

Case Report

Mucinous adenocarcinoma masquerading as acute appendicitis

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ABSTRACT

Appendiceal neoplasms are a rare entity with an incidence of unexpected findings in surgical specimen ranging as low as 0.7-1.7%. Carcinoid tumor of the appendix is the most common among all tumors while adenocarcinomas are rare and occur in 0.08-0.1% of appendectomies. It usually involves the base of the appendix and is treated by right hemicolectomy. As most carcinomas present as acute appendicitis, high degree of suspicion is required for diagnosis of neoplasm. Histopathology plays a vital role as it is the only way to confirm diagnosis. We present to you a case of 45-year-old male who came to the ER with pain and was diagnosed with acute appendicitis. Emergency appendectomy was done and on opening the specimen a mucocoele was seen which was positive for malignancy in histopathology.

Keywords: Acute appendicitis, Mucinous adenocarcinoma, Mucocoele

INTRODUCTION

Only less than 2% of appendectomy specimens prove positive for malignancy.¹ Primary neoplasms of appendix comprise of epithelial, mesenchymal tumors and lymphomas. Mucinous neoplasms are a complex group of epithelial neoplasms. Mucocoele of appendix can be defined as the accumulation of gelatinous material in appendix. It is imperative to recognize the underlying cause of mucocoele to rule out malignant adenocarcinoma as its rupture can lead to pseudomyxoma peritonei, which is a deadly complication.²

CASE REPORT

A 45-year-old male presented to OPD with complaints of pain in right iliac fossa. He had no similar complaints in the past nor did he have any other significant history. On examination, tenderness was present in the right iliac fossa. Patient was advised a USG which revealed an inflamed appendix of approximately 14 cm. The patient was then eventually planned for an interval appendicectomy. Patient presented with recurrent pain to

the emergency. Patient was worked up again and he underwent emergency appendectomy with drain insertion for the same. An inflamed appendix of around 15 cm was removed, which was thick, edematous and had a hard consistency.

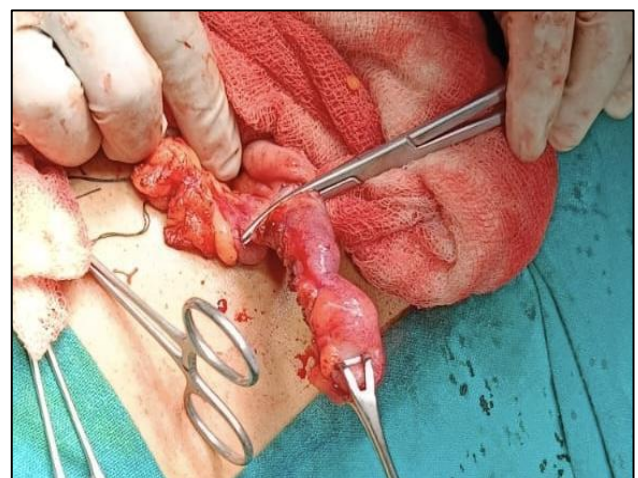


Figure 1: Intra-operative specimen of appendix.



Figure 2: Cut open specimen of appendix showing mucocoele.

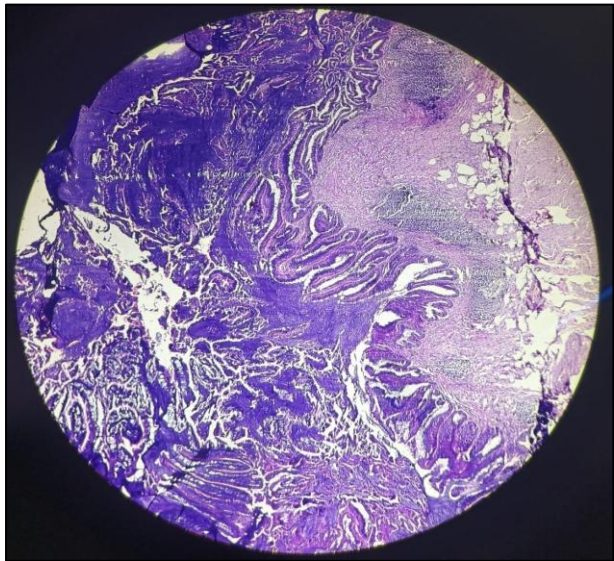


Figure 3: Malignant glands.

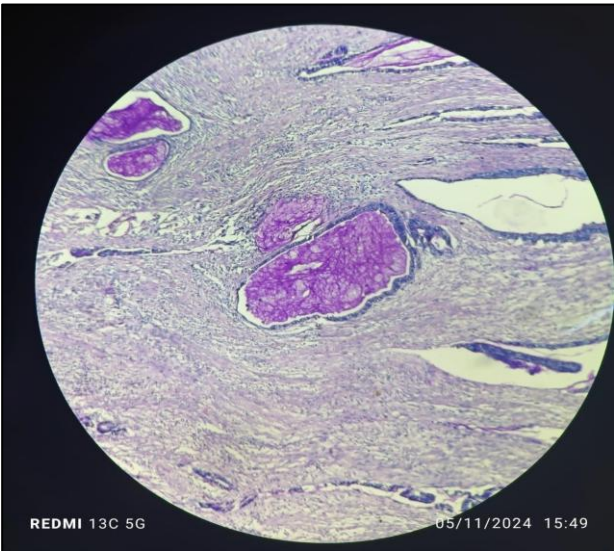


Figure 4: Lining of malignant gland and mucin content.

The appendicular specimen was opened and a mucocoele measuring 3×2 cm was found. The specimen sent for histopathological examination. It revealed that the specimen was positive for mucinous adenocarcinoma. Further confirmation about the involvement of base was obtained and results showed that the base of appendix was also involved. Patient was called on follow-up and was counselled about his condition and the need for a right hemicolectomy and which he underwent the surgery. The hemicolectomy specimen which was sent was free of the tumor. The intraoperative and postoperative periods for both the surgeries were uneventful. The patient recovered well and was healthy on follow up.

Table 1: Management of mucinous adenocarcinoma of appendix.

Grade	Rupture status	Recommended management
Low-grade	Non-ruptured	Appendectomy en bloc with cyst kept intact
High-grade	Non-ruptured	Right hemicolectomy with goal of ≥12 lymph nodes
Low-grade	Microscopic rupture	Laparoscopy for evaluation and resection of residual disease
High-grade	Microscopic rupture	Laparotomy with removal of residual disease, right hemicolectomy, omentectomy, right lower quadrant peritonectomy, *bilateral oophorectomy, and HIPEC
Any grade	Gross peritoneal disease	Imaging to evaluate eligibility for complete cytoreduction followed by complete cytoreduction and HIPEC

DISCUSSION

Malignant tumors of appendix comprise of mucinous epithelial neoplasms, neuroendocrine (carcinoid) tumors, goblet or composite carcinoid, lymphomas, adenocarcinoma, and mesenchymal sarcomas. Around 65% of appendiceal tumors are neuroendocrine tumors, making them the most common, while only around 20% are adenocarcinomas.^{3,4} 0.2-0.3% of appendectomy specimens reveal mucinous neoplasms.⁴ Patients present either with vague symptoms like weight loss, abdominal pain, anemia, infertility, new onset umbilical or inguinal hernia or can have acute appendicitis.⁵⁻⁷

While USG is the first line investigation for its high specificity for acute appendicitis, contrast enhanced CT is the investigation of choice as mucocoeles. These are frequently picked up on CT and mostly mimic acute appendicitis. But one major drawback is that it cannot differentiate between benign and malignant causes. If appendix has a diameter of more than 15 mm, soft tissue

mass, wall thickening or irregularity, it may raise a suspicion of mucinous neoplasm.⁸

MRI are useful in detecting incidental appendiceal adenomas, rupture and extra appendiceal mucin. Mucinous neoplasms are seen as hyperintense tubular distension of appendix in T2 weighted images.⁹ In our case, a basic USG was done for diagnosis of acute appendicitis. It was only after the specimen was cut open, a mucocoele like structure was identified and further histopathological examination revealed that the mucocoele had neoplastic origin.

Treatment is based on rupture of tumor and grade of the tumor. Non ruptured, low-grade tumors require appendectomy and high-grade tumors necessitate a right hemicolectomy. Diagnostic laparoscopy is carried out for microscopic ruptures, while gross ruptures have to be treated with laparotomy chemotherapy and radiotherapy are mainstay for unresectable tumors.¹⁰

CONCLUSION

Acute appendicitis is a common diagnosis but mucocoele in appendix is a rare occurrence. Neoplasm in appendicular mucocoele is even more rare. Early detection is very difficult as mucocoele are rarely identified on USG. It is important to identify these lesions as early as possible to prevent rupture and dissemination of the disease to reduce morbidity and mortality.

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