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#### MUCINOUS ADENOCARCINOMA OF THE APPENDIX: A CASE REPORT AND LITERATURE REVIEW

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## MUCINOUS ADENOCARCINOMA OF THE APPENDIX: A CASE REPORT AND LITERATURE REVIEW

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### SUMMARY

**Primary adenocarcinoma of the appendix is a rare disease as compared with cancer of the colon. It is common in patients in the middle age. Mucinous adenocarcinoma is one of the histological types seen. The usual presentation of patients is acute appendicitis or peri-appendicular abscess. Diagnosis is often made after the surgical specimen has been sent for histopathological review. It is important to rule out synchronous and metachronous tumour during surgery.**

**We present a case of a 42 year old female whom an appendectomy had been attempted, the procedure was aborted and she developed an enterocutaneous fistula prior to presentation in our hospital. At exploratory laparotomy, the fistulous tract was excised and an appendectomy done. Histological report was that of mucinous adenocarcinoma. There was no synchronous or metachronous tumour in the patient and she did well after surgery.**

**Mucinous adenocarcinoma of the appendix is a diagnosis made on histopathological assessment and is a very rare tumour. To the best knowledge of the authors, there is no known reported case of the disease in Nigeria. Thus, physicians and surgeons should entertain a diagnosis of the disease in middle aged patients such that exploration of bowel is performed with thorough surgical surveillance for synchronous and metachronous tumours.**

### INTRODUCTION

Primary adenocarcinoma of the appendix when compared with cancer of the colon is a rare disease that constitute < 0.5% of gastrointestinal neoplasm (1,2). As at 2002, less than 250 cases had been reported in literature (3). It is often seen in patients above the age of 50 years and presents as acute appendicitis or peri-appendicular abscess (4). The histological types seen includes mucinous adenocarcinoma, cystadenocarcinoma, adenocarcinoids, adenosquamous, signet ring type adenocarcinoma and colonic type non-mucin producing adenocarcinoma of the appendix (2,5,6). Mucinous adenocarcinoma is a difficult disease to diagnose pre-operatively and most patients are not identified till the disease is advanced (7). Niteck *et al* observed that in only 32% of cases studied were the diagnosis made intra-operatively (1). Diagnosis is often made after the histopathological examination of the surgical specimen (8). Rarely, frozen sections

have also been used in the diagnosis (6).

Malignant tumours of the appendix resemble colonic adenocarcinoma (9). Hence, there should be surveillance for synchronous or metachronous tumours of the gastro-intestinal and extra intestinal tract during surgery (1). The extent of the disease as at the time of surgery is a more important predictor of survival than the histological type (10). It has also been reported that mucinous adenocarcinoma and absence of carcinomatosis are good prognostic factors (8).

### CASE REPORT

A.M, a 42 year old woman referred from a peripheral hospital to our institution with leakage of feaculent fluid from abdominal operation site. She had earlier presented with recurrent pain in the right lower abdomen of one month duration, colicky, non-radiating with no aggravating or relieving factor. No accompanying abdominal swelling and no change in bowel habit. No associated fever, nausea, vomiting,

anorexia or weight loss.

A diagnosis of appendicitis was made and appendectomy was attempted at the referring hospital. However, intra-operative finding was an appendiceal mass with extensive adhesions involving terminal ileum, caecum and the ascending colon. Procedure was aborted, wound closed and she was referred to our institution for further management.

At presentation, she was febrile, dehydrated, and tachycardic with continuous leakage of feaculent material from the wound site but still passes feces per anus. An indurated mass was felt at the right iliac fossa with marked tenderness. A diagnosis of entero-cutaneous fistula was made.

*Initial management include:* rehydration, control of sepsis (antibiotics), enteral nutritional support (high calorie & protein diet), skin care with barrier cream and controlled collection of effluent using cellophane bag. The conservative management was done for four weeks but the feaculent leakage persisted despite moving her bowel adequately, sepsis controlled and adequate nutritional support. Abdomino-pelvic ultrasound scan revealed a mass in the appendiceal region.

She had exploratory laparotomy with findings of appendiceal mass adhered to the anterior abdominal wall. Fistulous tract from the anterior abdominal wall was traced to the ascending colon. Small intestine and other parts of the colon appeared grossly normal. No mesenteric lymphadenopathy. The liver, stomach, spleen and pelvic organs were grossly normal. She had appendectomy with excision of the fistulous tract. Post-operatively, patient improved considerably. Histology of the appendiceal mass was mucinous adenocarcinoma. She is being followed up and counselled for right hemicolectomy in outpatient clinic.

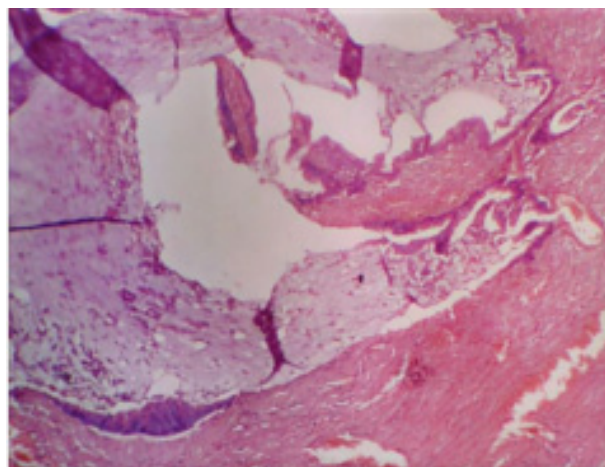
**Figure 1**

*Gross specimen of the appendix received*



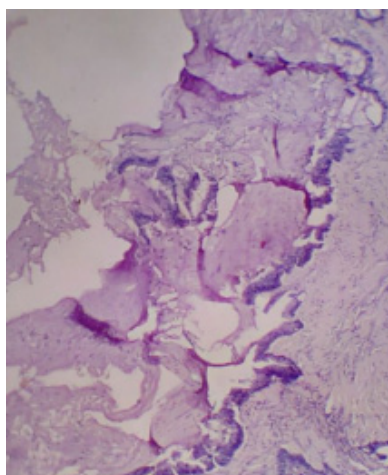
**Figure 2**

*H&E section of Mucinous Adenocarcinoma of the Appendix with malignant glands and mucin infiltrating the muscular layer*



**Figure 3**

*PAS section of Mucinous Adenocarcinoma of the appendix*



## DISCUSSION

Mucinous adenocarcinoma of the appendix is a rare disease (1). Data on it in Nigeria and Africa is scarce. It is a disease of the gastrointestinal tract that usually presents in patients above the middle age, although, primary adenocarcinoma of the vermiform appendix had been reported in a 36 year old woman (3). Thus, it is not surprising to have recorded it in this 42 year old female patient on whom an attempt at appendectomy had been made before referral to our hospital. The findings of a bulbous and firm appendix (4 cm in diameter)

without involvement of its base and caecum, absence of lymphadenopathy and metastatic lesions in other part of the bowel and the clinical state of the patient informed the decision to perform an appendectomy. There is controversy as to the superiority of right hemicolectomy over appendectomy (7). Some authors are of the opinion that a right hemicolectomy is the appropriate surgical treatment in patients diagnosed for mucinous adenocarcinoma of the appendix (9). The preferred surgical procedure and outcome are poorly understood (1).

The intra-operative diagnosis was that of carcinoid tumour, which buttress the fact that mucinous adenocarcinoma is not suspected most times; both before and during operations (1,7,10). However, the use of frozen section would have allowed the diagnosis of mucinous adenocarcinoma to be made intra-operatively. Furthermore, since female patients present with gynaecological signs and symptoms (8), it is advisable to exclude metachronous tumours such as metastatic adenocarcinoma from the ovary during surgery (1). This was painstakingly done at surgery.

The histological type of mucinous adenocarcinoma and absence of carcinomatosis could be the reason why the patient did well after surgery (7,10). She had right hemicolectomy done two months later and a proper long time follow up programme has been instituted for her. She has remained symptoms free till date.

In conclusion, mucinous adenocarcinoma of the appendix is a diagnosis made on histopathological assessment and is a very rare tumour. To the best knowledge of the authors, there is no known reported case of the disease in Nigeria. Thus, physicians and surgeons should entertain a diagnosis of the disease in middle aged patients such that exploration of the bowel should be performed with thorough surgical surveillance for synchronous and metachronous tumours. In addition, intra-operative diagnosis can be made using frozen sections.

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