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A Rare Clinical Finding of Pseudomyxoma Peritonei in a Case of Abnormal Uterine Bleeding

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ABSTRACT

Pseudomyxoma peritonei (PMP) is a rare condition marked by mucinous ascites and widespread peritoneal tumor deposits, most commonly arising from a perforated mucinous tumor of the appendix. This case report presents an atypical manifestation of PMP in a patient who initially presented with abnormal uterine bleeding. The report highlights the clinical presentation, diagnostic workup, and therapeutic approach.

The patient exhibited progressive abdominal distension, discomfort, and gastrointestinal symptoms. Diagnostic confirmation was achieved through detailed clinical evaluation, radiological imaging, and histopathological analysis. Surgical management involved cytoreductive surgery followed by hyperthermic intraperitoneal chemotherapy (HIPEC), which significantly enhanced the patient's prognosis.

Although PMP is uncommon, early recognition and prompt intervention are critical for improving outcomes. The combined approach of cytoreductive surgery and HIPEC remains the current gold standard in treatment. This case emphasizes the need for heightened clinical suspicion and a coordinated, multidisciplinary strategy to achieve optimal patient care.

Keywords: *Pseudomyxoma peritonei, abnormal uterine bleeding, appendicular mass, hysterectomy, HIPEC, case report*

1. INTRODUCTION

Case Presentation

Pseudomyxoma peritonei (PMP) is a rare clinicopathological entity characterized by mucinous ascites and widespread peritoneal tumor implants, most commonly originating from a ruptured appendiceal mucinous neoplasm. We report the case of a 40-year-old woman, Mrs. K, who presented to the Obstetrics and Gynecology Outpatient Department with complaints of persistent heavy menstrual bleeding and severe dysmenorrhea for 1.5 years. Despite various medical treatments, her symptoms remained unresolved. She had no known comorbidities. Clinical examination revealed a bulky uterus and fullness in the left fornix. Imaging studies identified a bulky uterus, a right hemorrhagic cyst, and a complex left ovarian cyst. Based on these findings, a Total Abdominal Hysterectomy with Bilateral Salpingo-Oophorectomy (TAH with BSO) was planned. Intraoperatively, an unexpected appendicular mass was identified and excised. Postoperative recovery was uneventful, and histopathological examination revealed features consistent with PMP. The patient was referred to oncology for further evaluation and long-term follow-up.

This case highlights the incidental diagnosis of PMP during gynecological surgery, underscoring the importance of intraoperative vigilance and multidisciplinary coordination. Although PMP often presents with non-specific symptoms and is typically diagnosed late, timely recognition and appropriate surgical management are essential for optimal outcomes. Cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (HIPEC) remains the cornerstone of treatment. Increased clinical awareness is vital, especially when encountering unexpected intra-abdominal masses.

Clinical Features and Diagnostic Approach to Pseudomyxoma Peritonei

Pseudomyxoma peritonei (PMP) is a slow-growing yet progressively debilitating condition characterized by the accumulation of mucinous material within the peritoneal cavity. The disease often develops insidiously over several months

or even years, with early symptoms that are vague and nonspecific. Common initial complaints include mild abdominal discomfort, bloating, and a gradual increase in abdominal girth. As mucinous ascites continue to accumulate, patients may experience altered bowel habits—such as constipation or signs of partial intestinal obstruction—due to the compression and encasement of bowel loops.

In advanced stages, symptoms may become more severe and include significant abdominal pain, weight loss, malnutrition, early satiety, and reduced gastrointestinal absorption. The disease can exert pressure on adjacent organs, contributing to urinary symptoms and even respiratory compromise in severe cases. Despite these effects, PMP rarely metastasizes beyond the peritoneal cavity, which often leads to a delay in diagnosis.

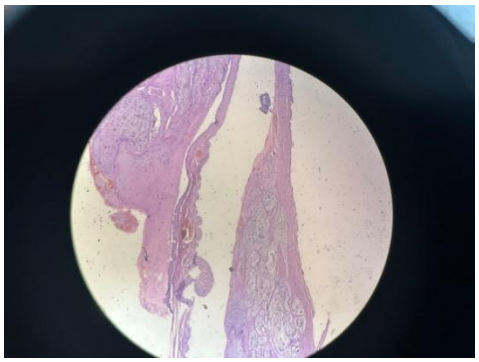
Accurate diagnosis of PMP requires a high index of clinical suspicion, especially when patients present with unexplained abdominal distension, persistent gastrointestinal symptoms, or adnexal masses. Imaging studies, particularly contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI), play a vital role in detecting mucinous ascites and assessing the extent of peritoneal involvement. These modalities may reveal characteristic features such as low-attenuation mucinous fluid, scalloping of the liver and spleen surfaces, and peritoneal implants.

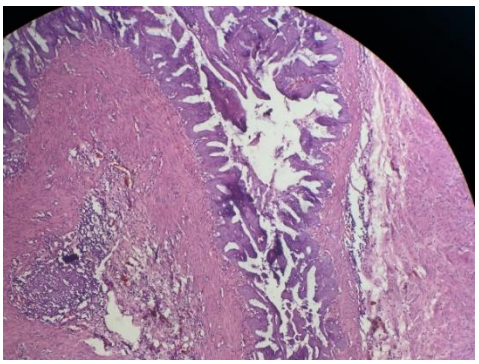
In many cases, definitive diagnosis is achieved intraoperatively during laparotomy or laparoscopy, which allows for direct visualization of mucinous deposits. Surgical exploration not only facilitates diagnosis but also provides the opportunity for tissue biopsy. Histopathological examination confirms PMP by identifying mucin-producing epithelial cells, usually of appendiceal origin, though occasionally from other gastrointestinal or ovarian sources.

Furthermore, tumor markers such as carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA 19-9), and cancer antigen 125 (CA-125) are frequently elevated in PMP and can support the clinical and radiological findings. While not diagnostic alone, these markers are useful for baseline assessment and postoperative monitoring of disease progression or recurrence.

Given its indolent progression and non-specific presentation, early detection of PMP remains a clinical challenge. However, timely diagnosis is crucial, as early and aggressive management—including cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC)—offers the best outcomes in terms of long-term survival and quality of life.

Histopathology Slides





2. DIFFERENTIAL DIAGNOSIS

Pseudomyxoma peritonei (PMP) can closely resemble several other intra-abdominal conditions, making accurate diagnosis challenging without thorough evaluation. The clinical presentation of progressive abdominal distension, ascites, and peritoneal thickening is not unique to PMP and may overlap with other pathologies. Hence, a broad differential diagnosis should be considered, especially in the early stages or in cases of incidental findings.

1. Peritoneal carcinomatosis: This condition, often secondary to gastrointestinal or gynecological malignancies (e.g., gastric, colorectal, or ovarian cancer), presents with peritoneal nodules and ascites. Unlike PMP, peritoneal carcinomatosis is typically associated with systemic metastases and may show more aggressive features on imaging.

2. Peritoneal mesothelioma: A rare malignancy originating from the peritoneal lining, mesothelioma can mimic PMP radiologically and clinically. It is usually linked to asbestos exposure and may show diffuse peritoneal thickening without the mucinous ascites characteristic of PMP.

3. Ovarian cysts or tumors: Complex ovarian neoplasms, especially mucinous cystadenomas or cystadenocarcinomas, can produce similar symptoms and radiological findings. However, PMP often involves the ovary secondarily, making histopathological correlation essential to determine the primary site.

4. Tuberculous peritonitis: Particularly in regions where tuberculosis is endemic, peritoneal TB must be considered. It may present with ascites, peritoneal thickening, and systemic symptoms like fever and weight loss. Diagnostic laparoscopy and

culture or PCR for *Mycobacterium tuberculosis* aid in differentiation.

5. Ascites from hepatic or cardiac origin: Ascites due to liver cirrhosis or congestive heart failure generally lacks the solid peritoneal deposits or mucinous material seen in PMP. Clinical history, liver function tests, and echocardiography help rule out these causes.

6. Mesenteric or omental cysts: These benign cystic lesions may lead to abdominal distension and mimic localized mucin accumulation. Imaging typically shows well-circumscribed cysts, and surgical excision confirms the diagnosis.

Ultimately, **histopathological examination** remains the gold standard for distinguishing PMP from these entities. Identification of mucin-secreting epithelial cells, often of appendiceal origin, and the characteristic mucin pools are diagnostic of PMP. Additionally, immunohistochemistry may aid in differentiating primary appendiceal neoplasms from metastatic tumors involving the peritoneum.

Histopathological Confirmation

Microscopic examination of pseudomyxoma peritonei (PMP) typically shows abundant extracellular mucin accompanied by scattered clusters or strips of mucin-secreting epithelial cells displaying low-grade cytological atypia. A hallmark feature of PMP is the limited invasion into adjacent tissues, distinguishing it from more aggressive malignancies. Immunohistochemical staining can assist in identifying the primary site of the tumor, thereby supporting a more precise and definitive diagnosis.

Treatment of Pseudomyxoma Peritonei

The cornerstone of treatment for pseudomyxoma peritonei (PMP) is cytoreductive surgery (CRS), a complex and meticulous procedure aimed at removing all visible tumor tissue and mucinous material from the peritoneal cavity. This surgical approach may involve extensive peritoneal stripping—the removal of the peritoneal lining—and in many cases, resection of affected organs, such as portions of the colon, spleen, ovaries, uterus, or even parts of the stomach or liver, depending on the extent of disease involvement.

Following CRS, patients typically undergo hyperthermic intraperitoneal chemotherapy (HIPEC). During HIPEC, heated chemotherapeutic agents are circulated within the abdominal cavity for a designated period, usually 60 to 90 minutes, immediately after tumor resection. The heat enhances the effectiveness of the chemotherapy and improves drug penetration, while direct administration within the peritoneal cavity allows for higher local concentrations with minimal systemic toxicity. HIPEC is specifically designed to eradicate residual microscopic disease and significantly reduces the risk of recurrence.

PMP management necessitates a multidisciplinary approach. In addition to specialized surgical oncologists with expertise in peritoneal surface malignancies, care teams often include medical oncologists, anesthesiologists, pathologists, radiologists, intensive care specialists, and supportive care providers. Preoperative optimization and postoperative care are critical and may include nutritional support, pain management, infection prevention, and monitoring for complications such as fistula formation, hemorrhage, or delayed wound healing.

Long-term follow-up is essential, as PMP can recur even after aggressive treatment. Surveillance typically involves periodic imaging and tumor marker monitoring (e.g., CEA, CA-125, CA 19-9).

3. DISCUSSION

The case of Mrs. K underscores the diagnostic challenges of pseudomyxoma peritonei (PMP), particularly when presenting with atypical features such as abnormal uterine bleeding, which is usually evaluated within gynecological practice. This atypical presentation often diverts clinical attention away from abdominal or gastrointestinal pathologies. A similar diagnostic dilemma has been described by Albright et al., who reported PMP presenting as an adnexal mass, initially suspected to be of ovarian origin [1].

In our case, the incidental intraoperative discovery of an appendicular mass highlights the importance of a high index of suspicion and the need for thorough preoperative evaluation. This correlates with the findings of Smeenk et al., who emphasized the appendix as the most common primary site in PMP, particularly in women undergoing gynecological procedures [2]. Esquivel and Sugarbaker also observed that the non-specific and slow-growing nature of PMP often leads to late diagnoses, as patients typically present with vague abdominal symptoms [3].

The literature consistently supports that PMP may be misdiagnosed preoperatively as ovarian or uterine pathology. Pranesh and Jayson discussed the risk of misinterpretation in radiological and clinical assessments, especially in women, resulting in delayed appropriate intervention [4]. In a comprehensive histopathological study, Ronnett et al. highlighted the importance of identifying disseminated peritoneal adenomucinosis (DPAM), the most common histological subtype of PMP, which shows minimal cytologic atypia and indolent behavior [5].

The current gold standard of PMP treatment involves cytoreductive surgery (CRS) followed by hyperthermic intraperitoneal chemotherapy (HIPEC). This combined approach was pioneered by Sugarbaker, who demonstrated a dramatic improvement in long-term survival, reporting 5-year survival rates exceeding 70% in carefully selected patients [6]. Chua et al. further

validated these findings in a large multicentric study showing that patients receiving complete cytoreduction and HIPEC had significantly better disease-free and overall survival [7].

However, CRS-HIPEC is a complex and high-risk intervention. Elias et al. emphasized the importance of patient selection, noting that complete cytoreduction is a key predictor of favorable outcomes, while incomplete cytoreduction correlates with early recurrence and poor survival [8]. In a comparative study, Yan et al. found that morbidity and mortality rates are acceptable when these procedures are performed in experienced centers with multidisciplinary support [9].

One ongoing challenge in PMP management is recurrence prevention and long-term surveillance. Baratti et al. explored the utility of tumor markers such as CEA, CA-125, and CA 19-9 as potential indicators of recurrence and treatment response, though their sensitivity and specificity remain limited [10]. Klemm et al. proposed that serial imaging combined with biomarker monitoring improves early detection of recurrence and guides secondary surgical interventions [11].

Looking to the future, Carr et al. advocated for the development of improved molecular and genetic biomarkers that could facilitate earlier diagnosis and better risk stratification. They also highlighted the need for refining treatment protocols to balance efficacy and morbidity, particularly in low-grade PMP cases [12].

In conclusion, Mrs. K's case reinforces the value of a multidisciplinary approach involving gynecology, oncology, pathology, and surgical expertise. Raising awareness about atypical presentations of PMP, particularly among gynecologists and general surgeons, could lead to earlier diagnosis and improved patient outcomes. Future research should continue to focus on enhancing early diagnostic tools and optimizing therapeutic strategies to improve the quality of life and survival for patients with this rare disease.

4. CONCLUSION

Pseudomyxoma peritonei is a rare but serious condition that requires prompt diagnosis and specialized management. Histopathological confirmation is vital in distinguishing PMP from other peritoneal disorders. The combination of cytoreductive surgery and HIPEC remains the most effective treatment strategy, significantly enhancing survival rates and quality of life. Given the complexity of PMP, a multidisciplinary approach and continued research are essential to improve treatment options and patient outcomes.

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