

# Mucinous Carcinoma Of The Appendix- Unveiled- A Case Report

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## Abstract:

Appendicular carcinoma though extremely rare in incidence may commonly present as acute appendicitis or perforation. Here we present a case of high-grade mucinous carcinoma of the appendix that presented as acute appendicitis with appendicular perforation in this 56yr old female. Initial imaging, USG, done in an outside hospital revealed tip of the appendix to be perforated along with appendicitis. Subsequently, patient was taken up for emergency open appendectomy. Intra-operatively, mucocele of the appendix was noted with an inflamed appendix. Post-operative HPE was followed up, which revealed high grade mucinous carcinoma of the appendix. This case report is a review of previous literature the hopes to emphasize the need for proper follow-up of patients post-operatively to watch out for the incidence of such rare conditions.

**Key Word:** Acute appendicitis, appendicular perforation, mucinous carcinoma of the appendix, carcinoma of the appendix, mucocele

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## I. Introduction

Malignant tumors of the appendix though rare, encompass a vast variety of subtypes including mucinous epithelial neoplasms, neuroendocrine (carcinoid) tumors, goblet or composite carcinoid, etc.

Appendiceal mucinous neoplasms are an incidental finding in roughly 0.2%–0.3% of appendectomy specimens. Appendiceal mucinous neoplasms include a wide range of diseases with varying malignant potential. Early stage AMNs are usually incidentally diagnosed during an appendectomy for suspicion of appendicitis. Advanced stage disease usually presents with abdominal distension, probably related to the accumulation of mucin in the peritoneal space. It is important to detect the disease to promptly refer the patient and plan for further management.

We present a 56-year-old female that presented with appendicular perforation that was then diagnosed to have mucinous carcinoma of the appendix.

## II. Case Report

A 56yr old female, Mrs. S was referred to the Emergency department of our hospital with abdominal pain localized to the right iliac fossa and lower abdomen for 2days; with associated vomiting, nausea and fever for 1day. She is a known Diabetic on regular OHAs and a known case of Bronchial Asthma on irregular treatment. On arrival, the patient was hemodynamically stable. Clinically on arrival, patient had tachycardia and was febrile and had Right iliac fossa tenderness at McBurney's point. A bedside Mantrel's score revealed a score of 6/10. Routine investigations were done and showed Hb of 13.6g/dl (Normal); WBC counts of 8,800; with normal RFT and LFT. An outside USG was taken and the report suggested 1cm thickened and fluid-filled appendix with severe surrounding fat stranding, tip of appendix has a rent with minimal collection surrounding it; suggestive of acute appendicitis with perforation at the tip of the appendix with focal peritonitis.

In view of the clinical signs and persistent tachycardia despite adequate analgesia, patient was taken up for Emergency Open appendectomy on the day of admission.

Intra-operatively, mucocele of the appendix was noted with perforated tip of the appendix. Appendix was inflamed and adherent to the ileum and caecum. Base of the appendix was healthy. We proceeded with total appendectomy.

Post-operatively, patient was closely monitored, was discharged and asked to follow-up with HPE reports.

HPE revealed High grade appendiceal Mucinous Carcinoma with dissection of mucous into peri-appendiceal surface with serosal reaction, base was free from tumor. Patient was then referred to Department of Surgical Oncology, Tamil Nadu Government Multi Super Specialty Hospital, Chennai.

1 month post-operatively, CECT abdomen and pelvis was taken along with tumor markers (CA125, CEA, CA19-9). CECT revealed post-op changes in RIF with no residual lesions; tumor markers were within normal limits (CEA-3.03; CA-125-16.8; CA-19/9-47.92)

After review of reports and discussion with the tumor board, patient was planned to continue on conservative management. Patient was serially followed up in our General Surgery OPD as well Surgical Oncology OPD.

Repeat CECT abdomen and pelvis and tumor markers done 3 months post-operatively revealed no residual disease as well. 7 months post-operatively, PET-CT was done for the patient which revealed no metabolically active peritoneal lesions or lesions elsewhere with a probable benign subcapsular lesion in segments VI, VII and II of the liver. Tumor markers were also repeated 7 months post-operatively, found to be within normal limits.

At present, patient has nil complaints and is on regular follow up.



### **III. Discussion**

Malignant tumors of the appendix include the following subtypes- (AJCC 8<sup>th</sup> edition) adenocarcinoma in-situ, Low grade appendiceal mucinous neoplasm (LAMN), mucinous carcinoma, colonic adenocarcinoma, goblet cell carcinoma, carcinoid tumors, signet cell carcinoma, mixed tumors, NOS, lymphoma [22]. Appendiceal mucinous neoplasms are randomly found in 0.2%–0.3% of post-appendectomy specimens [2]. The treatment of AMN is controversial in terms of extent of surgery and the role of chemotherapy, including choosing between early postoperative intraperitoneal chemotherapy (EPIC) and hyperthermic intraperitoneal chemotherapy (HIPEC).

Epidemiology-Appendiceal mucinous neoplasms account for 0.4%–1% of overall gastrointestinal malignancies in the U.S., roughly translating to 1-2 new cases per 1,00,000 people annually [1], [6-9],[22]. Women account for the majority of the cases; roughly 50%–55% of the appendiceal tumor population [5], [10].

Clinical manifestations-Patients with appendiceal tumors can present with nonspecific clinical manifestations thus delaying diagnosis [11]. An acute appendicitis- like presentation with right lower abdominal pain secondary to collection and distention of the appendix by mucin is the most common clinical presentation in early-stages disease contributing to about 32% of all cases [3,12]. Appendicitis or perforation of the appendix is another mode of presentation, specifically seen if the tumor obstructs the lumen of the appendix and is seen in less than 20% of all cases. Advanced stages of the disease presents either with increasing abdominal girth/distension due to the accumulation of mucinous ascites within the peritoneum or other clinical presentations like chronic abdominal pain, loss of weight, anemia, and new-onset umbilical or inguinal hernias [11], [13].

Classification-Well-recognized types of carcinoma appendix include mucinous adenocarcinoma, and ordinary colonic-type adenocarcinomas (non-mucinous), which are almost identical to those seen in the colorectal region [14]. A third category, called appendiceal crypt cell adenocarcinoma (or adenocarcinoma

ex- goblet- cell- carcinoid) is a less known type characterized that sometimes may have a mucinous component. Most non- mucinous carcinomas are colonic type adenocarcinoma.

Another system of classification, called the Ronnett's classification model [4] was subsequently created and revised and simplify classify mucinous carcinomas into low- and high- grade carcinoma, wherein any mucinous epithelium extending beyond the muscularis mucosa is considered as indisputable evidence of an invasive appendiceal malignancy [3], [12]. This classification is adapted and standardized into AJCC 8<sup>th</sup> edition classification, wherein 2 other entities are also included- serrated polyp with/without dysplasia and mucinous carcinomas with/without signet cell cytology [22].

Treatment of localized AMN-On review of most of the published literature,the general suggestion for treatment of localized AMN isto proceed with a simple appendectomy [15]. Right hemicolectomy is suggested for cases where clearance of the tumor margin in case of involvement post-appendectomy is needed, or it should be considered for tumors involving the peri- appendiceal area, tumor size of 2 cm or larger, high-grade histology, or tumor that invades through the muscularis propria.

Surgical management of low- grade AMN with peritoneal mucin spillage is a controversialtopic. Published literature suggests a significant prognostic difference between acellular and cellular mucin [3], [16]. Accordingly, the use of cytoreductive surgery (CRS) for relatively early lesions with localized cellular mucin spillage is advocated based on population- based [16] and pathology- based, series, showing high likelihood of progression to extensive intra- abdominal disease if treated by appendectomy or right hemicolectomy alone. For acellular mucin, regular follow up is advised due to the potential risk for peritoneal dissemination in the future. Adjuvant chemotherapy is not advised for low- grade well- differentiated mucinous tumors and should only be thought of in specific situations where the tumor shows invasive features such as lymphovascular invasion or lymph node involvement or has mixed- type histology.

Treatment of peritoneal AMN- In the late 90s, Sugarbaker [17] introduced the idea of a one- stage aggressive cytoreductive debulking surgery (CRS) via several macroscopic peritonectomies, followed by chemotherapy in the form of intraoperative HIPEC infusion. This is still considered to be the treatment of choice till date [18]. The aim of HIPEC is to deliver a regionally high intraperitoneal dosage of heated chemotherapy with as minimal systemic effects as possible. In the setting of peritoneal AMN, the most commonly used drugs are mitomycin C at 10–12.5 mg/m<sup>2</sup>, [19], oxaliplatin at 460 mg/m<sup>2</sup>, cisplatin, and 5- FU, as single agents or in various combinations [19]. Combining CRS and HIPEC [20] has showed immensely positive results in the 5- year survival rates of about 86% for DPAM (disseminated peritoneal adenomucinosis) a variant of high-grade AMN, and 50% for the more aggressive version of AMN called PMCA (Peritoneal mucinous carcinomatosis). The 5- year survival rate however drops to 20% with cases where there is incomplete cytoreduction[19], [20].

Role of systemic chemotherapy: Retrospective review of reports confirm that histopathology, pre- operative systemic chemotherapy, degree of CRS, and use of HIPEC are significant but independent predictors of the overall survival outcomes. Adequate CRS and HIPEC treatment without preoperative systemic chemotherapy remains the gold-standard in the management of peritoneal metastasis of low- grade AMN. [21]

#### **IV. Conclusion**

Although mucinous carcinoma of the appendix is a rare phenomenon, it may present as trivial as acute appendicitis or as severe as appendicular perforation. Only post-operative follow-up will reveal to us the occurrence of such cases. A review of the literature has reported very few cases presenting with appendicular perforation initially and a majority of cases presenting with either subacute or acute appendicitis worldwide. This study hopes to encourage prompt identification of patients at an early stage and referring them for further management.

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