Frontiers in Medical Case Reports, ISSN: 2582-8142

Intra-Operative Diagnosis of Mucinous Adenocarcinoma of The Appendix: A Case Report

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Received: 20 November 2020; **Accepted:** 05 December 2020

ABSTRACT

Primary Appendiceal neoplasms are rare and commonly difficult to diagnose due to their lack of specific clinical features. Here, we report an interesting case of an 86 year old otherwise fit and well lady presenting to this author's surgical assessment unit with a several week history of right sided abdominal pain and swelling, with the clinical features giving the clinical impression of a strangulated Spigelian Hernia. This was further backed up by the reported CT imaging of the abdomen. Intra-operative findings instead revealed extra-Peritoneal mucus with a macroscopically normal Appendix entering the abdominal wall defect. The pathological specimen was histologically confirmed to be a well differentiated Mucinous Adenocarcinoma of the Appendix.

Keywords: *Appendiceal Mucinous Neoplasms (AMNs), Strangulated Spigelian Hernia*

Introduction

Primary Appendiceal neoplasms are a rare pathological finding, present in >2% of Appendicectomies (Tirumani *et al*., 2013). They are commonly diagnosed after surgical intervention for acute Appendicitis or assumed Ovarian malignancy (Kelly, 2015), whilst specifically Appendiceal Mucinous Adenocarcinomas are even less common; presenting in 0.2% of Appendicectomies (Tirumani *et al*., 2013). Histologically, these neoplasms can be divided into Low Grade Appendiceal Mucinous Neoplasms (LAMN) and High Grade Mucinous Neoplasms (HAMN); both non-invasive, HAMN an intermediate stage between LAMN and Mucinous Appendiceal Adenocarcinoma (Ekta Sharma *et al*., 2020). LAMN itself can cause Pseudomyxoma Peritoni if the Appendix ruptures (Feely and Gonzalez, 2020). Clinical diagnosis of Appendiceal neoplasms is often challenging, with half of such cases asymptomatic, while other may present with, abdominal pain, weight loss, clinical features of acute Appendicitis, or a palpable mass (Tirumani *et al*., 2013).

Case Presentation

An 86 year old female patient was initially seen by her GP for new onset abdominal swelling, and was subsequently booked for an outpatient abdominal ultrasound, with no immediate further follow up. This patient was then referred directly to the on-call surgical team at this author's unit after being clinically examined by the GP, having given a 2 week history of worsening abdominal pain and swelling. Abdominal examination at this author's unit showed 10cm- 7cm irreducible swelling at the right lower quadrant, with moderate tenderness on palpation and erythematous skin changes. It should be noticed that the patient gave no history suggestive of bowel obstruction. The clinical impression from our initial clinical assessment was strangulated Spigelian Hernia.

Clinical Investigation

An urgent CT Abdomen and Pelvis with IV contrast was requested. This scan reiterated our earlier clinical impression, confirming the presence of an incarcerated Spigelian hernia with perforation of the bowel contents into the subcutaneous fat of the right abdominal wall. The CT report also described a small amount of fluid lying in an intra-abdominal location deep to the right abdominal wall and lateral to the hernia (**Fig. 1**).

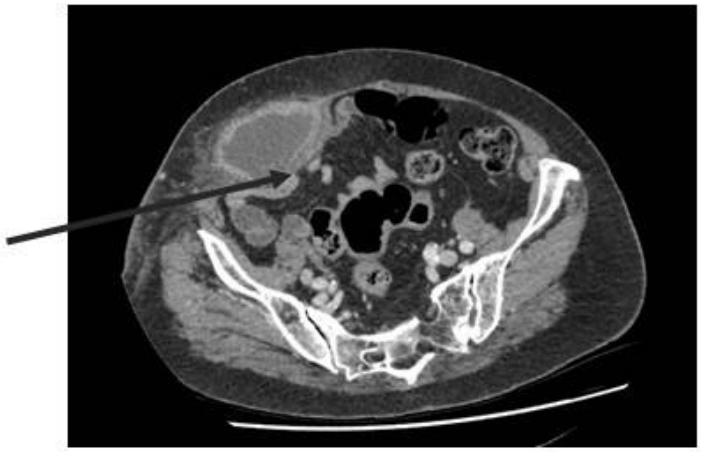

Figure 1: CT-AP - Axial cut showing protrusion of a mass into the right abdominal wall

Treatment

The patient was taken to the operating room with the intention of open hernia repair +/- further procedure. Initial transverse incision over the right sided abdominal wall swelling revealed infected extra-peritoneal mucus. The procedure was subsequently converted to a diagnostic Laparoscopy, which showed the Appendix and Omentum entering into the hernia sac. No intra-peritoneal mucin or intra--peritoneal malignant disease was seen. The base of the Appendix appeared macroscopically normal

with dilatation from the mid-appendix distally. The Appendix was removed using stapler at its base along with a cuff of Peritoneum and sent for Histopathological analysis. The extra-peritoneal mucin was aspirated, with wash out of the cavity site. The Peritoneal cavity was not contaminated with mucin. The initial incision of the right sided abdominal wall swelling was closed with interrupted sutures, with a Yeates drain left in situ to further facilitate drainage (**Fig .2** and **Fig. 3**).

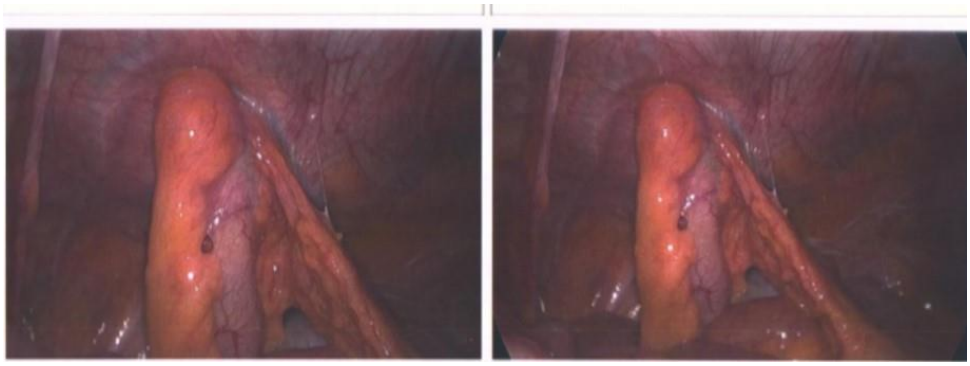

Figure 2 and Figure 3: Intra-operative photo during diagnostic Laparoscopy showing the Appendix perforating through the abdominal wall

Outcome and Follow Up

The patient was admitted to the Critical Care Unit post-operatively, and was stepped down to the surgical ward two days later. The patient suffered no post-operative complications, recovering well and discharged from hospital three days later. Histopathology of the Appendix confirmed well differentiated Mucinous Adenocarcinoma, G2 pT4b pNx.The case was discussed at the Multidisciplinary Colorectal Cancer Meeting at this author's Trust. The outcome reiterated the pathology as a tumour suggestive of a low grade Appendiceal mucinous neoplasm (LAMN), the albeit focal infiltrative pattern present within the adherent hernia sac more in keeping with a diagnosis of Mucinous Adenocarcinoma. The patient was then referred to a tertiary centre for further management.

Discussion

Reports on cases of Mucinous Adenocarcinomas remain few and far between. Diagnosis of the pathology remains difficult due to the overall lack of specific clinical features (Dang *et al*., 2017). A common theme in the literature describing such cases is that initial presentation may mimic acute Appendicitis or indeed present as a palpable abdominal wall mass. It is therefore important to consider such a diagnosis when encountering such presentations, particularly those with an atypical history or examination. As seen in this case, Mucinous Adenocarcinomas have a high risk of cavity planting, hence it is important to pay attention the operative field and wound in suspected or confirmed cases (Dang *et al*., 2017).

Learning Points

- Primary Appendiceal neoplasms, including Mucinous Adenocarcinoma, remain a rare pathological finding.
- Mucinous Adenocarcinoma is difficult to diagnose due to its non-specific clinical features; commonly features suggestive of acute Appendicitis or a palpable mass.
- It is important to consider this diagnosis, particularly when encountering such features with an atypical history or examination.
- Attention should be paid to the field and wound intra-operatively.

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