



Pseudomyxoma Peritonei, an Uncommon Tumor with Ongoing Debatable Nomenclature and Classification. Report of a Case and Review of the Literature

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Abstract

Pseudomyxoma peritonei is a rare condition, characterized by gelatinous ascites of the abdominal and pelvic cavities accompanied by mucinous implantations of the peritoneum and omentum. It is usually presented as a disseminated mucinous tumor with dissection of the abdominal and pelvic organs producing pseudomyxoma peritonei. Mucinous implants usually involve the serosal surface of multiple pelvic and abdominal organs. Although it is accepted that most investigators believe that the ovarian tumors are secondary in almost all cases to a primary appendiceal tumor, a synchronous origin in both organs has also been proposed. The agreement on the association of certain tumor morphologies with certain behaviors is persistently plagued by disagreement regarding tumor nomenclature and classification. We present a case of recurrent pseudomyxoma peritonei and review the literature of nomenclature, classification, diagnosis, and management of these tumors.

Keywords: Pseudomyxoma, Peritonei, Mucin, Appendix, Ovary, Peritoneum

ABBREVIATION

Pseudomyxoma peritonei (**PP**), disseminated peritoneal adenomucinosis (**DPAM**), peritoneal mucinous carcinomatosis (**PMCA**), hyperthermic intraperitoneal chemotherapy (**HIPEC**), cytoreductive surgery (**CRS**)

INTRODUCTION

Pseudomyxoma peritonei (PP) is a rare condition, with an incidence of 1 to 2 in 1 million per year [1]. PP represents a broad spectrum of neoplastic disorders ranging from benign to borderline and malignant lesions. It is originally reported as disseminated peritoneal adenomucinosis (DPAM), which describes the benign variant whereas peritoneal mucinous carcinomatosis (PMCA) describes the malignant variant [2,3]. Gelatinous ascites of the abdominal and pelvic cavities accompanied by mucinous implantations of the peritoneum and omentum characterize findings associated with PP [4].

Histopathologically, the primary tumor predominately

resembles a mucinous epithelial neoplasm of the appendix [1,5-7]. PP displays a wide range in the mucus/cell ratio in addition to the level of differentiation and grade of atypia of epithelial cells. However, proper histopathological characterization of PP remains difficult due to the heterogeneous appearance [1,2].

PP more commonly presents in women (male:female ratio = 9:11), with an average age of 53 years. In women, usual presentation is increasing abdominal girth and a primary ovarian lesion [8,9]. Most cases of ovarian involvement likely represent metastasis from an appendiceal or other gastrointestinal source [10]. Immunohistochemistry studies are useful in confirmation of appendiceal origin as they usually stained negative for CK-7 and positive for CK-20 and CDX2 in support of appendiceal origin. This pattern of immuno profile is reversed if it was of ovarian origin.

Diagnosis of PP requires the presence of mucinous neoplastic epithelial cells and diffuse intra-abdominal mucinous ascites. Some clinicians require the presence of diffuse mucinous peritoneal implants for definitive diagnosis [3].

PP warrants early intervention due to its high mortality rate when not properly managed [11]. Without treatment, mucin accumulation and large volume ascites compress vital internal organs such as the colon, liver, kidneys, stomach, spleen, and pancreas resulting in bowel obstruction and eventual death [1,5,9]. Proper diagnostic investigations include an ultrasonographic examination of the abdomen followed by computed tomography scans, showing the extent of the disease [12]. Additionally, the evaluation of tumor markers in serum, such as CA 19-9 and carcinoembryonic antigen (CEA), help determine prognosis [13]. The current optimal treatment involves macroscopic tumor excision cytoreductive surgery (CRS) combined with hyperthermic intraperitoneal chemotherapy (HIPEC) [14].

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CASE PRESENTATION

A 39-year-old woman presented to her surgeon with complaint suggestive of umbilical hernia. The patient has history of appendectomy with mucinous cystadenoma and pseudomyxoma peritonei (PP) two years prior to current presentation. Prior tumor was treated with repeated paracentesis of the mucinous ascites and surgical debulking of the primary tumor with appendectomy and removal of mucinous material. Thoracoabdominal contrast-enhanced MRI and CT scan showed no ovarian masses or other solid tumor. However, multiple peritoneal implants were noted with extension to the surface of the ovaries. Serum tumour markers showed elevated levels of CEA, CA-125, and CA19-9. The clinical presentation was consistent with recurrent disseminated PP with aggressive mucinous dissecting features.

The tumor was treated with surgical debulking with macroscopic tumor excision cytoreductive surgery (CRS) in addition to removal of both ovaries, spleen, portions of diaphragm, colon and small bowel as well as all peritoneal implants. Pathologic microscopic examination showed disseminated mucinous tumor with dissection of the ovarian and peritoneal stroma producing pseudomyxoma ovarii/pseudomyxoma peritonei (**Figure 1A**). Mucinous implants involved the serosal surface of multiple pelvic and abdominal organs including spleen, diaphragm, small intestine, colon and omentum (**Figure 1B**). No evidence of infiltration into skeletal muscle was noted and two removed lymph nodes were negative for malignant epithelium. Epithelial glandular structures were seen within the dissecting mucous. Some nuclear atypia was noted (**Figure 1C**). Although there was no definitive evidence of invasion, occasional epithelium within stromal fragments suggestive of possible invasion was noted. For this reason, the tumor was classified as “with intermediate

features’ (between low grade adeno mucinosis and well differentiated mucinous carcinomatosis). The epithelial atypical cells stained negative for CK-7 and positive for CDX2 (**Figure 1D**) and CK-20 (**Figure 1E**) in support of appendiceal origin. Surgery was combined with hyperthermic intraperitoneal chemotherapy (HIPEC) followed by postoperative chemotherapy utilizing cisplatin, and oxaliplatin.

The patient was followed up for five years with no evidence of recurrence or metastasis after which she was lost to follow up.

DISCUSSION

Pseudomyxoma peritonei (PP) is a rare disease characterized by disseminated mucinous ascites with peritoneal, serosal, and omental mucinous implants [3]. Typically, the pathologic process initiates with neoplastic proliferation of appendiceal goblet cells into the peritoneal cavity following perforation of the appendix [15]. Mucin overexpression results in intraperitoneal voluminous PP [16]. Gravity and physical factors such as movement of peritoneal fluid in the abdomen lead to accumulation of mucinous deposits at the omentum, retrohepatic region, and rectovesical pouch [17]. Visceral “scalloping” arises from mucinous compression and fibrosis of organs and is pathognomonic of PP and can be visualized on CT [18]. While generally regarded as benign, PP frequently demonstrates a borderline-malignant presentation with unfavorable progression of the disease and advanced-stage at diagnosis [19,20].

Although it is accepted that most investigators believe that the ovarian tumors are secondary in almost all cases to a primary appendiceal tumor, a synchronous origin in both organs has also been proposed [21]. The agreement on the association of certain tumor morphologies with certain behaviors is persistently

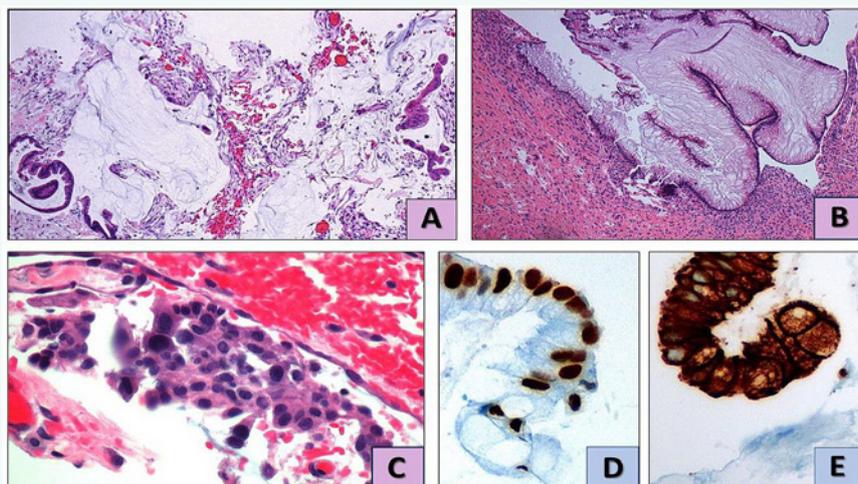


Figure 1 Pathological examination of the excised tumor

1A: disseminated mucinous tumor with dissection of the ovarian and peritoneal stroma producing pseudomyxoma ovarii/pseudomyxoma peritonei (H&E stain X20)

1B: Mucinous implants involved the serosal surface of multiple pelvic and abdominal organs (H&E stain X40)

1C: Tumor cells showing focal nuclear atypia (H&E stain X60)

1D: Tumor cells positive for CDX2 with nuclear staining

1E: Tumor cells positive for CK-20 with cytoplasmic staining



plagued by disagreement regarding tumor nomenclature and classification. Some investigators may consider the current tumor as a tumor with distinctive features of intermediate/hybrid tumors and even grouped it with the carcinoma. Other investigators may classify the current tumor with the concept of a clinically malignant, but pathologically benign, and in such case, they would classify it as well differentiated mucinous adenocarcinoma [22].

The clinical presentation of PP encompasses a broad spectrum with many non-specific findings resulting in up to 20% of cases diagnosed incidentally at laparoscopy or laparotomy [11,23]. In patients with an appendiceal mucocele, seeding of a mucinous epithelial neoplasm secondary to perforation, may not always appear macroscopically. Although small deposits tumor cells may appear on the surface of the appendix, frequently no sign of intraperitoneal tumor or mucus presents itself [1]. The lack of pathognomonic symptoms combined with the insidious clinical course results in local invasion of nearby structures, such as a reported case of urinary bladder involvement at the time of diagnosis [24]. Some common presenting features include increased abdominal girth (40%), bilateral or unilateral ovarian tumors (20%), hernia sac tumors (20%), appendicitis-like syndrome (10%) and infertility [8,9,26]. A retrospective review of 410 patients revealed acute appendicitis as the most common clinical presentation, occurring in 27% of male and female patients. However, ovarian mass presented most commonly in female patients (34%) [8].

Additionally, new-onset hernia caused by increased abdominal pressure often presents as a patient's chief complaint [20]. Abdominal exam may reveal a palpable omental cake or ovarian mass indicating peritoneal extension. Mucinous deposits in the pouch of Douglas or rectovesical pouch may be present upon rectal exam. In patients with vague abdominal complaints and distention CT served as the most common imaging technique used to diagnose PP. Abdominal and pelvic magnetic resonance imaging (MRI) may help determine any small bowel or hepatoduodenal ligament involvement [14].

Serum tumour markers may predict the aggressiveness and prognosis of PP [20]. A study of 519 PP patients revealed that elevation levels of CEA, CA-125, and CA19-9 prior to CRS and HIPEC correlated with decreased overall survival and disease-free survival [27].

PP rarely results in blood-borne or lymphatic metastasis but instead demonstrates a tendency for local recurrence [25,28,29]. Studies suggest that ovarian disease results from local invasion due to appendiceal perforation rather than as the site of origin [10]. In fact, most cases represent primary appendiceal mucinous neoplasms [2,30,31], while other reports indicate other cases with ovarian origin [32]. One literature review presented eighteen cases of the urachus as the tissue of origin [33]. Ultimately, the degree of local invasion throughout the peritoneal cavity represents a major factor in disease prognosis [11]. In our presented case, two lymph nodes were removed and showed no evidence of metastasis.

An early study conducted an aggressive treatment approach consisting of cytoreductive surgery with multiple operations. The center reported an average of 2.2 debulking operations required to reach complete cytoreduction in 55% of patients. Their treatment strategy resulted in a 10-years survival rate of 21%, with a 12% disease free rate at conclusion of follow-up. However, repetitive surgical debulking may result in imminent recurrence or disease progression due to intraperitoneal seeding of microscopic tumor residue [34; 1]. HIPEC showed reduced recurrence or progression of disease in patients with microscopic to minimal macroscopic tumor residue with some studies reporting a potential cytotoxic effect up to a tumor depth of 2.5 mm [1, 35].

Intraperitoneal chemotherapy provides the benefit of decreased systemic effects and therapeutic concentrations reached at lower doses [36]. Mitomycin C, 5-fluorouracil, cisplatin, and oxaliplatin represent the most commonly used chemotherapeutic agents [37]. Following CRS + HIPEC, a CT scan alongside physical examination and serum tumor markers represent invaluable tools for detecting disease progression. CEA, CA19-9 and CA12.5 tumor markers are measured every 3 months while a CT scan is every 6 months for 5 years status post treatment. Following, a CT scan is performed every 2 years for up to 10 years [38-40].

Previously, management of PP involved repeated paracentesis of ascites or surgical debulking of the primary tumor and mucinous material [41]. Repeated debulking surgery and intraperitoneal chemotherapy limited conventional treatment. The current optimal treatment involves macroscopic tumor excision, known as cytoreductive CRS, combined with HIPEC [14]. Further studies demonstrated the efficacy of this combined approach, with a 10-year morbidity and mortality in a specialized unit setting reaching 63% [41]. Another study observed that following CRS and HIPEC combined treatment, low-grade mucinous adenocarcinomas demonstrated a 5-year survival range from 62.5 to 100%, while high-grade mucinous adenocarcinomas demonstrated 0-65% [19]. Chua et al. (2012) demonstrated that HIPEC alone improved the rate of progression-free survival, but not overall survival. Therefore, while HIPEC improves disease control, long-term survival may require complete cytoreduction. Our patient was treated with debulking surgery combined with hyperthermic intraperitoneal chemotherapy (HIPEC) and followed by postoperative chemotherapy utilizing cisplatin, and oxaliplatin.

Pseudomyxoma peritonei secondary to appendicular mucinous tumors is an exceedingly uncommon tumor with a varied presentation and outcomes. It is our hope that this report raises awareness of clinicians and pathologists to these type of tumors with definitive diagnosis and management of various ovarian and peritoneal malignancies. Furthermore, that the continued investigation drives further development of efficacious diagnosis and safe treatments for improving patient outcomes.



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