Journal of Gastrointestinal Cancer (2019) 50:1037–1042
https://doi.org/10.1007/s12029-018-00192-8

CASE REPORT

Pseudomyxoma Peritonei: Case Report and Literature Review

Keishla M. García[1] · Karla M. Flores[1] · Alejandro Ruiz[1] · Frances L. González[2] · Ángel M. Rodríguez[1,3]

Published online: 8 January 2019

Abbreviations

PMP	Pseudomyxoma peritonei
DPAM	Disseminated peritoneal adenomucinosis
PMCA	Peritoneal mucinous carcinomatosis
CRS	Cytoreduction surgery
HIPEC	Hyperthermic intraperitoneal chemotherapy
CEA	Carcinoembryonic antigen
NCCN	National Comprehensive Cancer Network

Introduction/Background

Pseudomyxoma peritonei (PMP) is a disorder characterized by an intra-abdominal accumulation of mucin secondary to the growth of neoplastic mucin-secreting cells on peritoneal surfaces [1, 2]. PMP is a rare condition with an estimated incidence of one to two cases per million per year [3, 4]. Patients have an average age of 53 years at the time of diagnosis, being more commonly seen in females [5, 6]. In most cases, the neoplasm originates in the appendix and later spreads through the wall of the appendix into the peritoneal space [7, 8]. Reproduction and aggregation of free and implanted tumor cells lead to a progressive peritoneal mucinous tumor and ascites [9]. In other reported cases, the primary tumor origin has occasionally developed in other organs including the ovary, colorectum, gallbladder, stomach, pancreas, urachus, fallopian tube, lung, and breast [10, 11]. Initial presentation varies and consists of unspecified signs and symptoms that relate to the progression of the disease [12]. These include increased abdominal girth, an appendicitis-like syndrome, a new-onset hernia, presence of a pelvic mass, or nonspecific abdominal or pelvic pain [8, 13, 14]. Progressive accumulation of mucinous material gradually fills and can compress vital organs within the peritoneal cavity, which can result in abdominal distention, ascites, bowel obstruction, and nutritional compromise [8, 15, 16].

In 1995, two diagnostic categories were suggested by Ronnett et al. [17], which correlated the prognosis with the histological features (Table 1). These included disseminated peritoneal adenomucinosis (DPAM) and peritoneal mucinous carcinomatosis (PMCA). DPAM consisted of peritoneal lesions composed of abundant extracellular mucin containing scant simple to focally proliferative mucinous epithelium with little cytologic atypia or mitotic activity, with or without an associated appendiceal mucinous adenoma. PMCA was composed of peritoneal lesions containing more abundant mucinous epithelium with the architectural and cytologic features of carcinoma, with or without an associated primary mucinous adenocarcinoma [17].

In 2010, the World Health Organization (WHO) and the American Joint Committee on Cancer (AJCC) also proposed a pathological classification regarding a histological distinction between a low-grade or high-grade lesion (Table 1) [8]. This criterion is based on the histogenesis, molecular and cytological features [8]. Low-grade mucinous adenocarcinoma is characterized by mucin pools with low cellularity (< 10%), bland cytology, and non-stratified cuboidal epithelium [8]. High-grade mucinous adenocarcinoma contains mucin pools with cellularity, moderate/severe cytologic atypia, and signet ring morphology with desmoplastic stroma [8]. Studies correlating the histological classification to the prognosis of the disease determine that those with DPAM have much better outcomes when compared to patients with high-grade PMCA tumors [8, 18].

Treatment has generally consisted of repeating interventions of debulking procedures; however, long-term survival and possibility of cure have had limited expectations [7, 11]. Proposed treatment in the setting of peritoneal involvement consists of complete cytoreduction surgery (CRS) and hyperthermic intraperitoneal chemotherapy (HIPEC) [19, 20]. In this aggressive treatment approach, surgery aims to make a complete macroscopic cytoreduction and lysis of

✉ Ángel M. Rodríguez
rodriguez@cancerdoctorpr.com

[1] Department of Oncology, San Juan Bautista School of Medicine, Caguas, PR, USA

[2] University of Puerto Rico, San Juan, PR, USA

[3] Surgical Department, San Juan Bautista School of Medicine, Caguas, PR, USA

Table 1 Histological classifications of PMP

Classification	Year	Histological classification	Characterization
Ronnett et al. [4]	1995	Disseminated peritoneal adenomucinosis (DPAM)	Peritoneal lesions composed of abundant extracellular mucin containing scant simple to focally proliferative mucinous epithelium with little cytologic atypia or mitotic activity, with or without an associated appendiceal mucinous adenoma.
		Peritoneal mucinous carcinomatosis (PMCA)	PMCA was composed of peritoneal lesions containing more abundant mucinous epithelium with the architectural and cytologic features of carcinoma, with or without an associated primary mucinous adenocarcinoma [4].
AJCC and WHO [8]	2010	Low-grade mucinous adenocarcinoma	Mucin pools with low cellularity (< 10%), bland cytology and non-stratified cuboidal epithelium.
		High-grade mucinous adenocarcinoma	Mucin pools with high cellularity, moderate/severe cytologic atypia, and signet ring morphology with desmoplastic stroma

adhesions, while adjuvant HIPEC works to eradicate any macroscopic or microscopic tumor residue and, in this way, reduce tumor recurrence [21].

Case Presentation

This is the case of a 54-year-old male that presented with early satiety after meals. Patient has a history of hypothyroidism and no other previous medical history conditions. Patient denies tobacco, alcohol, or drug use. Family history is pertinent for unspecified cancer in his father and diabetes mellitus in his mother.

Colonoscopy was performed and was pertinent for diverticulosis and internal hemorrhoids. An esophagogastroduodenoscopy (EGD) performed revealed a small hiatal hernia and a prominent extrinsic compression seen at proximal gastric body. Further evaluation with an abdominopelvic CT scan with contrast showed a very large 15-cm multi-cystic multi-septated mass in the left upper quadrant, infiltrating the distal pancreas and spleen with scattered calcified septations and abundant upper abdominal fluid. Also present was a 3- to 4-cm similar rim calcified cystic lesion in the right paracolic gutter, and likely a related peritoneal implant. Carcinoembryonic antigen (CEA) was 17.8 ng/ml. A distal pancreatectomy and splenectomy were performed. The excision of an abdominal cavity mass revealed mesothelium lined fibrous tissue with extensive mucin deposits, chronic inflammation, and hemosiderin laden macrophages. In addition, a mucous cyst composed of mesothelium lined fibrous tissue with extensive mucin deposits was identified. Pathology resection of the distal pancreas and spleen revealed a low-grade mucinous carcinoma, of at least 14 cm in size. Immunohistochemical stains for CK7 and CK20 show staining for CK20 only. The patient received adjuvant systemic chemotherapy of 5-FU/LV. CEA marker 2 months after surgery was at 16 ng/ml. The patient was followed with serial imaging and tumor markers. Diagnosis of appendiceal adenocarcinoma was not suggested at this time.

Abdominopelvic CT scan performed 17 months after surgery revealed an abnormally dilated appendix, measuring up to 26 mm in thickness (Fig. 1). It had some mural calcifications, suspicious for an appendiceal mucocele. CEA levels at the time were 11.8 ng/ml. A diagnostic laparoscopy was performed. Findings were consistent with mucin deposits without gross evidence of solid organ or peritoneal metastasis. Pathology from peritoneal fluid and peritoneum biopsy revealed mucoid material with atypical cells, consistent with pseudomyxoma peritonei. Immunohistochemical staining was negative for CK20, CDX-2, WT1, and desmin.

A right hemicolectomy, cytoreductive surgery, and HIPEC with mitomycin C were performed. Final pathology revealed primary high-grade mucinous appendiceal adenocarcinoma. Evaluation 6 months after CRS and HIPEC revealed no major postoperative complications. Imaging, laboratory testing, and CEA levels were without evidence of disease following 3 and 7 months after CRS and HIPEC procedure.

Discussion

Pseudomyxoma peritonei (PMP) is a rare and poorly understood disease characterized by disseminated mucinous ascites and peritoneal, serosal, and omental mucinous implants [22]. PMP is difficult to diagnose clinically or by current imaging options due to its variable presentation, course, and nature; nevertheless, CT scan is able to identify the tumor in an early state and is considered the best diagnostic noninvasive modality [23]. Typical appearance on CT includes areas of low attenuation, with islands of higher attenuation due to solid elements within mucinous material. Also, visceral surfaces show

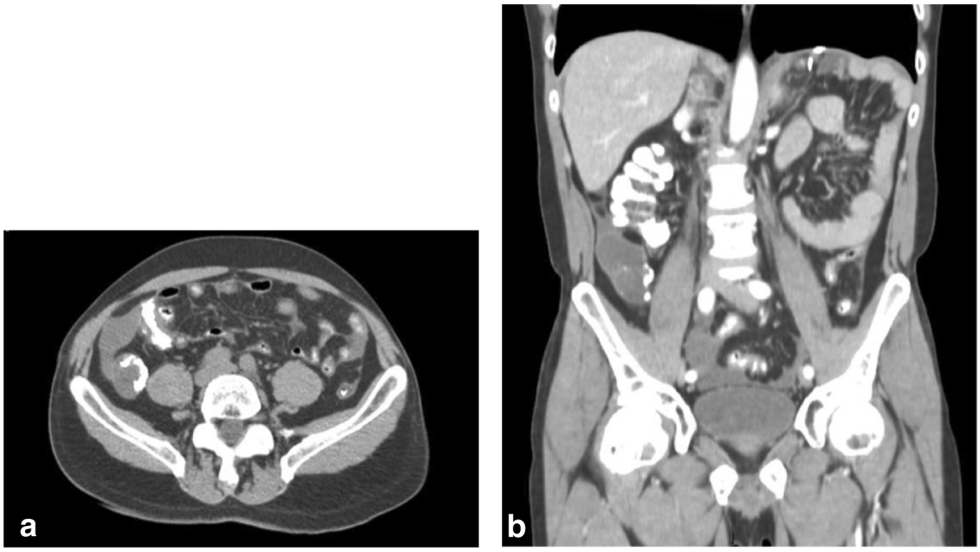

Fig. 1 Abdominopelvic CT scan. Dilated appendix measuring 26 mm in thickness, with mural calcifications, concerning for an appendiceal mucocele. **a** Transverse plane. **b** Coronal plane

a "scalloping" form, especially of the liver and spleen, that distinguishes mucinous from fluid ascites [23]. The peritoneal cancer index (PCI) has been used for the detailed evaluation of the peritoneal spread in tumors of gastrointestinal origin and has been found to be a prognostic indicator of survival [24]. To determine the PCI, the peritoneal cavity must be divided into 13 quadrants, and the presence or absence of tumors within each quadrant is assessed. If no tumor is detected, the score is "0," and if a tumor is detected, the lesion size score given can fluctuate from 1 to 3, depending on the size of the tumor. Bigger tumors have a higher number of lesion size score [25, 26]. Scores of PCI go from 1 to 39, and the higher the number calculated, the worse the prognosis [27]. If the estimated preoperative PCI is greater than 20, in the presence of high-grade histology, complete surgical resection can become very challenging [23]. Therefore, high PCI might be prognostic of operative time and morbidity, but it is still associated with a good long-term outcome [28].

In the past, management of pseudomyxoma peritonei involved repeated drainage of mucinous ascites or surgical debulking through removal of mucinous material and the primary tumor [5]. It was not until the1990s when Sugarbaker peritonectomy procedures showed better outcomes when CRS treatment and HIPEC were used combined [29]. Further studies conducted nowadays by Chua et al. [5] demonstrated the efficacy of this combined modality strategy approach, with acceptable morbidity and mortality in a specialized unit setting of 63%, with patients surviving beyond 10 years. Bevan et al. [4] observed that following a CRS and HIPEC combined treatment, low-grade mucinous adenocarcinomas had a 5-year survival range from 62.5 to 100%, while high-grade mucinous adenocarcinomas had 0–65%. Therefore, this approach is utilized as the current medical care treatment; clinical response of reported patients who received CRS in combination with HIPEC is summarized in Table 2.

HIPEC is used at 42 °C as it elicits a cytotoxic effect, as well as it synergistically enhances the effect of chemotherapy [55]. Intraperitoneal chemotherapy provides the benefit of decreased systemic effects and therapeutic concentrations reached at lower doses [56]. The most common chemotherapeutic agents used are mitomycin C, 5-fluorouracil, cisplatin, and oxaliplatin [57]. Chua et al. [5] demonstrated that HIPEC, despite being associated with an improved rate of progression-free survival, was not shown to be a statistically significant independent factor, when the overall survival was analyzed. Therefore, although HIPEC may improve disease control, complete cytoreduction seems to be an influential factor associated with long-term survival.

Cytoreduction surgery involves a complex series of peritonectomies executed in one single operation, with the purpose to resect all visible tumors from intra-abdominal surfaces [58, 59]. The degrees of cytoreduction completeness (CC score) assess the presence or absence of peritoneal carcinomatosis after CRS [60]. The score goes from 0 to 3, "0" to "1" meaning a complete CRS with no nodules visible or nodules less than 0.25 cm, and "2" to "3" meaning an incomplete CRS with nodules greater than 0.25 cm [61]. Extensive disease is prone to scores of incomplete cytoreduction and complicated postoperative recoveries [21]. In a study conducted in 2004, it was shown that patient with complete CRS had a greater median survival time (32.4 months) than those with incomplete CRS (8.4 months) [61]. Therefore, essentially complete cytoreduction surgery is preferred for long-term survival outcomes [58].

Comparing patients with incomplete cytoreduction, Sugarbaker et al. [62] reported a significant 5-year survival difference of 86% in favor of patients with a complete cytoreduction. However, complete cytoreduction and postoperative complications may be affected by prior operations due the formation of intra-abdominal adhesions

Table 2 Clinical response after combining surgical cytoreduction with HIPEC in patients diagnosed with PMP

Author	Year	Sex	Age	Procedure	Intraperitoneal treatment	Clinical response
Li et al. [22]	2006	M	63	CRS	Given, not specified	Recurrence at 4 years
Haase et al. [30]	2009	F	30	CRS	HIPEC—mitomycin C	NED at 60 months
Zappa et al. [31]	2009	F	48	DS and a subsequent CRS	HIPEC—mitomycin C	Disease progression at 28 months
Khan et al. [32]	2010	M	48	Complete CRS	HIPEC—mitomycin C	NED at 18 months
El-Safadi et al. [33]	2010	F	24	DS	HIPEC—mitomycin C	Recurrence at 5 months
Da Fonseca et al. [34]	2010	F	40	CRS	Mitomycin C	Deceased at 1 month
Morris-Stiff et al. [35]	2011	M	56	DS	HIPEC—mitomycin C	NED at 6 months
Bouchereau et al. [36]	2011	F	46	Complete CRS	HIPEC—oxaliplatin	NED at 5 years
Laskov et al. [37]	2012	F	33	DS	HIPEC—mitomycin C	Recurrence at 18 months (treated with CRS and intraperitoneal infusion of 5-FU; NED at 7 years)
Kitai [38]	2012	F	60	CRS	HIPEC	Deceased at 12 months
Sugarbaker et al. [39]	2014	F	44	DS	HIPEC—mitomycin C	Death at 1 month
Singh et al. [40]	2014	F	26	CRS	HIPEC—mitomycin C	NED at 12 months
Murughan et al. [41]	2014	F	52	DS	HIPEC—mitomycin C	–
Tempfer et al. [42]	2014	F	62	–	PIPAC—cisplatin and doxorubicin	Stable after 6 months
Peixoto et al. [43]	2014	F	53	Complete CRS	HIPEC	Metastatic carcinoma at 18 months
Campanati et al. [44]	2014	F	35	CRS	HIPEC—mitomycin C	NED at 5 years
Sabbagh et al. [45]	2015	F	55	DS	HIPEC—mitomycin C	NED at 12 months
Chauhan et al. [46]	2015	M	75	DS	HIPEC—mitomycin C	Clinical remission
Anania et al. [47]	2015	F	45	DS	HIPEC	NED at 1 month
		F	42	DS	HIPEC	–
Chiruvella et al. [48]	2016	F	61	CRS	HIPEC—mitomycin C	NED at 14 months
Gohda et al. [49]	2016	F	38	Complete CRS	HIPEC—mitomycin C	No recurrence at 2 years
Ababneh et al. [50]	2016	M	44	CRS	HIPEC—oxaliplatin, 5-FU, leucovorin	Mucinous adenocarcinoma 9 months after surgery
Mathur et al. [51]	2016	F	27	CRS	HIPEC—mitomycin C and EPIC	NED at 5 months
Vavinskaya et al. [52]	2016	F	41	Complete CRS	HIPEC	NED at 2 years
McGrath et al. [53]	2017	F	54	CRS	HIPEC—mitomycin C	Deceased at 8 years
Liang et al. [54]	2017	M	44	CRS	Given, not specified	NED at 7 months

F, female; *M*, male; *HIPEC*, hyperthermic intraperitoneal chemotherapy; *CRS*, cytoreductive surgery; *DS*, debulking surgery; *PIPAC*, pressurized intraperitoneal aerosol chemotherapy; *NED*, no evidence of disease

and scars [5]. The adhesions create multiple barriers that limit access of the chemotherapy solution to visceral surfaces and prevent direct contact with the cancer [5, 63]. Moreover, free intraperitoneal cancer cells can become entrapped within the scar tissue matrix, making it impenetrable by intraperitoneal chemotherapy [63].

An elevation of tumor markers CEA, CA 19.9, and CA 125 in mucinous malignancies may indicate the presence or progression of the disease or, after treatment, an increased risk of recurrent disease [23]. Therefore, tumor marker measurements after surgery are highly recommended to correlate their levels with treatment outcomes. Epithelial appendiceal cancer was observed to have a 56.1% positive CEA and a 67.1% positive CA 19.9 [64]. An elevated CEA tumor marker at the time of recurrence might be indicative of a worsening prognosis, and decreased or normal levels can be suggestive that there is no recurrence of the condition [23]. However, despite the fact that CEA was initially increased in a higher percentage of PMP cases, van Ruth et al. [65] reported there was no relation between preoperative CEA and extent of tumor or recurrence. Instead, only CA 19.9 increased in patients with recurrent disease [65].

Performing a CT scan as soon as the disease is suspected, followed by an accurate histological sampling, is critical for early and proper identification and diagnosis of PMP, and it directly influences the outcomes and treatment of course. Patients with an inappropriate diagnosis may have non-definitive treatments like debulking surgery or chemotherapy, affecting long-term outcomes and survival [8]. Clinical awareness of this variable and rare condition is essential for a successful management and improved outcomes. Case reports are pivotal for improving our understanding of the disease, and for the clinical awareness needed to treat patients with PMP.

With the present data available, we recommend the proposed standard of care, the combination of CRS followed by HIPEC for all PMP classifications, in order to avoid malignant progression and morbidity of subsequent debulking surgeries.

In March 2018, the National Comprehensive Cancer Network (NCCN) published a guideline for patients with colon cancer [66] and included information regarding PMP. They indicated that in recent studies, HIPEC did not show a correlation with an improvement in the overall survival of patients, whereas completeness of CRS did [66]. Because the present studies are still inconclusive and unclear as to determine a standard approach, we stand by the optimal treatment for patients with pseudomyxoma peritonei, the combination of HIPEC with CRS. However, because this combination therapy can be challenging for many patients, clinical consideration of both classifications (Ronnet's and WHO classifications) should be contemplated to ensure the treatment is strictly appropriated. Indeed, because the current existing classifications can be considered within a broad umbrella, a revised and integrated category of these two classifications can prove to be beneficial resulting in a more effective and a less adverse effect-related therapy approach for selected patients, avoiding unnecessary harm. Also, a palliative therapy should be included as part of the treatment to provide relief for eligible patients and to those whose treatment is more aggressive.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

References

1. Misdraji J. Mucinous epithelial neoplasms of the appendix and pseudomyxoma peritonei. Mod Pathol. 2015;28(S1):S67–79.
2. Carr NJ (2014). Current concepts in pseudomyxoma peritonei. In Annales de pathologie Elsevier Masson. (Vol. 34, No. 1, pp. 9-13)
3. Fernandes ACDO, Rocha GRMD, Oliveira ADD, Guimarães MD, Carvalho SCD, Chojniak R. Pseudomyxoma peritonei in a pediatric patient: a case report and literature review. Revista da Associação Médica Brasileira. 2018;64(2):195–9.
4. Bevan KE, Mohamed F, Moran BJ. Pseudomyxoma peritonei. World J Gastrointest Oncol. 2010;2(1):44–50.
5. Chua TC, Moran BJ, Sugarbaker PH., Levine EA, Glehen O, Gilly FN, ... & Liauw W (2012). Early-and long-term outcome data of patients with pseudomyxoma peritonei from appendiceal origin treated by a strategy of cytoreductive surgery and hyperthermic intraperitoneal chemotherapy. J Clin Oncol, 30(20), 2449–2456.
6. Homeag M, Enachescu V, Pavel LE, Zaharia B, Manescu N. Pseudomyxoma peritonei. Curr Health Sci J. 2009;35(3):193–6.
7. Smeenk RM, Verwaal VJ, Zoetmulder FAN. Pseudomyxoma peritonei. Cancer Treat Rev. 2007;33(2):138–45.
8. Ramaswamy V. Pathology of mucinous appendiceal tumors and pseudomyxoma peritonei. Indian J Surg Oncol. 2016;7(2):258–67.
9. Sugarbaker PH. Pseudomyxoma peritonei. A cancer whose biology is characterized by a redistribution phenomenon. Ann Surg. 1994;219(2):109.
10. Prayson RA, Hart WR, Petras RE. Pseudomyxoma peritonei. A clinicopathologic study of 19 cases with emphasis on site of origin and nature of associated ovarian tumors. Am J Surg Pathol. 1994;18(6):591–603.
11. Agrawal AK, Bobiński P, Grzebieniak Z, Rudnicki J, Marek G, Kobielak P, ... & Hałoń A (2014). Pseudomyxoma peritonei originating from urachus—case report and review of the literature. Curr Oncol, 21(1), e155.
12. Nawaz A, Karakurum A, Weltman D, Shehata A, Mohammed I, Hadjiyane C, et al. Pseudomyxoma peritonei manifesting as intestinal obstruction. South Med J. 2000;93(9):891–3.
13. Touloumis Z, Galyfos G, Kavouras N, Menis M, Lavant L. Aggressive pseudomyxoma peritonei: a case report with an unusual clinical presentation. Case Reports Oncol Med. 2013;2013:1–4.
14. Esquivel J, Sugarbaker PH. Clinical presentation of the pseudomyxoma peritonei syndrome. Br J Surg. 2000;87(10):1414–8.
15. Noguchi R, Yano H, Gohda Y, Suda R, Igari T, Ohta Y, et al. Molecular profiles of high-grade and low-grade pseudomyxoma peritonei. Cancer Med. 2015;4(12):1809–16.
16. Amini A, Masoumi-Moghaddam S, Ehteda A, Morris DL. Secreted mucins in pseudomyxoma peritonei: pathophysiological significance and potential therapeutic prospects. Orphanet J Rare Dis. 2014;9(1):71.
17. 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(12):1390–408.
18. Bradley RF, Stewart JH IV, Russell GB, Levine EA, Geisinger KR. Pseudomyxoma peritonei of appendiceal origin: a clinicopathologic analysis of 101 patients uniformly treated at a single institution, with literature review. Am J Surg Pathol. 2006;30(5):551–9.
19. Sugarbaker PH. New standard of care for appendiceal epithelial neoplasms and pseudomyxoma peritonei syndrome? Lancet Oncol. 2006;7(1):69–76.
20. Sugarbaker PH. Cytoreductive surgery and intraperitoneal chemotherapy with peritoneal spreadof cystadenocarcinoma. Eur J Surg Suppl. 1991;561:75–82.
21. Winder T, Lenz HJ. Mucinous adenocarcinomas with intra-abdominal dissemination: a review of current therapy. Oncologist. 2010;15(8):836–44.
22. Li C, Kanthan R, Kanthan SC. Pseudomyxoma peritonei–a revisit: report of 2 cases and literature review. World J Surg Oncol. 2006;4(1):60.
23. Kelly KJ. Management of appendix cancer. Clin Colon Rectal Surg. 2015;28(4):247–55.
24. Tentes AA, Tripsiannis G, Markakidis SK, Karanikiotis CN, Tzegas G, Georgiadis G, et al. Peritoneal cancer index: a prognostic indicator of survival in advanced ovarian cancer. Eur J Surg Oncol (EJSO). 2003;29(1):69–73.
25. Heaney RM, Shields C, Mulsow J. Outcome following incomplete surgical cytoreduction combined with intraperitoneal chemotherapy for colorectal peritoneal metastases. World J Gastrointest Oncol. 2015;7(12):445–54.
26. Di Giorgio A, Naticchioni E, Biacchi D, Sibio S, Accarpio F, Rocco M, et al. Cytoreductive surgery (peritonectomy procedures) combined with hyperthermic intraperitoneal chemotherapy (HIPEC) in the treatment of diffuse peritoneal carcinomatosis from ovarian cancer. Cancer. 2008;113(2):315–25.
27. Gilly FN, Cotte E, Brigand C, Monneuse O, Beaujard AC, Freyer G, et al. Quantitative prognostic indices in peritoneal carcinomatosis. Eur J Surg Oncol (EJSO). 2006;32(6):597–601.
28. Morris DL. Peritonectomy HIPEC—contemporary results, indications. Chin J Cancer Res. 2013;25(4):373–4.

29. Sugarbaker PH. Peritonectomy procedures. Ann Surg. 1995;221(1):29–42.
30. Haase E, Yoo D, Sugarbaker PH. Management of appendiceal pseudomyxoma peritonei diagnosed during pregnancy. World J Surg Oncol. 2009;7(1):48.
31. Zappa L, Savady R, Humphries GN, Sugarbaker PH. Interstitial pneumonitis following intrapleural chemotherapy. World J Surg Oncol. 2009;7(1):17.
32. Khan AB, Al Suhaibani Y, Al Mohaimed K, Morris DL. Application of advanced multimodality care to pseudomyxoma peritonei patient: report of first patient treated at a tertiary center. Indian J Surg Oncol. 2010;1(3):270–3.
33. El-Safadi S, Stahl U, Tinneberg HR, Hackethal A, Muenstedt K. Primary signet ring cell mucinous ovarian carcinoma: a case report and literature review. Case Reports Oncol. 2010;3(3):451–7.
34. Fonseca LMD, Lacerda-Filho A, Silva RGD. Pseudomyxoma peritonei syndrome 12 months after an intact resection of malignant mucocele of the appendix: a case report. Clinics. 2010;65(8):817–8.
35. Morris-Stiff G, Falk GA, Joyce D, Chalikonda S. Unusual presentation of more common disease/injury: unusual contents of a scrotal swelling. BMJ Case Reports. 2011;2011:bcr0320114023.
36. Bouchereau M, Gervais MK, Sideris L, Loriot MH, Ahern SP, Dubé P. Hepatic necrosis and hemorrhage following hyperthermic intraperitoneal chemotherapy with oxaliplatin: a review of two cases. J Gastrointest Oncol. 2011;2(2):113–6.
37. Laskov I, Michaan N, Many A, Amit A, Azem F. Successful pregnancy following transfer of frozen-thawed embryos in a patient with pseudomyxoma peritonei who underwent peritonectomy and bilateral oophorectomy. J Gynecol Oncol. 2012;23(2):129–32.
38. Kitai T. Pulmonary metastasis from pseudomyxoma peritonei. Gastroenterol Res Pract. 2012;2012:*1–6*.
39. Sugarbaker PH, Rangole AK, Carr NJ. Peritoneal metastases from mucinous endocervical adenocarcinoma. Gynecol Oncol Reports. 2014;10:5–8.
40. Singh VK, Nishant K, Sharma BK, Lamichaney R. Case report: a weeping umbilical hernia: bilateral ovarian mucinous cystadenoma with disseminated peritoneal adenomucinosis. BMJ Case Reports. 2014;2014:bcr2014204748.
41. Murughan K, Tiwari M, Balakrishnan K. Cytoreduction with hyperthermic intraperitoneal chemotherapy: an anaesthetic challenge. Indian J Anaesth. 2014;58(1):59–62.
42. Tempfer CB, Solass W, Buerkle B, Reymond MA. Pressurized intraperitoneal aerosol chemotherapy (PIPAC) with cisplatin and doxorubicin in a woman with pseudomyxoma peritonei: a case report. Gynecol Oncol Reports. 2014;10:32–5.
43. Peixoto RDA, Wilson S, Schaeffer DF, Lim HJ. Pseudomyxoma peritonei metastatic to the bone: case report and review of systemic management. Gastrointest Cancer Res: GCR. 2014;7(3–4):108.
44. Campanati RG, Hanan B, Souza SSD, Gomes da Silva R. Pregnancy after cytoreductive surgery and hyperthermic intraperitoneal chemotherapy in pseudomyxoma peritonei. Case Reports Surg. 2014;2014:*1–3*.
45. Sabbagh C, Vaillandet C, Tuech JJ, Regimbeau JM. A new etiology for the abdominal compartment syndrome: pseudomyxoma peritonei. Case Reports Gastroenterol. 2015;9(3):307–10.
46. Chauhan A, Patodi N, Ahmed M. A rare cause of ascites: pseudomyxoma peritonei and a review of the literature. Clin Case Reports. 2015;3(3):156–9.
47. Anania G, Giaccari S, Solfrini G, Scagliarini L, Vedana L, Resta G. Appendicular mucocele: two case reports and literature review. Il Giornale di chirurgia. 2015;36(6):276–9.
48. Chiruvella A, Staley CA, Khanna N, Russell M, Maithel SK, et al. Pseudomyxoma Peritonei from a borderline mucinous tumor arising in an ovarian mature cystic teratoma: a rare case report. Arch Surg Oncol. 2016;2:114. https://doi.org/10.4172/2471-2671.1000114.
49. Gohda Y, Noguchi R, Horie T, Igari T, Nakamura H, Ohta Y, ... & Furukawa Y (2016). Pseudomyxoma peritonei of a mature ovarian teratoma caused by mismatch repair deficiency in a patient with Lynch syndrome: a case report. BMC Med Gen, 17(1), 94.
50. Ababneh R, Piso P, Hofmann HS, Dumitrovici A, Buiga R, Samasca G, et al. An invasive treatment of pseudomyxoma peritonei with intrathoracic involvement. Maedica. 2016;11(1):72–5.
51. Mathur S, Kelly E. Pseudomyxoma peritonei: a case report of an incidental finding at cesarean section. J Clin Gynecol Obstet. 2016;5(1):32–6.
52. Vavinskaya V, Baumgartner JM, Ko A, Saenz CC, Valasek MA. Low-grade appendiceal mucinous neoplasm involving the endometrium and presenting with mucinous vaginal discharge. Case Rep Obstet Gynecol. 2016;2016:*1–4*.
53. McGrath C, Linden K, Hube P, Adamiak A, Dennis K. Palliative radiation therapy for symptom control in an advanced case of pseudomyxoma peritonei. Cureus. 2017;9(6):e1407.
54. Liang L, Zhou N, Xu H, Liu D, Lu Y, Li F, et al. Urachal mucinous adenocarcinoma with pseudomyxoma peritonei: a case report. Medicine. 2017;96(35):e7548.
55. González-Moreno S, González-Bayón LA, Ortega-Pérez G. Hyperthermic intraperitoneal chemotherapy: rationale and technique. World J Gastrointest Oncol. 2010;2(2):68–75.
56. Markman M. Intraperitoneal chemotherapy in the management of malignant disease. Expert Rev Anticancer Ther. 2001;1(1):142–8.
57. Loungnarath R, Causeret S, Bossard N, Faheez M, Sayag-Beaujard AC, Brigand C, et al. Cytoreductive surgery with intraperitoneal chemohyperthermia for the treatment of pseudomyxoma peritonei: a prospective study. Dis Colon Rectum. 2005;48(7):1372–9.
58. Sugarbaker PH. Peritonectomy procedures. Surg Oncol Clin N Am. 2003;12(3):703–27.
59. Sugarbaker PH, Gianola FJ, Speyer JL, Wesley R, Barofsky I, & Myers CE (1985). Prospective randomized trial of intravenous v intraperitoneal 5-FU in patients with advanced primary colon or rectal cancer. In Seminars in oncology. Elsevier. Vol. 12, no. 3, pp. 101-111
60. Ortega-Deballon P, Glehen O, Levine E, Piso P, Sugarbaker PH, Hayes-Jordan A, et al. Childbearing after hyperthermic intraperitoneal chemotherapy: results from an international survey. Ann Surg Oncol. 2011;18(8):2297–301.
61. Glehen O, Kwiatkowski F, Sugarbaker PH, et al. Cytoreductive surgery combined with perioperative intraperitoneal chemotherapy for the management of peritoneal carcinomatosis from colorectal cancer: a multi-institutional study. J Clin Oncol. 2004;22:3284–92.
62. Sugarbaker PH, Chang D. Results of treatment of 385 patients with peritoneal surface spread of appendiceal malignancy. Ann Surg Oncol. 1999;6(8):727–31.
63. Sugarbaker PH. Managing the peritoneal surface component of gastrointestinal cancer. Part 2. Perioperative intraperitoneal chemotherapy. Oncology (Williston Park, NY). 2004;18(2):207–19.
64. Pablo Carmignani C, Hampton R, Sugarbaker CE, Chang D, Sugarbaker PH. Utility of CEA and CA 19-9 tumor markers in diagnosis and prognostic assessment of mucinous epithelial cancers of the appendix. J Surg Oncol. 2004;87(4):162–6.
65. Van Ruth S, Hart AAM, Bonfrer JMG, Verwaal VJ, Zoetmulder FAN. Prognostic value of baseline and serial carcinoembryonic antigen and carbohydrate antigen 19.9 measurements in patients with pseudomyxoma peritonei treated with cytoreduction and hyperthermic intraperitoneal chemotherapy. Ann Surg Oncol. 2002;9(10):961–7.
66. NCCN (National Comprehensive Cancer Network). Colon Canr: NCCN Clinical Practice Guidelines in Oncology (Version 4.2018). https://www.nccn.org/professionals/physician_gls/pdf/colon.pdf. Accessed 29 July 2018.