An Unusual Etiology of Increased Abdominal Girth in a 42-Year-Old Man

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A 42-year-old man with diabetes and hypertension presented to the emergency room after experiencing a several month history of gradually increasing abdominal girth with the sudden onset of abdominal pain.

Figure 1. Axial CT

Figure 2. Axial PET/CT

Figure 3. Sagittal PET/CT

Figure 4: Coronal PET/CT
Radiologic Diagnosis: *Pseudomyxoma Peritonei*

Interpretation of Findings: Figure 1 demonstrates mucinous peritoneal deposits with scalloping of the liver, spleen, and stomach. A septum divides the mucinous material adjacent to the liver (arrow). Figures 2-4 demonstrate decreased metabolic activity within these areas.

**DISCUSSION**

Pseudomyxoma peritonei (PMP) is a rare disease, which involves progressive growth of diffuse mucinous fluid deposits in the peritoneum. Rokitansky first described PMP as “jelly belly,” in 1842. The term “PMP” was first coined in 1884 by Werth. It has an overall incidence of 1-2 per million per year, more common in women than men, and has an average age of diagnosis of 53 years. The most likely site of origin is the appendix, though other primary sites include the large and small bowels, stomach, bile ducts, gallbladder, pancreas, ovary, fallopian tubes, urachus, urinary bladder, breast, and lung.

PMP most commonly originates as a cystadenoma or cystadenocarcinoma of the appendix. Spreading of the tumor involves its overgrowth in the appendiceal lumen with subsequent rupture of the organ. The peritoneal cavity and abdominopelvic organs are then seeded with mucin-producing cells, which gradually proliferate. PMP can manifest anytime between a few years to a few decades after the initial appendiceal rupture, often leading to a delayed diagnosis. If left untreated, the gelatinous growths may eventually compress the gastrointestinal tract, causing bowel obstruction and death.

PMP is often encountered unexpectedly during surgery, appearing grossly in two of every 10,000 laparotomies. In many women with PMP, the bilateral ovaries feature multilocular mucin-filled cysts, with the appendiceal primary not being grossly visible. Mucinous pockets may also be incidentally discovered in surgical specimens such as hernia sacs. Pathohistological confirmation of PMP is made using samples from either the primary tumor or peritoneal mucin pools. Tumor cells of appendiceal origin feature low-grade differentiation and may be elongated and hyperchromatic. Immunohistochemically, tumors of ovarian epithelium origin stain positive for cytokeratin 7 (CK7) and stain negative for cytokeratin 20 (CK20), while tumors of appendiceal epithelium origin stain positive for CK20 and stain negative for CK7. Some tumors will stain neither for CK7 nor CK20 but may stain positive for other markers such as CEA, CA19.9, and CDX2, which is expressed by the gastrointestinal tract and pancreato-biliary systems.

Prognosis depends on the histopathologic categorization of the disease, of which there is no definite consensus in the literature. One such classification divides PMP into three pathological grades. Grade I, or disseminated peritoneal adenomucinosis (DPAM), has a 75 percent five-year and an 86 percent ten-year survival rate. Grade III or peritoneal mucinous carcinomatosis (PMCA) has a 14 percent five-year and a 3 percent ten-year survival rate. Grade II, which is a hybrid form named peritoneal mucinous carcinomatosis with intermediate or discordant features (PMCA-I/D), has a five-year and ten-year survival rate of 50 percent and 21 percent, respectively.

Clinically, increasing abdominal girth is the most common symptom seen in both men and women with PMP. Men may present with an inguinal hernia, and women may present with a palpable ovarian mass. Narrowing or obstruction of the bowels, occurring later in the disease process, most commonly occurs at retroperitoneal sites such as the pyloric antrum, ileocecal valve, and the Pouch of Douglas. Treatment of PMP is usually performed with cytoreductive surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC), with varying degrees of success and associated morbidity.

CT remains the conventional imaging modality for PMP, and it appears to be more sensitive than positron emission tomography (PET) for preoperative staging of peritoneal metastases such as those found in PMP. Furthermore, it allows direct visualization of anatomical findings, such as visceral scalloping, septae in mucinous material, calcification, omental caking, low attenuation ascites, and in some instances an appendiceal mucocele.

However, PET imaging may play an important role in the management of PMP. As Passot et al. proposed, by differentiating patients with an absence of uptake from those with uptake, PET scanning suggests those who may have DPAM versus PMCA or PMCA-I/D. As such, PET imaging may help predict the pathological grade and effectiveness of therapy, which is important due to the morbidity and mortality between the histopathologic categories and with the CRS and HIPEC treatment options. However, PET scanning alone is insufficient to distinguish between DPAM, PMCA, and PMCA-I/D. An important potential pitfall with PET imaging is that residual inflammatory changes after surgery may cause uptake of tracer, leading to false positive results. Ultimately, studies suggest that the sensitivity and specificity for pretreatment evaluation of patients with PMP is improved when combining the aforementioned anatomic data of CT with the functional information of PET.

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