Mucinous Adenocarcinoma 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 appendicectomy, 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 appendicectomy and right sided salpingo-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 types. 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 years. Its histological classification is broad, as either low grade or high grade, which is extremely important in the management and prognosis of the disease.

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% . This commonly presents when the invasive tumor is ruptured, and causes a clinical syndrome of abdominal distension with mucinous ascites, known as pseudomyxoma 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.
High grade appendiceal mucinous neoplasms carry 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 CT or MRI. 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 metastasis.

Management of adenocarcinoma of the appendix 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, appendicectomy with cystectomy without spilling mucin intraperitoneally is recommended. If the mucin cyst is ruptured but there is no evidence of mucosal invasion, a right hemi-colectomy is preferred. For high grade carcinoma with evidence of distant metastasis, the gold standard recommendation is surgical cytoreduction and intraperitoneal chemotherapy. Surgical cytoreduction refers to complete gross resection of invaded tissue, which routinely consists of omentectomy, right quadrant peritonectomy and bilateral salpingo-oophorectomy. For widespread disease with extensive lympho vascular metastasis, such as the liver or porta hepatitis, intra peritoneal chemotherapy in the form of Hypothermic Intra-Peritoneal Chemotherapy (HIPEC) should be administered. A median 5 year survival rate of 75%-80% for low grade tumours, and 45%-65% for high grade tumours has been demonstrated.

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