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Case Report

A rare case of mucinous adenocarcinoma of appendix presenting as acute small obstruction

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ABSTRACT

Mucinous appendiceal neoplasms are rare, occurring in <0.3% of appendectomies, often presenting incidentally. A 61-year-old female with periumbilical pain and vomiting was diagnosed with small bowel obstruction and mucinous adenocarcinoma of the appendix. Imaging showed bowel thickening and obstruction. Surgery revealed a fibrous band, serosal tears, and appendicitis with appendicolith. Histopathology confirmed adenocarcinoma with perineural invasion. Despite intervention, the patient succumbed to septicemia and multiple organ dysfunction syndrome (MODS). Appendiceal tumors are often incidental, requiring computed tomography (CT) for diagnosis. Right hemicolectomy is standard treatment. Early recognition and surgical management are key to improving prognosis in these rare cases.

Keywords: Mucinous neoplasms, Open appendicectomy

INTRODUCTION

Mucinous appendiceal neoplasms are rare and not well understood, accounting for fewer than 0.3% of all appendectomy cases.[1,2] These tumors form a varied and complex group of epithelial neoplasms that often lead to cystic enlargement of the appendix due to the buildup of a gelatinous substance, a condition known as a mucocele. First identified by Rokitansky in 1842, mucoceles are frequently discovered incidentally during clinical or radiological evaluations in patients without symptoms.

It is crucial to note that the cause of a mucocele can range from a benign retention cyst to a malignant adenocarcinoma. If a mucocele ruptures, it can lead to a serious complication known as pseudomyxoma peritonei (PMP).

CASE REPORT

A 69-year-old male came to svp hospital with chief complaint of lower abdominal pain since 7 days associated with non-bilious non projectile vomiting containing food particles since 3/4 days, constipation on 06 June 2024.

On per abdomen examination, abdomen was distended, and non-tender.

On digital rectal examination (DRE), the anal tone was normal, with no active bleeding observed. Stool stain was present. Mild roominess present.

All routine pre-operative blood investigations were within normal limits except low potassium.

X-ray abdomen showed multiple air fluid levels.

Ultrasonography (USG) whole abdomen indicated fluid and content filled ileal and jejunal loops noted with maximum diameter of ileal loop in pelvis measures 28 mm. Fluid filled right sided colon and caecum noted, diameter of caecum measures 30 mm. Sluggish peristalsis noted in the small bowel.

Minimal interbowel free fluid noted.

Contrast enhanced computed tomography (CECT) indicated multiple air and contrast filled dilated small bowel loops predominantly jejunal and ileal loops are noted in periumbilical region, bilateral lumbar, bilateral iliac fossae and hypogastric region with smooth transition point as described above; possibility of subacute small bowel obstruction. There is evidence of blind ending tubular fluid density lesion with peripherally enhancing wall, having maximum diameter of approximately 15 mm in right iliac fossa arising from the base of caecum; possibility of mucocele of appendix likely. Mild ascites was observed.

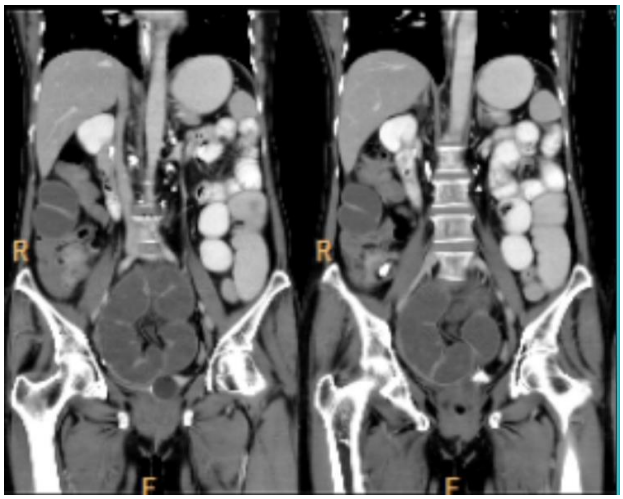

Figure 1: Radiological image.

Management

Patient presented with the aforementioned complaints and was managed conservatively. Features of obstruction present on admission were henceforth relieved and further radiological investigations were done. Patient was operated on 07 June 2024 for exploratory laparotomy + open appendicectomy. Postoperatively patient developed features of intestinal obstruction also supported by radiological findings and patient was managed conservatively for the same and discharged on 22 June 2024.

HPE

Sections show the tumor to be composed of neoplastic glands lined by pleomorphic hyperchromatic cells. Extracellular mucin with floating tumor cells are seen. Tumor cells are invading muscularis propria and adipose tissue. Occasional signet ring cells are seen. Perineural invasion is present. Lymphocytic response is moderate.

Diagnosis

Acute small bowel obstruction due to mucinous adenocarcinoma of appendix.

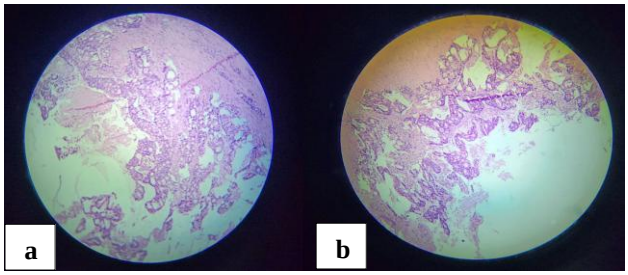

Figure 2 (a and b): Histological image.

Follow up

On follow up patient had no complaints, and in view of low Eastern Cooperative Oncology Group (ECOG) score patient was not operated for right hemicolectomy.

DISCUSSION

Appendiceal neoplasms are among the least common tumors of the gastrointestinal tract, identified in approximately 0.7% to 1.4% of appendectomy specimens. Within this group, mucinous adenocarcinomas of the appendix are particularly rare, with an incidence of only 0.01% to 0.08%.[3-5] Primary appendiceal carcinomas are classified into two main categories: adenocarcinomas (originating from epithelial cells) and neuroendocrine tumors (of neuroendocrine origin, previously referred to as "carcinoids"). Adenocarcinomas are further divided into mucinous and nonmucinous (colonic) subtypes, while neuroendocrine tumors can be classified into signet ring cell, malignant, and goblet cell variants.

Fewer than one-third of mucinous appendiceal adenocarcinomas present as acute appendicitis. More commonly, they are incidentally discovered through imaging as a cystic mass in the right lower abdomen or in patients with abdominal distension due to pseudomyxoma peritonei.[6] CT imaging is particularly effective in identifying appendiceal tumors, typically revealing features such as cystic enlargement of the appendix or a localized soft-tissue mass.[7] While an appendiceal diameter exceeding 15 mm is not definitively diagnostic, it should raise strong suspicion of malignancy. Although ultrasound may assist in evaluating abdominal masses, CT provides superior anatomical detail, particularly in differentiating between a mucocele and adjacent cecal structures, and in detecting mural calcifications.[8]

The standard treatment for appendiceal adenocarcinoma is a right hemicolectomy, either performed initially or as a follow-up after the diagnosis is confirmed histologically. When an appendiceal mucocele is suspected, debate exists regarding whether an open or laparoscopic appendectomy is preferable.[9] Some studies, such as that by Gonzales et al, have reported peritoneal dissemination following laparoscopic resection, suggesting open surgery may be safer.[10] Rupture of a mucocele poses a significant risk by allowing epithelial cells to spread within the peritoneal

cavity, potentially leading to pseudomyxoma peritonei—a serious and often life-threatening complication.

CONCLUSION

Mucinous adenocarcinoma of the appendix presenting as obstruction is a rare yet clinically significant condition that warrants prompt recognition and intervention. primary appendiceal tumors are histologically diverse and have an insidious onset and few specific clinical manifestations. In the majority of cases, these tumors are discovered after appendectomy during pathological exam of the resected tissue. Treatment may include appendectomy (simple or radical) and right hemicolectomy depending on factors such as histological type, tumor size and lymph node/organ involvement. This case emphasizes the importance of thorough clinical evaluation and imaging, like CT scans, for diagnosing acute abdominal symptoms. Surgical intervention, often exploratory laparotomy, is crucial for both diagnosis and treatment. The challenging prognosis of mucinous adenocarcinoma of the appendix, highlighted here, underscores the impact of comorbidities and complications like sepsis. Clinicians must be aware of diverse presentations, including obstruction, to ensure early recognition and multidisciplinary care, optimizing outcomes and reducing morbidity and mortality.

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