be challenging. An accurate preoperative identification with the help of radiological modalities is helpful in assessment of the severity and adjacent tissue structures. This case highlights the importance of early diagnosis and laparoscopic management in better health outcomes as accurate diagnosis can help avoid any intraoperative rupture.

KEYWORDS: Mucinous neoplasm. mucin producing tumors, mucocele, computed tomography, LAMN, HAMN, Appendicular neoplasm, appendix

1. INTRODUCTION

Case presentation

A 30-year-old female presented to the outpatient department of our tertiary care hospital located at South India with right-sided abdominal pain from last two weeks. The pain was localized, dull and intermittent, non-radiating and associated with occasional nausea. No other symptoms such as vomiting, fever, change in bowel habits or weight loss were observed. There were no addictions and any significant medical history noted. Gynecological history was regular with last menstrual period 15 days back. Abdominal examination showed mild tenderness in the RIF with elicited rebound tenderness without palpable masses. Her laboratory findings were found to be within normal limits. Ectopic pregnancy was ruled out by a beta-HCG test of the urine. Computed tomography (CT) scan of the abdominal and pelvic region showed a well-defined, fluid-filled tubular structure in the appendiceal region. The walls of the lesion exhibited calcification specks highly suggestive of mucocele. The ovaries and adnexa appeared normal, and no regional lymphadenopathy was detected [Fig 1].

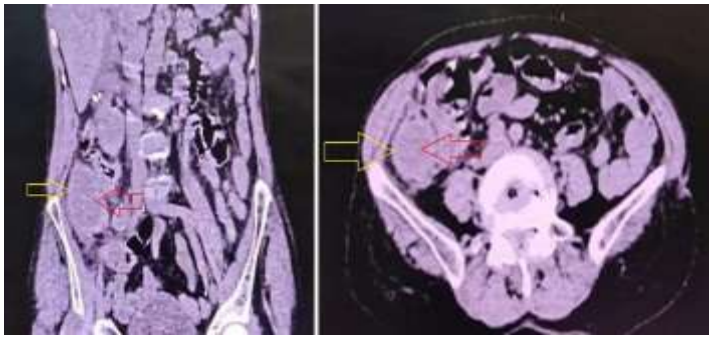


Fig 1 Computed tomography image showing yellow arrow: Cystic dilatation of the Appendix consistent with appendiceal mucinous neoplasm; Red arrow: Intraluminal mucin within the dilated appendix

In view of the reported association between appendiceal mucinous lesions and synchronous colorectal neoplasia, a preoperative screening colonoscopy was performed, which demonstrated normal colonic mucosa without any intraluminal lesion or suspicious pathology

The patient was managed by laparoscopic appendectomy. Intraoperative findings showed a dilated, pale appendiceal mass measuring approximately 7.0 x 3.0 cm, which was identified in the RIF [Fig 2].

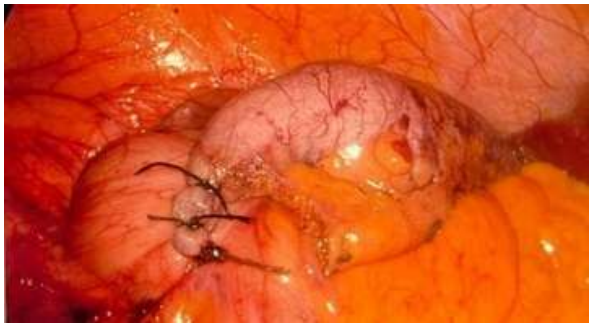


Fig 2 Intraoperative view of appendiceal mucinous neoplasm before resection. Laparoscopic image demonstrating a tense, cystically dilated appendix arising from the cecum with intact serosa, surrounding omental adhesions and mesoappendix were also noted.

The appendix walls appeared thinned but intact, with no evidence of perforation or mucinous spillage. The appendix was resected and retrieved using an entrapment bag with care to avoid any rupture and spillage [Fig 3].

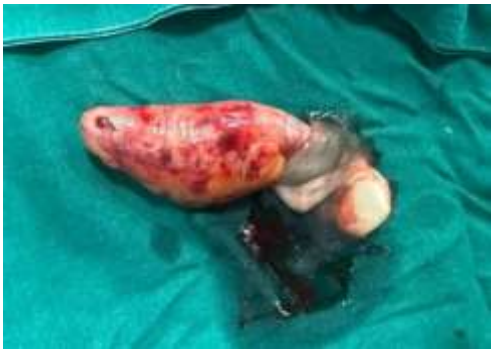
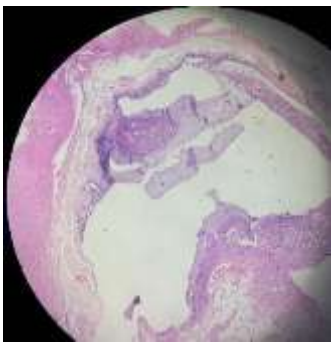


Fig 3 Gross specimen showing resected appendix as markedly dilated with cystic enlargement and smooth congested serosa.

Gross examination of the laparoscopic appendectomy specimen revealed an appendix measuring 6 * 3 * 2.5 cm with a congested external surface and attached periappendiceal fat. The cut surface showed a dilated lumen filled with clear gelatinous fluid and a wall thickness of 0.3 cm.

Microscopic examination demonstrated a low-grade appendiceal mucinous neoplasm (LAMN) diffusely involving the appendix. The tumor measured 6 * 3 cm in greatest dimension. Histological analysis identified acellular mucin invading the visceral peritoneum (serosa) and present in the periappendiceal fat. There was no evidence of lymphovascular or perineural invasion, and surgical margins were negative for non-invasive tumor. The case was staged according to the AJCC 9th Edition as pT4a (tumor invades through the visceral peritoneum, including acellular mucin involving the serosa of the appendix or mesoappendix) [Fig 4].



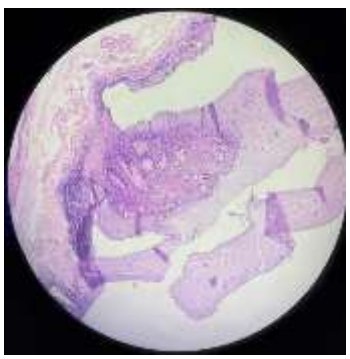


Fig 4 [H&E X 40 and H&E X 100] Hematoxylin and eosin-stained slide showing: Appendiceal lumen with low grade neoplastic epithelium with large amount of pools of mucin

The patient had an uneventful postoperative course and was discharged on the third postoperative day. There were no associated complications noted at first follow-up at fifteen days.

DISCUSSION

Appendiceal mucinous neoplasms are commonly observed in individuals >50 years with no gender predominance. Though, appendiceal mucoceles have been associated with lower risk of malignancy [2]. An incidence of 0.2 to 0.7% has been reported in total appendectomy samples. Abdominal pain is commonly noted as the presenting clinical symptom for appendiceal mucinous neoplasm, with an incidental diagnosis in most of the patients [3]. Though, there are some uncommon clinical presentations of adjacent organ involvement [4]. It is observed as a palpable mass in ~50% of the patients [5]. Common radiological modalities such as ultrasound and computed tomography can be helpful in diagnosis of severity of the condition and study the adherence to the adjacent layers, which can provide useful information in management and prevention of post-operative complications such as pseudomyxoma peritonei. Though, in few instances, surgical exploration is needed [6,7]. A case series reporting incidentally diagnosed appendiceal mucinous neoplasm, it to be associated right-sided abdominal pain. It is recommended to maintain a high suspicion index in cases with right-sided lower quadrant abdominal pain, as also noted in this patient [7].

The clinical picture often mimics acute appendicitis, appendicular plastron, or, in female patients, ovarian pathology. While abdominal ultrasound may show an "onion skin" sign (concentric layers of echogenicity), a CT scan remains the gold standard for diagnosis. The presence of mural calcification on CT scanning is seen in up to 50% of cases, is a specific indicator distinguishing mucocele from simple appendicitis. The choice of surgical approach is controversial. Though, laparotomy can minimize rupture risk, laparoscopic surgery is increasingly favored for its benefits in recovery and cosmetic outcomes, provided the specimen is handled carefully [8]. The primary surgical goal is to prevent iatrogenic rupture, which can lead to pseudomyxoma peritonei, a condition requiring aggressive management, including peritonectomy and intraperitoneal chemotherapy. In this case, the laparoscopic approach provided excellent visualization and allowed for safe resection without spillage, confirming that laparoscopy is a safe option for selected patients with unruptured mucinous neoplasm.

Management and Surveillance of T4a Low-Grade Appendiceal Mucinous Neoplasms

According to current National Comprehensive Cancer Network (NCCN) guidelines, low-grade appendiceal mucinous neoplasms (LAMN) classified as T4a are defined by tumor expansion through the muscularis propria that breaches the visceral peritoneum (serosa) of the appendix or mesoappendix. This staging requires the presence of either acellular mucin or neoplastic mucinous epithelium on the serosal surface, without adjacent organ invasion (T4b) or distant peritoneal spread (M1a/M1b).

The primary surgical objective in managing T4a LAMN is achieving a complete gross resection (R0) while meticulously avoiding iatrogenic rupture. Appendectomy serves as the standard definitive treatment; however, a partial cecectomy may be indicated to secure negative margins if the tumor involves the appendiceal base. Crucially, current guidelines strongly advise against routine formal right hemicolectomy for T4a LAMN. Extensive colonic resection does not confer survival benefits nor reduce the risk of recurrence in LAMN, and unnecessarily subjects the patient to increased operative morbidity. Right hemicolectomy is strictly reserved for instances where an R0 margin cannot be achieved via cecectomy (such as massive cecal involvement), when synchronous right-sided colon malignancies are identified, or if the final pathology unexpectedly reveals an invasive mucinous adenocarcinoma. Furthermore, contemporary consensus favors conservative management over re-resection for patients with a purely T4a LAMN who have microscopically positive margins for neoplastic epithelium, as positive margins in this context do not reliably predict recurrence.

Despite the indolent nature of LAMN, the serosal breach inherent to T4a lesions stratifies these patients into a high-risk category for developing pseudomyxoma peritonei (PMP), mandating rigorous long-term surveillance. Optimal follow-up imaging incorporates magnetic resonance imaging (MRI) of the abdomen and pelvis, specifically utilizing diffusion-weighted imaging (DWI), which offers superior sensitivity for detecting subtle mucinous implants. Computed tomography (CT) with intravenous contrast remains an acceptable and widely utilized alternative, whereas PET/CT is discouraged due to its low sensitivity for mucinous disease. Standard surveillance protocols necessitate cross-sectional imaging combined with a tumor marker panel (CEA, CA 19-9, and CA-125) every six months for the first five years, transitioning to annual assessments up to ten years post-resection. Additionally, a baseline screening colonoscopy is recommended for all patients to exclude synchronous colorectal malignancies.

CONCLUSION

Appendiceal mucinous neoplasm are rare in young adults. This case report highlights the need to add appendiceal mucinous neoplasm in non-specific clinical presentations even in younger adults, as delayed or misdiagnosis might result in adverse outcomes such as Pseudomyxoma peritonei.

CONFLICTS OF INTEREST

None.

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