Pseudomyxoma Peritonei (PMP)
"Jelly Belly"
A Postoperative Home Palliative-Hospice Case Study

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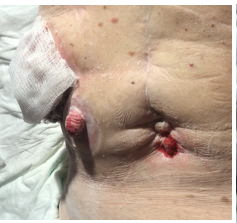
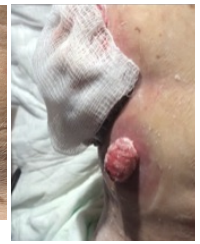
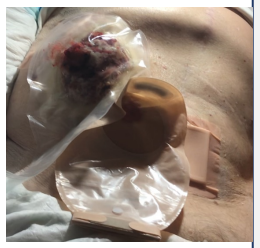
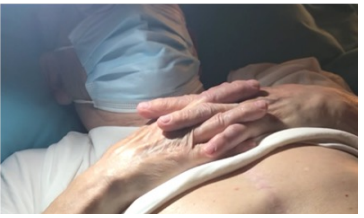
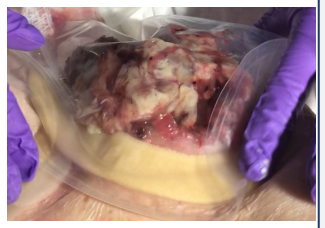
CASE STUDY

- DS 80y/o male, alert, oriented independent ADLs
- Hospice Services 06/2021 with a primary diagnosis of Malignant Neoplasm of ascending colon, Specifically: Metastatic Pseudomyxoma Peritonei (PMP) "jelly belly"
- Prior to hospice, s/p diverting laparoscopic ileostomy, due to concern for ischemic colon from increased abdominal pressure from PMP
- PMP progressed; mucus and mucin increased the abdominal pressure creating tumor and fistulas out onto the patient's abdomen; primarily at the ileostomy site and failed to heal laparoscopic sites

Defining PMP:

- *"A condition characterized by poorly-circumscribed gelatinous masses filled with malignant mucin-secreting cells"* (The National Library of Medicine)
- PMP appears as a distinct histologic diagnosis in the classification of tumors of the digestive system, and it is also used as a description of the macroscopic appearance of mucinous ascites (WHO, 2010) PMP evolved into a high- and low-grade disease classifications (WHO, 2019)
- High controversy in definition and classification, that has evolved since 1884 when first thought as originating from ruptured ovarian cysts.
- PMP arises most often from appendiceal neoplasms, and rarely from other tumors such as neoplasms from the colon, urachus, or pancreas.
- Takes the appearance of a mucocele as its clinical presentation.
- PMP is classified according to the histology of the peritoneal disease rather than the primary tumor.

Epidemiology/Prevalence:

- Approx. 1/3 of appendiceal epithelial lesions progress to PMP
- Unclear data of those afflicted, 1-4 people per 1,000,000 people.
- 1-2 million cases per year (national organization for rare disorders)
- Occurs equally between men and women with an average onset of 48y/o

Physical Presentation:

- PMP derives from cells of low-grade malignancy, with poor aggressive behavior in terms of growth rate and systemic metastasis.
- Mucin around tumor cells allows them to disseminate and redistribute within the peritoneal cavity.

Treatment:

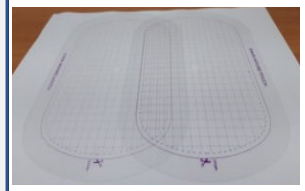
- Cytoreductive surgery, is main treatment, it is the only therapy with a potential curative option. There is low indication for systemic chemotherapy, hyperthermic intraperitoneal chemotherapy (HIPEC), due to PMP's slow growth and divisional rate, and considered for patients who have no surgical options.
- When surgery/chemotherapy are not an option, no effective treatments exist.

Survival Rate:

- 10-year survival of around 63%, almost all PMP patients have recurrences; 98.7% are reported after 10 years. If no reoccurrence, considered cured.
- Patients may live for many years with active disease, slow worsening symptoms

Evolution of Pouching as Disease Evolved & Expanded


As PMP continued to grow, larger wound pouches were used to contain tumor sites. Ostomy pouches transitioned from a 1-piece drainable pouch with a large wafer/flange pouching surface to a 1-piece high output pouch due to large output, to a 2-piece system that supported a pediatric wafer and a high output pouch with a spout, to manage shrinking pouching surface area. Eventually the pouching system was transitioned to two large wound pouches and the Saddlebag technique was implemented.



Challenges

- Shrinking pouching surface areas, as tumor burden enlarged
- Extreme high liquid output; despite stool thicken agents, PO intake adequate but could not stay hydrated, required daily 500cc NS IV fluid bolus. Portable suction at bedside in the home, to manage during pouching changes.
- Goals to go to a Red Sox Game, and a High School Graduation, without leaking from pouching system.
- Patient VERY alert and involved in care, his decision to keep stool and tumor separate, with goals of care (GOC) to respect that decision, and to have peaceful pouch changes that he "zened out" to.

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