North Clin Istanb 2024;11(3):261–268
doi: 10.14744/nci.2023.50374

REVIEW GENERAL SURGERY

Pseudomyxoma peritonei: The struggle of a lifetime and the hope of a cure - a rare diagnosis with review of the literature

Ahmet Askar, Asli Arpat, Vedat Durgun

Department of General Surgery, Istanbul University Cerrahpasa-Cerrahpasa Faculty of Medicine, Istanbul, Turkiye

ABSTRACT

Pseudomyxoma peritonei is a rare pathological condition characterized by mucinous tumor tissue implants on the peritoneal surface. Although the cause of Pseudomyxoma peritonei has been extensively studied, the prevailing agreement is that it stems from mucinous tumors that occur in the ovaries or appendix. The tumor tissue typically remains localized to the peritoneum and does not exhibit extraperitoneal spread. Patients with Pseudomyxoma peritonei may present with symptoms such as abdominal pain, bloating, loss of appetite, and shortness of breath. Computerized Tomography is commonly used for diagnostic purposes. The treatment of Pseudomyxoma peritonei typically involves surgical evacuation of the tumoral tissue, followed by cytoreduction and Hyperthermic Intraperitoneal Chemotherapy. While effective treatment options are available, some patients may require repeated surgeries over an extended period. This paper reports on a case study of a patient with a history of recurrent Pseudomyxoma peritonei, necessitating multiple surgical interventions over a decade. The paper concludes with a review of the relevant literature.

Keywords: HIPEC; peritoneum; PMP; pseudomyxoma peritonei.

Cite this article as: *Askar A, Arpat A, Durgun V. Pseudomyxoma peritonei: The struggle of a lifetime and the hope of a cure - a rare diagnosis with review of the literature. North Clin Istanb 2024;11(3):261–268.*

Pseudomyxoma peritonei (PMP) was initially described by Karl F Rokitansky in 1842 [1]. Subsequently, in 1884, Werth classified PMPs as ovarian-associated mucinous carcinomas [2], while Frankel, in 1901, established the link between PMPs and ruptured appendiceal mucoceles [3]. Current research indicates that PMPs are typically low-grade tumors that originate from the appendix, characterized by the production of copious amounts of mucinous tumor tissue [4]. The mucinous material consists of an acid mucopolysaccharide, known as Pseudomucin, which may be cell-free or may contain benign or malignant cells [5].

The abundant production of mucin by PMPs results in marked irritation of the peritoneum, leading to the formation of peritoneal exudates and adhesions, which can cause abdominal pain and even intestinal obstruction. Most patients affected by PMPs are females, with abdominal distention being the most common symptom [6]. Computerized Tomography (CT) has been established as the gold standard for the diagnosis of PMP [7]. Despite numerous attempts to develop a standard treatment for PMPs, paracentesis or aspiration of the tumors has proven unsuccessful due to the highly viscous nature of the mucin [8]. Surgery remains the primary treatment for PMPs, involving the careful removal of the primary focus and evacuation of the mucinous contents [9]. Despite the high recurrence rate, modern treatment options such as cytoreduction surgery and Hyperther-

Received: April 18, 2023 *Accepted:* June 02, 2023 *Online:* June 28, 2024

Correspondence: Ahmet ASKAR, MD. Istanbul Universitesi Cerrahpasa-Cerrahpasa Tip Fakultesi, Genel Cerrahi Anabilim Dali, Istanbul, Turkiye.
Tel: +90 536 408 63 61 e-mail: achkar.dr@yahoo.com

Istanbul Provincial Directorate of Health - Available online at www.northclinist.com

mic Intraperitoneal Chemotherapy (HIPEC) have improved survival rates, with some patients remaining recurrence-free for several years [10]. The 5-year survival rate for PMPs ranges from 6.7% to 37.6%, depending on the subtype of the disease [11].

CASE PRESENTATION

An 81-year-old woman with no significant medical history except for well-controlled hypertension presented with abdominal discomfort and was diagnosed with bilateral ovarian cystic lesions at another clinic. Due to suspicion of ovarian cancer, she underwent total abdominal hysterectomy and bilateral salpingo-oophorectomy. Upon pathological examination following her initial surgery in 2005, the patient was found to have bilateral mucinous cystic tumors and PMP in the paratubal area. The source of the PMP was suspected to be a mucocele in the appendix that had spread over time to involve both ovaries. However, the patient was discharged without any further intervention or investigation. Four years later, the patient returned to our outpatient clinic with recurring symptoms. A CT scan revealed a 6 cm cystic mass in the right lower quadrant of the abdomen, along with fluid collections in the right paracolic area and pelvis. Notably, the appendix was not identifiable on imaging at that time (Fig. 1A, B).

Based on the patient's medical history, an exploratory laparotomy was immediately performed. The surgery revealed diffuse gelatinous fluid with tumoral implants filling the abdominal and pelvic cavity. The intention was to identify and remove the appendix or perform a right hemicolectomy; however, severe adhesions in the abdomen restricted access to the right lower quadrant. Due to the patient's age, general health status, and to avoid any intestinal injury, the operation was terminated without resection. The postoperative period was uneventful. A PET/CT scan was obtained during the hospital stay to detect any possible distant metastatic lesions, but no hypermetabolic lesions were observed. The low FDG affinity of tumors made it difficult for the PET/CT scan to identify metastatic status. Upon examination of the excised specimen, mucinous lakes containing malignant epithelial cells were observed, some of which had a signet ring cell appearance (Fig. 2A–D). These findings were consistent with low-grade mucinous neoplasm and led to a pathological diagnosis of PMP.

In 2012, the patient presented to our clinic with an incisional hernia and underwent surgery for hernia repair. During the procedure, the abdominal cavity was found to

Highlight key points

- Pseudomyxoma peritonei is a rare and difficult to treat disease so optimal management of it demands a multidisciplinary approach.
- Hyperthermic Intraperitoneal Chemotherapy therapy has emerged as a viable alternative for the management of advanced intra-abdominal malignancies, peritoneal mesothelioma, and pseudomyxoma peritonei.
- It is crucial to explore alternative treatment modalities, such as laparoscopic evacuation of povidone-iodine washings, which can deliver favorable outcomes for patients who are not viable candidates for Hyperthermic Intraperitoneal Chemotherapy treatment or decline this intervention due to financial limitations.

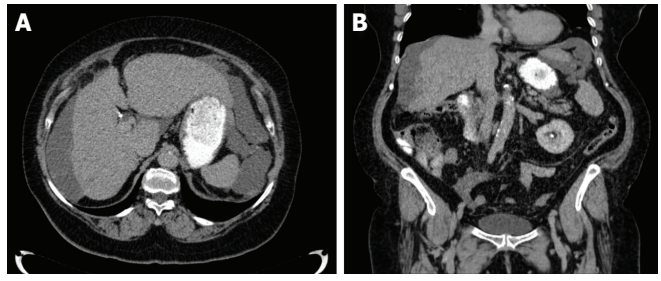

Figure 1. **(A)** Axial CT scan view of the abdomen, with the cystic masses in the right perihepatic and left paracolic area. **(B)** Coronal CT scan view of the abdomen, with the cystic mass located at the subdiaphragmatic area compresses the liver.

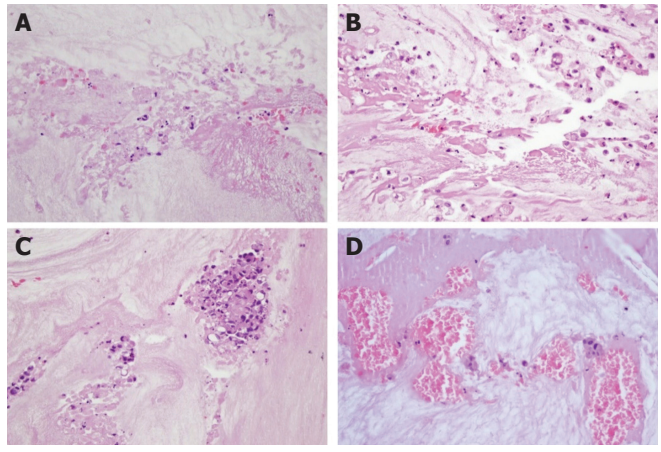

Figure 2. Microscopic examination using hematoxylin and eosin staining (X400); Malignant epithelial cells within mucinous lakes, some of these cells have an atypical morphology as signet ring cells. The pathology confirmed low-grade mucinous neoplasm.

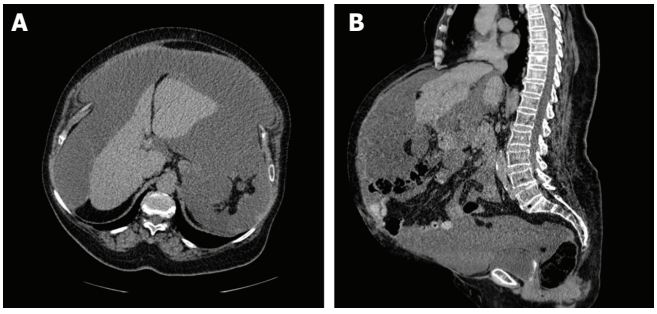

FIGURE 3. **(A, B)** The axial and sagittal CT scan views of the abdomen, reveal large amounts of mucinous contents and scalloping in the liver.

be filled with mucinous fluid, which was evacuated, and the hernia defect was repaired with a Prolene mesh. The patient had an uneventful recovery and was discharged. In 2015, the patient returned to the emergency department with symptoms of abdominal distension, nausea, and vomiting, and was diagnosed with an incarcerated umbilical hernia. During surgery, the abdominal cavity was again filled with mucinous fluid, which was evacuated, and the necrotic umbilicus was excised and repaired with an inlay Prolene mesh. In 2018, the patient presented to our outpatient clinic with symptoms of abdominal distension, discomfort, and shortness of breath. The CT scan revealed a large volume of mucinous contents and scalloping in the liver (Fig. 3A, B). The patient underwent a surgical procedure in which the large amount of gelatinous fluid in the abdomen and pelvic cavity was drained, with approximately 15 liters of fluid being removed. She was discharged without any notable postoperative complications. In 2020, the patient presented with similar symptoms and only underwent mucin evacuation. In 2021, she was admitted to the outpatient clinic with tenderness in the umbilical region, where the umbilicus was discolored and showed signs of necrosis, along with fecaloid discharge. The patient then underwent exploratory laparotomy, during the exploration, a significant amount of mucinous fluid was observed filling the abdomen and pelvis. The amount of fluid evacuated was close to 20 liters (Fig. 4A–D). Additionally, during the exploration, it was observed that the sigmoid colon was adherent to the Prolene mesh with a small fistula formation to the abdominal wall, a segmental colon resection and end-to-end anastomosis were performed. Following an uneventful postoperative recovery, the patient was discharged and followed-up in our outpatient clinic. A timeline of the patient's medical history is presented in (Fig. 5).

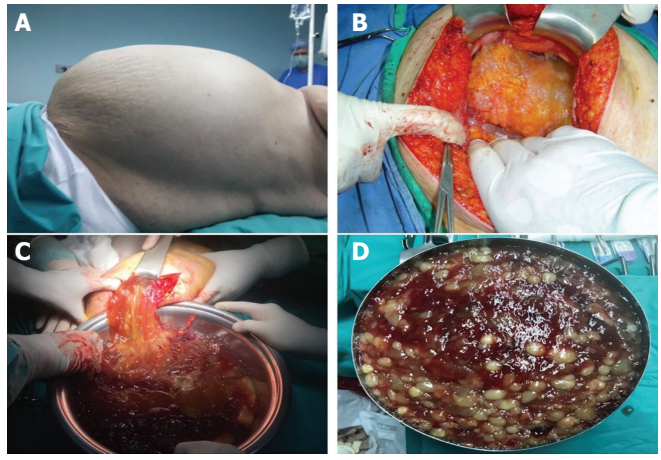

FIGURE 4. **(A)** Preoperative distended abdomen. **(B)** diffuse of gelatinous fluid with tumoral implants that filled the abdominal and pelvic cavity, **(C)** Evacuation of mucinous material from the abdominal cavity. **(D)** Large volume of mucinous material evacuated from the abdomen.

DISCUSSION

The incidence of PMP remains unclear; however, it is estimated to be around one or two cases per million per year based on various studies [12]. This rare disease is characterized by an excessive production of mucin in the abdominal and pelvic cavities. Despite continuous research efforts, the etiology and pathogenesis of PMP are still not fully understood [13]. It is commonly accepted that cystadenomas, which produce mucus and remain dormant in their primary site, may rupture eventually due to increasing distension and intra-tumoral pressure. Such an occurrence could promote the spread of malignant cells within the peritoneal cavity and surrounding anatomical structures. An important feature of PMP is the high proportion of mucin to tumoral cells, with a Mucus:Tumor cell ratio of 10:1 [14]. Metastasis of PMPs to distant sites is exceedingly uncommon, as evidenced by several studies [2, 11, 15]. The origin of PMPs is still a subject of debate among researchers, with some proposing the ovaries as the site of origin, while others argue for the appendix vermiformis. Ronnett et al. [11] conducted a study with 30 female patients diagnosed with PMP and concluded that the appendix or intestinal mucinous tumors were the primary source of PMP. Although PMPs are typically classified as mucinous neoplasms of the appendix [11, 13, 16], there are reports in the literature of PMPs originating from other organs, including the ovaries, colon, and pancreas [2, 5, 9, 11, 13, 14, 16, 17].

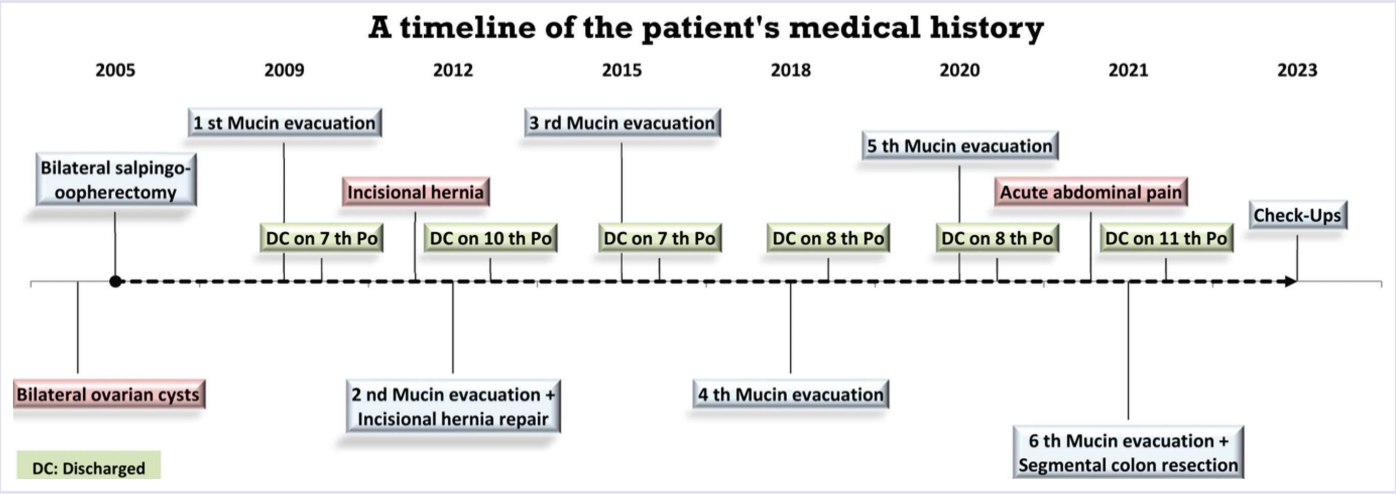

FIGURE 5. Timeline diagram outlining patient's clinical course over 13 years period.

The term PMP encompasses a heterogeneous group of lesions with varying behaviors, including benign, borderline, and malignant lesions. Therefore, defining a lesion solely as PMP can be challenging and imprecise. To address this issue, Ronnett et al. [11] classified PMPs into two diagnostic categories: Disseminated Peritoneal Adenomucinosis (DPAM) and Peritoneal Mucinous Carcinoma (PMCA). DPAMs are peritoneal lesions that contain fewer cells and more extracellular mucin, with focal mucinous epithelium. Tumoral cells have low mitotic activity and cellular atypia. In contrast, PMCAs are peritoneal lesions with more epithelial cells within the mucin, with tumoral cells showing features and structures more typical of carcinoma and may or may not be associated with adenocarcinoma.

In 2010, the WHO and AJCC classified PMPs as low, or high-grade lesions based on histologic, molecular, and cytological features [18]. To make a definitive diagnosis of PMP, the presence of mucinous neoplastic cells and epithelial glandular cells should be demonstrated histologically in the pathologic specimen, and diffuse mucinous material and implants in the abdomen should also be present clinically. If epithelial cells are absent, the term "mucinous ascites" may be more appropriate [13, 19]. Mucin is produced by appendiceal goblet cells and has tight junctions with the surrounding stroma. Studies have shown that goblet cells of PMP patients selectively express the MUC-2 gene, which is considered a specific marker of appendicular PMP. This finding supports the theory that PMPs originate from the appendix rather than the ovaries. O'Connell et al. [1] have reported these findings.

PMP typically affects female patients, and its most distinctive clinical manifestation is abdominal distension and discomfort, which can give rise to a "Jelly Belly" appearance characterized by diffuse abdominal swelling. In some cases, milder forms of the disease may present with abdominal pain without accompanying abdominal swelling, which can potentially be mistaken for acute appendicitis [12, 14, 16, 19]. Due to increased intra-abdominal pressure, PMP may sometimes present as a newly developed hernia or uterine prolapse, and some patients may be incidentally diagnosed with PMP during surgery for these aforementioned reasons. PMP can also present as a palpable pelvic mass, which can be mistaken for a tubo-ovarian abscess or malignancy [2, 19, 20].

Abdominal CT scan is considered the gold standard diagnostic tool for PMP. The characteristic finding of PMP on CT scan is intraabdominal and pelvic low attenuation diffuse mucinous ascites. The Scalloping sign, which is a distinctive feature of PMP on a CT scan, refers to an indented appearance on the liver surface resulting from the compression of a large volume of mucinous ascites against the liver capsule. This sign helps differentiate between PMP and ascites. Other potential findings on a CT scan include calcifications and septations [14, 19, 20].

Sulkin et al. [20] conducted a study and reported that in cases of large-volume PMP tumors, the appendix was not visible on CT scans. Additionally, in patients with small PMP tumor volumes, only a few individuals had a visible appendix. The researchers noted a correlation between the CT scan findings and the observations made during surgery. Similarly, in the present case, neither CT scan nor laparotomy could visualize the appendix.

PMP tumors and mucin are influenced by various factors, such as the flow and absorption of peritoneal fluid and gravity. Consequently, tumoral cells and mucin accumulate significantly in some regions of the abdominal cavity, such as the omentum (resulting in an omental cake appearance), the right hemidiaphragm, the right retrohepatic area, the left paracolic region, and the pelvis. Conversely, they rarely localize in other areas [4, 9, 11, 14, 16, 20].

The diagnostic utility of serum tumor markers such as CA125, CA 19-9, and CEA in PMP is limited. However, they are helpful in follow-up and can provide insight into disease recurrence [14, 12, 21]. Prior to the emergence and widespread adoption of Hyperthermic Intraperitoneal Chemotherapy (HIPEC), the treatment options for PMP were limited to repetitive debulking surgeries. The introduction of HIPEC as a treatment modality in the field of surgical oncology has revolutionized the treatment protocols for various malignancies, including PMP. Consequently, HIPEC has become a new treatment option for PMP [2, 9, 14, 17, 22]. Sugarbaker and Chang [4] conducted a study with 385 patients diagnosed with appendiceal malignancies that had spread to the peritoneal surfaces. The researchers combined cytoreductive surgery with HIPEC and concluded that the 5-year survival rate for patients with adenomucinosis could reach up to 86% with the use of HIPEC. The desired cytoreductive surgery comprises parietal and visceral peritonectomy, along with intraoperative HIPEC. While the procedure appears promising, the average operative time of 10,5 hours for radical surgical resection is associated with higher rates of morbidity and mortality [12]. The use of systemic chemotherapy alone is not recommended for treating PMP due to its relatively poor circulation and the excessive presence of mucin, which hinders the penetration of chemotherapeutic agents to reach therapeutic levels. Based on a study by Kojimahara et al. [22], systemic chemotherapy does not affect the survival rates in PMP patients, while intraperitoneal administration of chemotherapy has a favorable effect on survival rates. Therefore, systemic chemotherapy may only be considered as a treatment option when no other alternative treatments are suitable for the patient [2, 9, 14, 19, 21, 22]. Systemic chemotherapy may be considered as a treatment modality in cases where alternative treatment options are not suitable for the patient [9, 21]. In cases where patients with PMP exhibit medical comorbidities and limited performance status that preclude aggressive cytoreductive surgery and HIPEC, or in resource-limited settings where access to such sophisticated treatments is restricted, repetitive surgical debulking for palliative purposes may represent a viable treatment option.

Kelly et al. [8] conducted a study on patients diagnosed with recurrent PMP disease and found that percutaneous drainage to alleviate patient symptoms was not beneficial. Repeated laparotomies for debulking surgery may pose a challenge to patients and require a recovery period of 4–6 weeks. The authors proposed laparoscopic mucin evacuation as a minimally invasive alternative method. However, in this particular case, we encountered obstacles to laparoscopic mucin evacuation due to the patient's advanced age, extensive adhesions and fibrosis resulting from prior abdominal surgeries, and the potential for iatrogenic bowel injury. Consequently, we opted to perform open debulking surgery multiple times to relieve the patient's symptoms. It is important to note that repeated debulking surgeries have been linked to high rates of morbidity and mortality [9, 22]. In patients with an Eastern Cooperative Oncology Group (ECOG) performance score of 2–3 and 0–1 who received HIPEC, the average overall survival was 9.5 and 21.7, respectively [10, 19]. In the current case, the patient's Eastern Cooperative Oncology Group (ECOG) score was 3, and as a result, HIPEC was not utilized due to concerns regarding patient safety.

Wheeler et al. [23] reported that in patients with DPAM, cytoreductive surgery without intraperitoneal chemotherapy leads to a poorer prognosis, but does not affect patient survival. They contend that combining cytoreductive surgery with intraperitoneal chemotherapy in DPAM patients could add to the burden of treatment, and that cytoreductive surgery alone, without intraperitoneal chemotherapy, may be more advantageous for patients with pseudomyxoma peritonei with PMCA.

Prognosis in PMP is significantly influenced by factors such as tumor size, success of cytoreductive therapy, and degree of neoplastic cell differentiation. Regrettably, despite appropriate treatment, PMP patients with high preoperative tumor marker levels tend to experience recurrence at some point in the future [12, 13, 21]. Elevated levels of tumor markers such as CEA, CA19-9, and CA 125 during follow-up are directly correlated with PMP recurrence. Conversely, patients with low tumor marker levels during follow-up tend to have better prognoses [12, 13, 14, 19, 21]. While the 5-year and 10-year survival rates for PMP patients treated with classical debulking surgery are 53–75% and 32–60%, respectively, an aggressive cytoreduction combined with intraoperative chemotherapy can increase the 10-year survival rate to 90% [14].

Table 1. Brief summary of the clinical response for patients diagnosed with PMP who underwent surgical intervention and received various chemotherapeutic treatments

Author	Year	Age	Sex	Procedure	Chemotherapy	Outcome	Origin	Location	Histology	CEA	CA19-9	CA125
de Oliveira et al. [19]	2014	76	F	DS	N/A	Recurrence at 2 yr.+8 mo.	Appendix/ Intestinal	Diffuse	Mucinous neoplastic cells	177	<2.5	57.5
Wrafter et al. [6]	2015	52	F	DS	N/A	No recurrance within 2 yrs.	Appendix	Right paracolic	LAMN	6.4	–	27.3
Liang et al. [17]	2017	44	M	CRS	Perioperative chemotherapy	No recurance within 7 mo.	Urachal	Pelvis	Urachal mucinous adeno-carcinoma with HAMN	25.12	73.7	–
Pantiora et al. [7]	2018	59	M	CRS	HIPEC with Oxaliplatin, 5-FU and Leukovorin	No recurrance within 2 years	Appendix/ Intestinal	Diffuse	LAMN	210	NL	–
		62	M	CRS	HIPEC with Oxaliplatin, 5-FU and Leukoverin	No recurrance within 2 yrs.	Appendix/ Intestinal	Right paracolic	HAMN	NL	NL	–
García et al. [18]	2019	54	M	CRS	HIPEC with Mitomycin C	No recurance within 7 mo.	Appendix	Perisplenic and peripancreatic	Low grade mucinous adenocarcinoma	17.8	–	–
Huang et al. [25]	2019	63	F	CRS	HIPEC with Mitomycin C and 5-FU + Apatinib	Recurrence within 1 yr.	Ovary	Perihepatic	LAMN	138	99.6	133
Azzam et al. [26]	2020	62	M	CRS	HIPEC with Mitomycin C + IORT	Recurrence within 2 yrs.	Appendix	RLQ	Mucinous adenocarcinoma	8.9	NL	NL
Padma-nabhan et al. [27]	2020	72	F	CRS	DS + NIPT with Cisplatin, Docataxel + HIPEC with Oxaliplatin and 5-FU	–	Appendix	RLQ and pelvis	HAMN	7.1	453	73.8
Hirano et al. [28]	2021	62	M	DS	N/A + Adjuvant systemic Bevacizumab and TAS-102	Remission of the disease	Unknown	Diffuse	HAMN	100–200	–	–
Yazawa et al. [15]	2023	26	F	Staging laparotomy	Intraperitoneal chemotherapy	Remission of the disease	Ovary	Left paraopvarian	Mucinous borderline ovarian tumor	16.6	171	55.7
		73	F	N/A	N/A	Recurrence within	Appendix	Left paraovarian	LAMN	15.2	2.2	55.5
		82	F	N/A	N/A	Palliative care	Appendix	Diffuse	LAMN	40.1	10.4	141.1
		48	F	CRS	HIPEC with Mitomycin C	Follow ups	Appendix	Diffuse	LAMN	21.8	3.7	84.4
		81	F	DS	N/A	Remission of the disease	Appendix	Diffuse	LAMN	59	102	25
Our case	2023											

F: Female; M: Male; HIPEC: Hyperthermic intraperitoneal chemotherapy; CRS: Cytoreductive surgery; DS: Debulking surgery; NIPT: Neoadjuvant intra-peritoneal chemotherapy; IORT: Intra-operative radiotherapy; LAMN: Low grade appendiceal mucinous neoplasm; HAMN: High grade appendiceal mucinous neoplasm; RLQ: Right lower quadrant; NL: Normal; N/A: Not applicable.

Fiorelli et al. [24] demonstrated the antitumor activity of povidone-iodine (commonly known as Betadine®) on mesothelioma cell lines in their experimental study. While povidone-iodine is typically used at a 10% concentration for its antiseptic properties, a concentration of 0.1% was recommended for its antitumor effects. We propose that patients who are not suitable candidates for aggressive cytoreductive surgery and HIPEC may undergo debulking surgery with the addition of povidone-iodine washing, with the hope that the antitumor effects may be of benefit to the patient.

In Table 1, we provide a literature review of the clinical responses of PMP patients who underwent surgical intervention and received various chemotherapeutic treatments over the past decade [6, 7, 15, 17–19, 25–28]

Conclusion

PMP is a rare disease with a relatively mild clinical course, which can make it difficult to diagnose. It is believed to originate from primary ovarian or appendiceal mucinous tumors. Despite numerous attempts to develop an optimal treatment protocol, many modalities have failed to achieve satisfactory results. Recurrence is a common issue in PMP and can be troublesome for both patients and clinicians. HIPEC, a recently introduced treatment modality, has shown promise in PMP management by achieving low recurrence rates and prolonged survival rates. However, it is limited by factors such as prolonged operative time, high costs, and high rates of morbidity and mortality, which limit its routine use. In cases such as the one presented, where patients with recurrent PMP disease are elderly, have high morbidity, or reside in developing countries with limited access to advanced treatment modalities, repetitive laparoscopic or open debulking surgery followed by betadine washing may be a viable option. While HIPEC is considered the standard treatment for PMP, its use in elderly patients or those with specific medical conditions may still be controversial. Although HIPEC has a promising effect, there remains a need for increased experience, reduced costs, and more time to improve its universal applicability.

Acknowledgements: We would like to thank to Dr. Sefa Ergun for his invaluable contributions to this project; Dr. Ayse Hanim Yavuz for providing the pathologic images and confirming the diagnosis, which were crucial to the success of this work; Dr. Sirin Askar for her insightful feedback, critical analysis, and unwavering support throughout the entire project.

Authorship Contributions: Concept – AAS, AAr, VD; Design – AAS, AAr; Supervision – AAS, VD; Fundings – AAS, AAr; Materials – AAS, AAr; Data collection and/or processing – AAS, AAr; Analysis and/or interpretation – AAS, AAr, VD; Literature review – AAS, AAr; Writing – AAS; Critical review – AAS.

Conflict of Interest: No conflict of interest was declared by the authors.

Use of AI for Writing Assistance: AI was employed solely for minor editing purposes to ensure clarity and coherence. The essence and originality of the content remain entirely human-generated.

Financial Disclosure: The authors declared that this study has received no financial support.

Peer-review: Externally peer-reviewed.

REFERENCES

1. O'Connell JT, Tomlinson JS, Roberts AA, McGonigle KF, Barsky SH. Pseudomyxoma peritonei is a disease of MUC2-expressing goblet cells. Am J Pathol 2002;161:551–64. [CrossRef]
2. Gough DB, Donohue JH, Schutt AJ, Gonchoroff N, Goellner JR, Wilson TO, et al. Pseudomyxoma peritonei. Long-term patient survival with an aggressive regional approach. Ann Surg 1994;219:112–9. [CrossRef]
3. Franckel E. Uber das sogennate pseudomyxome peritonei. Med Wochenschr 1901;48:965–70.
4. Sugarbaker PH, Chang D. Results of treatment of 385 patients with peritoneal surface spread of appendiceal malignancy. Ann Surg Oncol 1999;6:727–31. [CrossRef]
5. Morera-Ocon FJ, Navarro-Campoy C. History of pseudomyxoma peritonei from its origin to the first decades of the twenty-first century. World J Gastrointest Surg 2019;11:358–64. [CrossRef]
6. Wrafter PF, Connelly T, Khan JS, Joyce WP. Pseudomyxoma peritonei diagnosed 19 years after appendicectomy. BMJ Case Rep 2015;2015:bcr2015211706. [CrossRef]
7. Pantiora EV, Massaras D, Koutalas J, Melemeni A, Fragulidis GP. Pseudomyxoma peritonei: presentation of two cases and challenging issues in the literature. Cureus 2018;10:e3732. [CrossRef]
8. Kelly KJ, Baumgartner JM, Lowy AM. Laparoscopic evacuation of mucinous ascites for palliation of pseudomyxoma peritonei. Ann Surg Oncol 2015;22:1722–5. [CrossRef]
9. Sugarbaker PH. Pseudomyxoma peritonei. A cancer whose biology is characterized by a redistribution phenomenon. Ann Surg 1994;219:109–11. [CrossRef]
10. Stewart JH 4th, Shen P, Levine EA. Intraperitoneal hyperthermic chemotherapy for peritoneal surface malignancy: current status and future directions. Ann Surg Oncol 2005;12:765–77. [CrossRef]
11. Ronnett BM, Zahn CM, Kurman RJ, Kass ME, Sugarbaker PH, Shmookler BM. Disseminated peritoneal adenomucinosis and peritoneal mucinous carcinomatosis. A clinicopathologic analysis of 109 cases with emphasis on distinguishing pathologic features, site of origin, prognosis, and relationship to "pseudomyxoma peritonei." Am J Surg Pathol 1995;19:1390–408. [CrossRef]
12. Alexander-Sefre F, Chandrakumaran K, Banerjee S, Sexton R, Thomas JM, Moran B. Elevated tumor markers prior to complete tumor removal in patients with pseudomyxoma peritonei predict early recurrence. Colorectal Dis 2005;7:382–6. [CrossRef]
13. Li C, Kanthan R, Kanthan SC. Pseudomyxoma peritonei – a revisit: report of 2 cases and literature review. World J Surg Oncol 2006;4:60. [CrossRef]

14. Smeenk RM, Bruin SC, van Velthuysen ML, Verwaal VJ. Pseudomyxoma peritonei. Curr Probl Surg 2008;45:527–75. [CrossRef]
15. Yazawa R, Yazawa H, Fukuda K, Ohara M, Osuka F. Four cases of pseudomyxoma peritonei with ovarian tumors at our hospital. Fukushima J Med Sci 2023;69:57–65. [CrossRef]
16. Young RH. Pseudomyxoma peritonei and selected other aspects of the spread of appendiceal neoplasms. Semin Diagn Pathol 2004;21:134–50. [CrossRef]
17. Liang L, Zhou N, Xu H, Liu D, Lu Y, Li F, et al. Urachal mucinous adenocarcinoma with pseudomyxoma peritonei: a case report. Medicine (Baltimore) 2017;96:e7548. [CrossRef]
18. García KM, Flores KM, Ruiz A, González FL, Rodríguez ÁM. Pseudomyxoma peritonei: case report and literature review. J Gastrointest Cancer 2019;50:1037–42. [CrossRef]
19. de Oliveira AM, Rodrigues C, Borges A, Martins A, dos Santos S, Pires F, et al. Pseudomyxoma peritonei: a clinical case of this poorly understood condition. Int J Gen Med 2014;7:137-41. [CrossRef]
20. Sulkin TV, O'Neill H, Amin AI, Moran. CT in pseudomyxoma peritonei: a review of 17 cases. Clin Radiol 2002;57:608–13. [CrossRef]
21. Kusamura S, Hutanu I, Baratti D, Deraco M. Circulating tumor markers: predictors of incomplete cytoreduction and powerful determinants of outcome in pseudomyxoma peritonei. J Surg Oncol 2013;108:1–8. [CrossRef]
22. Kojimahara T, Nakahara K, Shoji T, Sugiyama T, Takano T, Yaegashi N, et al. Identifying prognostic factors in Japanese women with pseudomyxoma peritonei: a retrospective clinico-pathological study of the Tohoku Gynecologic Cancer Unit. Tohoku J Exp Med 2011;223:91–6. [CrossRef]
23. Wheeler BR, Reddy SK, Kenwright D, Keating JP. Treating pseudomyxoma peritonei without heated intraperitoneal chemotherapy--a first look in New Zealand. N Z Med J 2014;127:31–9.
24. Fiorelli A, Pentimalli F, D'Urso V, Di Marzo D, Forte IM, Giordano A, et al. Antineoplastic activity of povidone-iodine on different mesothelioma cell lines: results of in vitro study. Eur J Cardiothorac Surg 2014;45:993–1000. [CrossRef]
25. Huang R, Shi XL, Wang YF, Yang F, Wang TT, Peng CX. Apatinib for treatment of a pseudomyxoma peritonei patient after surgical treatment and hyperthermic intraperitoneal chemotherapy: a case report. World J Clin Cases 2019;7:3881–6. [CrossRef]
26. Azzam AZ, Al-Hamadah FH, Azzam KA, Amin TM. Mucinous appendiceal adenocarcinoma penetrated the quadratus lumborum muscle posteriorly and presented as gluteal abscess managed aggressively with drainage followed by cytoreductive surgery and hyperthermic intraperitoneal chemotherapy combined with intraoperative radiotherapy. a case report and review of the literature. Indian J Surg Oncol 2020;11 Suppl 1:128–30. [CrossRef]
27. Padmanabhan N, Ishibashi H, Nishihara K, Sako S, Katayama K, Wakama S, et al. Complete pathological response of high grade appendicular neoplasm induced Pseudomyxoma Peritonei (PMP) after neoadjuvant intra-peritoneal chemotherapy: a case report. Int J Surg Case Rep 2020;72:117–21. [CrossRef]
28. Hirano S, Gohda Y, Miyazaki H, Hayama N, Shimizu S, Igari T, et al. A case of pseudomyxoma peritonei successfully treated with trifluridine/tipiracil (TAS-102) and bevacizumab after palliative debulking surgery. Chin Clin Oncol 2021;10:29. [CrossRef]