CLINICAL VIGNETTE

Jelly Belly: A Case of *pseudomyxoma peritonei*

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A 35-year-old male presented to the clinic to establish primary care. Patient was diagnosed with pseudomyxoma peritonei 6 years ago after he presented with increasing abdominal girth for about a year prior. Patient had been generally healthy with no previous medical problems. He had a past surgical history of tonsillectomy and removal of a benign cyst on the chin. He is a non-smoker, drinks alcohol, 1.2 oz per week, and has no history of illicit drug use. There is no family history of cancer.

On exam, he had normal vital signs, normal cardiac and pulmonary findings. Chest wall was asymmetric. Abdomen was non-distended with normoactive bowel sounds. He had multiple scars from his previous surgeries.

Seven years ago he noted increasing abdominal girth. Abdominal CT showed umbilical hernia for which he underwent repair. Surgery findings were "a small to moderate umbilical hernia with omental caking with mucinous peritoneal implants in and around the sac" with 1000 cc of cloudy yellowish ascites.

Peritoneal tissue biopsy showed "fibroadipose tissue with dissecting acellular mucin" and second peritoneal biopsy, showed "fibrous tissue with dissecting mucin and focal low grade epithelial cluster". Ascites fluid showed mucin and many glandular aggregates with mild to moderate atypia. Clinical and pathologic findings were "consistent with pseudomyxoma peritonei; the glandular component shows mild to moderate atypia with no high grade cytologic and architectural features."

Two months later, patient underwent cytoreductive surgery (CRS) with hemicolectomy, splenectomy, distal pancreatectomy, omentectomy, cholecystectomy, peritonectomy, removal of terminal ileum and partial gastrectomy and heated intraperitoneal chemotherapy (HIPEC). Pathology reported low-grade appendiceal mucinous adenocarcinoma.

Patient received intraperitoneal chemotherapy with mitomycin. He had FOLFOX therapy for 8 cycles but this was stopped after 8 cycles due to peripheral neuropathy.

The patient did well until 10 months later when his chest CT showed "new and increased complex pleural-based collections in the right lower chest suspicious for pleural-based metastatic disease". PET scan showed "extensive pleural-based abnormalities invoking the right hemithorax consistent with a neoplastic process".

Patient underwent pleurectomy, resection of the rib, diaphragm, with a pericardial window, intrapleural chemotherapy. Pathology was consistent with low grade well-differentiated appendicial mucinous adenocarcinoma with four lymph nodes biopsied showing no involvement.

Two years later, he was noted to have a right subcutaneous abdominal nodule. He was given antibiotics without any improvement. The nodule continued to grow and eventually ruptured, releasing purulent drainage which grew Strep viridans. The decision was made to surgically remove his mesh in the diaphragm. He also underwent flexible bronchoscopy with bronchoalveolar lavage, exploratory redo right thoracoscopy, removal of diaphragm patch, pleural biopsy, washout, and drain placement.

Discussion

Pseudomyxoma peritonei (PMP) refers to a "clinical syndrome characterized by diffuse mucinous peritoneal involvement, often associated with a mucinous appendiceal lesion that has presumably ruptured".[1]

PMP is a rare disease with incidence of one per million per year and the name "jelly belly" came from the copious production of gelatinous, mucinous ascites that fills the peritoneal cavity.[2] While often considered a benign condition, over time, the disease persists and recurs, and should be considered a borderline malignant disease .

It is believed that PMP originates in the appendix in men and recent data suggests that it is also the most common site of origin for women. PMP have been associated with ovarian tumors, but there are reports originating from the colon, stomach, gallbladder, small intestine and urinary bladder, fallopian tube and pancreas, although rare, accounting for less than 5%.[2]

PMP is more common in females. The most common presentation is increasing abdominal girth in both sexes, followed by inguinal hernia and ovarian mass in women noted during routine pelvic exam.[1] In men, the other clinical presentation is acute appendicitis.[3] However, the majority are diagnosed after laparotomy or during surgery.

CT findings may lead to suspicion of PMP such as scalloping of the liver, spleen, mesentery. A load of tumor surrounding the diaphragmatic surface of the liver gives it a "scalloped"

appearance. Another significant finding is the "redistribution phenomenon" whereby the tumor is located at the periphery in the abdomen and pelvis with sparing of the small intestine and mesentery.[1] In one study, patients with scalloping with greater mucin deposition significantly correlated with poorer disease-free survival.[4]

Pathology

Mucus is the predominant element with sparse cells with features of low-grade adenocarcinoma when lymph node metastasis are absent. Thus, it is believed to be "benign" and has been treated with by debulking procedures. However, due to tumor recurrence and progression, it should be regarded as "borderline malignant" Lymphatic and hematogenous spread may not occur, even with terminal stages of the disease.

Some features that show increase aggressive disease include short interval from first to subsequent presentations, cachexia and wasting, findings of solid lumps in exam and elevated CEA, CA 12-5 and CA 19-9.[2]

Management of PMP

The mainstay of treatment is surgery and the role of radiation is not clear. The two surgical approaches to PMP are "debulking" and peritonectomy. With debulking, as much as possible of the mucus and tumor are removed by blunt dissection with right hemicolectomy and partial omentectomy. In females, this also includes hysterectomy and bilateral oophorectomy. An alternative approach is complete tumor removal based on the position of the tumor. The mucinous tumors coat parietal peritoneal surfaces with surgery involving 6 peritonectomy procedures: greater omentectomy with splenectomy, stripping of left and right hemidiaphragm, cholecystectomy and lesser omentectomy, distal gastrectomy and pelvic peritonectomy with resection of the rectosigmoid.[2]

Surgery usually lasts 10 hours with significant morbidity and mortality. Mortality rates with experienced surgeons is 3- 5%, with sepsis as the most cause of death or cardio pulmonary complications. Morbidities arise from fistula, anastomotic leakage, re-operation for bleeding and thromboembolic problems. Quality of cytoreductive surgery (CRS) is dependent on the skills and experience of the surgeon.[5]

One aggressive approach includes cytoreductive surgery with heated intraperitoneal chemotherapy (HIPEC). The use of both these treatments improves survival, quality of life and preserves function.[6] Drug penetration can be enhanced by heating the perfusate containing chemotherapy.[6] In the US, mitomycin is used in most centers and oxaliplatin is used in Europe.[1] The importance of surgical technique and surgeon experience to the success and safety of this approach is important.[1]

Many post-surgical patients do not have symptoms but the disease eventually recurs in the form of small bowel obstruction or gross distention. Recurrences are treated with further laparotomy and debulking, although these becomes more dangerous with increased risk of bowel injury from fistula or peritonitis.

The 5-year survival can range from 53 to 75%, depending on benign and malignant subgroups.[7] A retrospective study with well-selected patients who had undergone initial resection for low grade mutinous tumor of the appendix with limited spread, 5 year overall survival was 95% and progression free survival was 82%.[8]

In this case, patient had both CRS and HIPEC. Recurrence was noted 10 months post-surgery. At six years post-surgery, patient is doing well except for occasional respiratory issues treated with maintenance inhalers. He has had no other septic episodes post-splenectomy.

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