Pseudomyxoma Peritonei - “Jelly Belly”: To Tap or Not?

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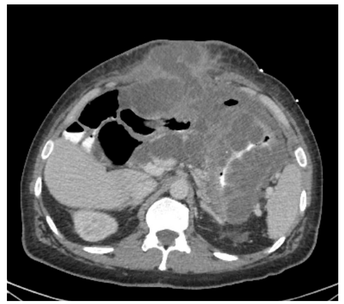
Introduction

Appendiceal mucinous neoplasms are rare tumors accounting for less than 1% of all cancers. (1)

Pseudomyxoma peritonei is an extremely rare complication of appendiceal mucinous adenocarcinomas with an estimated incidence rate of one to two people per million per year. (2)

It is characterized by production of mucinous and gelatinous masses and is associated with rupture of appendiceal mucinous tumors and other mucinous tumors of the gastrointestinal tract. (2)

Here-in we present a unique case of enterocutaneous fistula formation secondary to percutaneous biopsy of an enlarging omental mass in the setting of pseudomyxoma peritonei.



Picture 1: CT abdomen and pelvis with PO and IV contrast axial imaging demonstrating multi-cystic mass lesion in the peritoneal cavity consistent with pseudomyxoma peritonei, as well as continuation of cystic lesions from the peritoneal cavity into the abdominal wall

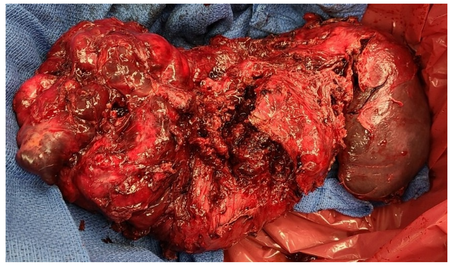
Case Description

A 50-year-old male with a past medical history of metastatic appendiceal mucinous adenocarcinoma presented to the ED with abdominal pain, nausea, and vomiting. The patient had previously undergone 2 debulking surgeries over the past two years prior to admission and had since been on FOLFOX (folinic acid, fluorouracil, and oxaliplatin) therapy.

Due to the COVID pandemic, the patient did not follow-up in the two years period from previous admission. A CT scan was now notable for a new enlarging omental mass despite the recent debulking surgery. Given the enlarging mass, a decision was made to pursue a percutaneous biopsy of the mass due to concern for potential new malignancy.

Two weeks after the biopsy, the patient presented to our facility due to worsening erythema and drainage from the biopsy site. The patient met SIRS criteria, thus broad-spectrum antibiotics were initiated. A CT of the abdomen and pelvis with oral and IV contrast was obtained, which demonstrated a 9 cm abscess or continuation of intra-abdominal multilocular cystic lesion/ pseudomyxoma peritonei. The surgical team was consulted for evaluation of the fistula.

Patient had 100 cc of purulent and mucinous drainage expressed from biopsy site. The patient was then placed for transfer to a hospital capable of advanced surgical management for evaluation and potential resection of fistula formation. The patient had a successful reductive surgery and intraoperative chemotherapy.



Picture 2: Resected omental mass with notable adherent mucinous masses, characteristic of pseudomyxoma peritonei

Discussion

Given the rarity of pseudomyxoma peritonei, appropriate management is not always straightforward.

Due to the heterogeneity of these tumors, treatment varies, with peritoneal debulking and HIPEC (hyperthermic intraperitoneal chemotherapy) being the choice of treatment in patients with advanced stage disease (1)

A literature review yielded no previous reports of enterocutaneous fistula as a complication of percutaneous biopsy in the setting of pseudomyxoma peritonei.

We recommend that percutaneous drainage not be sought in individuals with this diagnosis due to potential for fistula formation.

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References

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