

FEATURE ARTICLE

Pseudomyxoma Peritonei Syndrome of Appendiceal Origin: An Overview

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Pseudomyxoma peritonei (PMP) syndrome is an uncommon, slowly progressive condition that usually arises from perforation of an adenoma in the appendix. PMP syndrome is characterized by mucin accumulation in the peritoneal cavity. Mucinous implants are found on all peritoneal surfaces and the omentum. PMP syndrome rarely metastasizes outside the abdominal cavity but remains a fatal illness as the space in the abdomen and pelvis required for normal function of the gastrointestinal tract becomes filled with copious amounts of the mucinous tumor. Treatment options include observation, aggressive debulking surgery, intraperitoneal chemotherapy, radiotherapy, and mucolytic agents.

Pseudomyxoma peritonei (PMP) syndrome is a rare condition characterized by large amounts of mucinous ascites that accumulate in the abdomen and pelvis. Historically, the term “jelly belly” has been used to describe the gelatinous-like fluid that implants on the peritoneal surfaces and omentum (Harshen, Jyothirmayi, & Mithal, 2003; Hinson & Ambrose, 1998) (see Figure 1). PMP syndrome is considered to be a borderline malignancy because the tumor usually is not aggressive and metastasis to solid organs via the lymphatic system or bloodstream does not occur. PMP syndrome remains a fatal condition. Eventually, space in the abdomen and pelvis becomes filled with a tumor of varying consistency that causes intestinal obstruction (Hinson & Ambrose; Sugarbaker, Fernandez-Trigo, & Shamsa, 1996; Witham, 2003).

Originally, PMP syndrome was used to describe mucinous ascites associated with a ruptured appendix mucocele. Many oncologists and pathologists have applied the term to any disease in the abdomen and pelvis characterized by mucinous fluid (Sugarbaker et al., 1997). As a result, prognosis of the disease has been variable and unpredictable.

PMP syndrome has been histologically classified into three categories: disseminated peritoneal adenomucinosis (DPAM), peritoneal mucinous carcinomatosis (PMCA), and PMCA with intermediate or discordant features (PMCA-I/D). Classification is based on the morphologic features of the epithelium identified in peritoneal lesions and the nature of the underlying lesion. DPAM describes lesions that have little cytologic atypia or mitotic activity, whereas cytologic features of carcinoma can be found in PMCA and PMCA-I/D, which are more aggressive. The classification system is important because survival rates have been found to differ significantly for each category and may affect treatment choices (Ronnelt, Zahn, et al., 1995).

At a Glance

- ◆ Pseudomyxoma peritonei (PMP) syndrome is an uncommon, slowly progressive disease characterized by large amounts of mucinous ascites, which accumulate in the abdomen and pelvis. Five-year survival rates for PMP syndrome range from 75%–86%.
- ◆ Treatment options for PMP syndrome are multimodal and include observation, aggressive debulking surgery, intraperitoneal chemotherapy, radiotherapy, and mucolytic agents.
- ◆ Treating PMP syndrome with intraperitoneal chemotherapy allows higher concentrations of cytotoxic agents to be administered directly into abdominal and pelvic surfaces where the tumor is located without producing toxic systemic levels.

PMP syndrome should be applied only to histologically benign peritoneal tumors that are slow growing and associated most commonly with appendiceal mucinous adenoma. Pathologically, mucinous lesions are classified as DPAM or adenomucinosis (Sugarbaker et al., 1997). This article will focus on the DPAM pathologic classification.

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