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Mucinous Adenocarinoma of the Appendix
A retrospective note:

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Introduction
Carcinoma of the appendix is very rare, and almost always a retrospective diagnosis which requires revision and reconsideration of the original approach. In this case report, we discuss an elderly female who presented with mucinous adenocarcinoma of the appendix, and the diagnostic and curative challenges the case team faced in the management.

Case Presentation
A 75-year-old female was referred to the surgical unit, with intractable, dull abdominal pain& distension for 1 month. The pain originated as an intermittent, non-radiating, colicky pain which escalates at night, but was well-tolerated. She also complained of recent onset subjective loss of weight, and loss of appetite. She did not have any urinary symptoms or fever. Examination findings revealed a distended, tense abdomen with guarding. There was clinical evidence of free fluid. Rest of the examination was normal.

The basic laboratory work-up including the white blood cell count, platelets, urine full report and culture, C-reactive protein, serum amylase, liver and renal function tests was normal. The capillary blood sugar level and the subsequent fasting blood sugar level were also normal. X ray abdomen did not reveal any distended bowel segments or air fluid interfaces. Ultrasound scan of the abdomen revealed gross ascites and a suspicious appendiceal mass.

During appendisectomy, she was found to have gross ascites of non-mucinous nature macroscopically, and an appendiceal mass incorporating the right ovary and fallopian tube. There was no clinical or radiological evidence of any regional or distant metastasis. Following appendisectomy and right sided salphingo-oophorectomy, histological analysis was done and the mass proved to be a well differentiated mucinous adenocarcinoma of the appendix.

The agreed upon definitive option of management is total surgical resection with a right hemi-colectomy, & if needed, surgical cytoreduction, following which the patient should be referred to an oncology unit for administration of intra peritoneal chemotherapy. She was waiting surgical debulking at the time of writing this case report (November 2018), and remains healthy up to date.

Discussion
Carcinoma of appendix is one of the rarest types of malignancies in the gastro intestinal tract, accounting for less than 0.5%, and although infrequent, presents in an array of histologically distinct sub types1. The diagnosis is almost always retrospective in nature, making it quite a challenge to diagnose and treat prospectively.
Epidemiologically, mucinous adenocarcinoma accounts for less than 0.5%of all gastro intestinal cancers, and does not demonstrate any gender predilection, nor any substantial risk factors, and has a mean age of onset of 60 years1. Its histological classification is broad, as either low grade or high grade, which is extremely important in the management and prognosis of the disease[1,2,3.]

In its commonest clinical presentation, appendicular adenocarcinoma mimics acute appendicitis, which seldom leads to a pre-op CT, and is almost always retrospectively diagnosed either intra-operatively or later as a histological diagnosis. This requires us to 'go back' and re-investigate to stage the disease and plan for a modus of definitive management.

Low Grade Appendiceal Mucinous Neoplasm (LAMN) is a slowly progressive indolent disease, where the 5 years survival rate for patients even with a high disease burden is around 81.3%1,2,3,4. This commonly presents when the invasive tumor is ruptured, and causes a clinical syndrome of abdominal distension with mucinous ascites, known as pesudomyxoma peritonei. However, local invasion of surrounding organs can be observed in LAMN, which would require debulking maneuvers such as right hemi-colectomy or en mass resection of ovaries.

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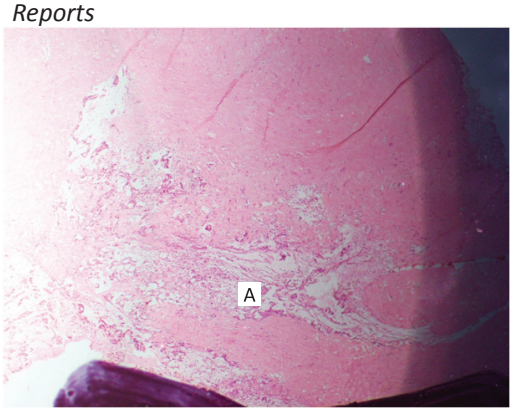

Figure 1. A – Appendicealacinar demonstrating atypical dysplastic cells

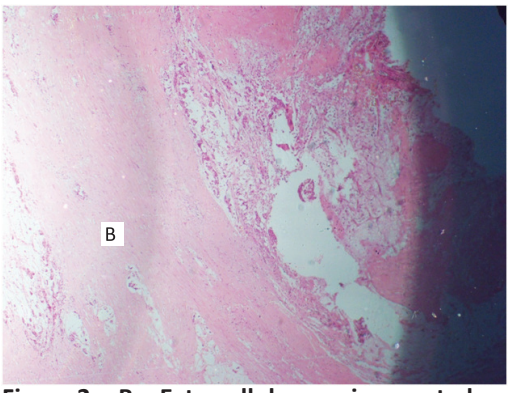

Figure 2 - B – Extracellular mucin secreted by carcinomatous cells

High grade appendiceal mucinous neoplasms carries a high index of mortality,with extensive invasion of regional and distant tissue, making a wide excision challenging. The Peritoneal Carcinomatosis Index (PCI) described by Jacquet and Sugarbaker is a useful clinical tool to assess the index of disease burden, aided by radiological investigations such as pre/post-operative contrast enhanced CTor MRI4. The disease is then staged according to the American Joint Commission on Cancer (AJCC) criteria on the basis of tumour size, extent of involvement of lymphatic, and extent of metastasis1,3,4,6.

Management of adenocarcinoma of the appendixis often consists of surgical resection combined with adjuvant chemotherapy. The specificities depend on the histological sub type, and the staging of the disease. For low grade adenocarcinoma which hasn't ruptured, appendisectomy with cystectomy without spilling mucin intraperitoneally is recommended7,8. If the mucin cyst is ruptured but there is no evidence of mucosal invasion, a right hemi-colectomy is preferred7.
For high grade carcinoma with evidence of distant metastasis, the gold standard recommendation is surgical cytoreduction and intraperitoneal chemothearapy.7,8 Surgical cytoreduction refers to complete gross resection of invaded tissue, which routinely consists of omentectomy, right quadrant peritonectomy and bilateral salphingo-oopherectomy. For widespread disease with extensive lympho vascular metastasis, such as the liver or portahepatis, intra peritoneal chemotherapy in the form of Hypothermic Intra-Peritoneal Chemot herapy(HIPEC)should be administered7,8. A median 5 year survival rate of 75%-80% for low grade tumours, and 45%-65% for high grade tumours has been demonstrated.3,7,8

In conclusion, although almost impossible to diagnose pre-operatively, appendiceal carcinoma should be investigated and treated aggressively, without hesitating to re-visit and reconsider the original approach.

References

1. McCusker M E, Coté T R, Clegg L X, Sobin L H. Primary malignant neoplasms of the appendix: a population-based study from the surveillance, epidemiology and end-results program, 1973–1998. Cancer. 2002;94(12):3307–3312.
2. Alakus H, Babicky M L, Ghosh P. et al. Genome-wide mutational landscape of mucinous carcinomatosisperitonei of appendiceal origin. Genome Med. 2014;6(5):43.
3. Benedix F Reimer A Gastinger I Mroczkowski P Lippert H Kube R; Study Group Colon/Rectum Carcinoma Primary Tumor. Primary appendiceal carcinoma—epidemiology, surgery and survival: results of a German multi-center study Eur J SurgOncol 2010368763–771.
4. Davison J M, Choudry H A, Pingpank J F. et al. Clinicopathologic and molecular analysis of disseminated appendiceal mucinous neoplasms: identification of factors predicting survival and proposed criteria for a three-tiered assessment of tumor grade. Mod Pathol. 2014;27(11):1521–1539.
5. Jacquet P, Sugarbaker P H. Clinical research methodologies in diagnosis and staging of patients with peritoneal carcinomatosis. Cancer Treat Res. 1996;82:359–374.
6. Low R N, Barone R M. Combined diffusion-weighted and gadolinium-enhanced MRI can accurately predict the peritoneal cancer index preoperatively in patients being considered for cytoreductive surgical procedures. Ann SurgOncol. 2012;19(5):1394–1401.
7. Turaga K, Levine E, Barone R. et al. Consensus guidelines from The American Society of Peritoneal Surface Malignancies on standardizing the delivery of hyperthermic intraperitoneal chemotherapy (HIPEC) in colorectal cancer patients in the United States. Ann SurgOncol. 2014;21(5):1501–1505.
8. Baratti D, Kusamura S, Nonaka D. et al. Pseudomyxomaperitonei: clinical pathological and biological prognostic factors in patients treated with cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) Ann Surg Oncol. 2008;15(2):526–534.