Retrospective study of different methods for managing Egyptian patients with pseudomyxoma peritonei: feasibility and overall outcome

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Introduction
Pseudomyxoma peritonei (PMP) is a rare disease with poor outcome when not treated properly and is characterized by mucinous ascites and peritoneal implants. Treatment for PMP is variable, both because of the rarity of the disease and because of its often slow-growing nature. There is no consensus regarding the proper management of aggressive cases. Our study was designed to analyze the symptoms and signs of PMP, as well as the diagnostic tools, and evaluate the effect of treatment and factors influencing postoperative recurrence of and survival from PMP in Egyptian patients.

Patients and Methods
We reviewed consecutive cases of PMP that presented at the surgical department of the Main Alexandria University Hospital from January 1990 to December 2012.

Results
This study included 62 patients with PMP: 43 were women (69%) and 19 were men (31%). Their mean age at the time of diagnosis was 47.3 ± 11.6 years (median 49, range 29–67). The predominance of women was statistically significant ($P = 0.08$). A total of 69 surgical procedures had been performed in 46 cases, including 46 primary operations, 10 secondary operations, one tertiary cytoreduction and peritonectomy, and 12 debulking procedures for recurrence.

Conclusion
Surgical debulking is the standard treatment for PMP in primary and recurrent tumors. Intraperitoneal chemotherapy either intraoperative or postoperative is accompanied by better disease-free survival and overall survival. Referring of patients to specialized centers that treat these patients on a regular basis is essential to prevent high morbidity and mortality. Recurrence is common and requires reoperation with or without adjuvant chemotherapy.

Keywords:
aggressive peritoneal tumors, intraperitoneal chemotherapy, surgical treatment

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Introduction

Pseudomyxoma peritonei (PMP) is an uncommon locoregional surgical entity with an estimated incidence of one or two per million per year or two per 10 000 laparotomy cases. It is characterized by the presence of a mucinous tumor on peritoneal surfaces producing a progressive amount of mucinous ascites. The primary tumor is thought to be predominately a mucinous epithelial neoplasm of the appendix [1].

The term PMP was first introduced by Werth in 1884 [2] describing it in association with a mucinous tumor of the ovary. In 1901, Frankel [3] described a case associated with a cyst of the appendix. If PMP is left untreated, mucin will eventually build up to the point where it compresses vital structures, such as the colon, the liver, kidneys, stomach, spleen, and pancreas [4]. Many cases present unexpectedly at laparoscopy or laparotomy, or may be suspected at cross-sectional imaging, or staging, of another pathological entity. PMP is a disease more commonly seen in women (male : female ratio = 6 : 11); the mean age at presentation is 58 years [5].

Regarding its classification, PMP is a broad descriptive term embracing a wide spectrum of biological behaviors of the neoplasm, from the benign to the frankly malignant lesion. Ronnett *et al.* [6] proposed a classification distinguishing 'disseminated peritoneal adenomucinosis' from 'peritoneal mucinous carcinomatosis'.

A definitive diagnosis of PMP requires the presence of (a) mucinous neoplastic cells/epithelium and (b) mucinous ascites with diffuse intra-abdominal mucin. Some authors also consider the presence of diffuse mucinous implants for a diagnosis [7]. Viable epithelial glandular cells must be identified within the mucin pools by histological analysis to diagnose PMP.

PMP is a heterogeneous lesion, which may develop from mucinous metaplasia of the peritoneum or from appendiceal or ovarian lesions. Careful examination

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of the ovary and appendix with appendectomy is advised in every case of PMP. Immunohistochemical examination of the peritoneal, ovarian, or appendiceal lesions using antibodies, in particular, that for CK7, would help in defining the origin of mucin production.

Recently, MUC 2 overexpression was suggested as a molecular marker for PMP of intestinal rather than of ovarian origin [8]. PMP has been reported as originating from the colon and rectum, stomach, gallbladder and bile ducts, small intestine, urinary bladder, lung, breast, fallopian tube, and pancreas.

Abdominal pain is the most common complaint of PMP with an incidence of 23% in the initial evaluation, whereas a newly onset hernia is seen in 12% of cases [9]. The usual clinical features of this tumor are increasing abdominal girth, ovarian tumors, hernia sac tumors, appendicitis-like syndrome, and infertility [10]. Primary PMP rarely causes complications even in the presence of large-volume disease. Rarely, ureteric obstruction and lower limb edema secondary to venous obstruction have been reported. Recurrent disease, however, may occur on bowel surfaces and can cause fibrosis and intestinal adhesions. This often leads to intestinal obstruction or obstructive jaundice, which may prove fatal [11].

Narrowing, but rarely complete obstruction, of the gastrointestinal tract frequently occurs at three well-defined anatomic sites: the pyloric antrum, the ileocecal valve, and the cul-de-sac of Douglas [9]. These are three portions of the gastrointestinal tract that are attached to the retroperitoneum and are relatively motionless. As mentioned above, PMP has multiple clinical manifestations that lead to difficulties in definitive diagnosis and timely treatment [12]. As symptoms remain nonspecific, the disease presents a great diagnostic challenge to clinicians. PMP can present with unspecific symptoms that mislead diagnosis. Patients usually experience a long period of deterioration in health before an accurate diagnosis is made [13].

The prognosis of PMP is poor. The 5-year survival rate is less than 50% [14]. Recently, heated intraperitoneal chemotherapy (HIPEC) was reported to provide 62.5–100% survival rates for low-grade and 0–65% rates for high-grade disease [1]. Ten percent of patients die of PMP within 5.5 years of their initial presentation. Overall survival (OS) of patients is about 75 and 68 for 5 and 10 years, respectively, as revealed by Ronnett *et al.*[6].

Treatment for PMP is variable, both because of the rarity of the disease and because of its often slow-growing nature [9]. Current treatment strategies range from follow-up with palliative supportive care to cytoreductive surgery with HIPEC or early postoperative intraperitoneal chemotherapy [15]. Palliative care includes monitoring with computed tomography (CT) scans, tumor marker tests, and physical symptoms, to determine when, and if, surgery is warranted.

There is no consensus regarding the proper management of aggressive cases. Recent studies support that cytoreduction with peritonectomy plus HIPEC is a safe procedure that suggests an improvement in survival rates, even in aggressive cases [9]. Our study was designed to analyze the symptoms and signs of PMP, as well as the diagnostic tools, and evaluate the effect of treatment and factors influencing postoperative recurrence and survival in PMP in Egyptian patients.

Patients and methods

We reviewed consecutive cases of PMP that presented to the surgical department of the Main Alexandria University Hospital from January 1990 to December 2012. We analyzed patient characteristics, clinical manifestations, surgical indications, preoperative radiological investigations, and patient management.

All analyses were performed using SPSS for Windows 16.0.1 software [16]. The sex preponderance was analyzed with the χ^2 method. The data were presented in proportions (percentages) for categorical variables and as means ± SEs (medians) for continuous variables.

Results

This study included 62 patients with PMP: 43 were women (69%) and 19 were men (31%). Their mean age at the time of diagnosis was 47.3 ± 11.6 years (median 49, range 29–67). The predominance of women was statistically significant (P = 0.08). There were five clinical presentations among our patients (Table 1).

Table 1 Different clinical presentations among patients with PMP

Chief complaint	*N* (%)		
	Females	Males	Total
Abdominal pain	13 (21)	6 (9)	19 (31)
Acute abdomen	4 (6)	5 (8)	9 (15)
Increasing abdominal girth	11 (18)	4 (6)	15 (24)
Newly onset hernia	4 (6)	4 (6)	8 (13)
Coincidental diagnosis	10 (16)	1 (1)	11 (17)
Total	43 (69)	19 (31)	62 (100)

PMP, pseudomyxoma peritonei.

The chief complaint was abdominal pain [28 patients (46%)], either chronic [19 cases (31%)] or acute [nine cases (15%)]. This was followed by increase in abdominal girth (abdominal distension) [15 cases (24%)] (Fig. 1), development of a new hernia [eight cases (13%), and finally coincidental diagnosis [11 cases (17%)]. During gynecological examination the condition was seen to be present in six women. Individual reasons for four women were cystocele, fever, mucoid feces, and palpable tumor; one man presented with a hydrocoele.

Seven patients (11%) had only a definitive preoperative histopathological diagnosis of PMP, which had been done by ultrasonography-assisted Tru-cut needle biopsy. The diagnosis was confirmed by open surgical biopsy taken at the initial surgery in 44 cases (71%), at the second exploratory operation in 10 cases (16%), and at the third exploratory operation in one case.

The time interval between symptoms and definitive diagnosis ranged from 2 to 8 months, with a mean of 4 months. Various methods had been used for radiological investigation of our patients, some of them had undergone more than one technique. All patients had been investigated with abdominal ultrasonography. Of the 62 patients, 35 (57%) had undergone CT before initial surgery, three patients (5%) had undergone MRI, and another three patients (5%) had undergone gastrointestinal contrast film. PMP had been diagnosed from 18 (51%) of the 35 CT scans, and hence CT had a sensitivity of 51% in recognizing PMP (Fig. 2).

For the 46 patients who had undergone initial surgery with a suspicion or diagnosis of PMP, diagnosis had been made in several ways using different tools, and some of them had more than one proven diagnosis. The diagnosis had been made using CT in 14 patients (30%), with ultrasonography in 16 patients (35%), by MRI in two patients (4%), by a combination of ultrasonography-assisted fine needle biopsy and gastrointestinal contrast film in one patient (2%), and with ultrasonography-assisted fine needle biopsy in six patients (13%). Suspected ovarian tumor was the most common cause for PMP, comprising 26 of 46 initial surgeries (56%).

During operations, synchronous ovarian lesions were detected in nine cases, appendiceal mass in four cases, metastatic adenocarcinoma in nine cases, adenocarcinoma of the pancreas in two patients, and gastric carcinoma in one patient. The origin of mucin production was not detected in most of the cases [21 cases (46%)].

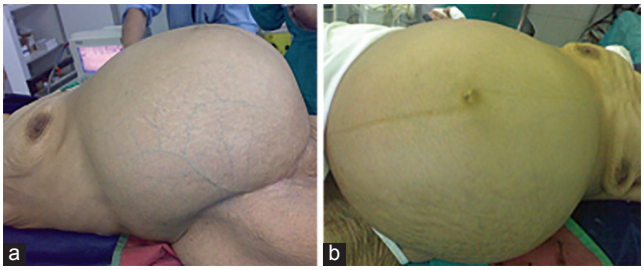
Normal preoperative CA125, carcinoembryonic antigen (CEA), and CA19.9 levels were detected in 29 patients (47%). On the other hand, high levels of CA125 were correlated with ovarian neoplasm in 12 cases, high CA19.9 was correlated with the two cases of pancreatic carcinoma, and CEA levels were high in seven cases of appendicular neoplasm.

Surgical debulking has been the traditional protocol at Alexandria Main University Hospital for managing patients with PMP. However, since 2007, an aggressive cytoreductive surgery (CRS) using peritonectomy has also been used in selected patients.

Treatment methods

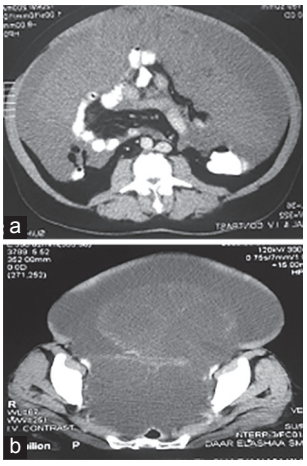
(a) Watch and wait: This approach was adopted in four patients (11%) whose general health condition and age were not suitable for surgical or adjuvant therapy.
(b) Surgery: The traditional surgical approach was debulking to remove as much of the tumor as possible, which had been applied in 46 cases (74%) with additional removal of one or more organs,

Figure 1



A female patient who presented with a huge distended abdomen. (a, b) The patient was diagnosed preoperatively as having PMP and received chemotherapy without response. PMP, pseudomyxoma peritonei.

Figure 2



(a) CT of the abdomen and pelvis showing a huge abdominal mass (omental cake). (b) Tense abdominal ascites (jelly belly). CT, computed tomography.

including removal of the uterus and ovaries in 25 cases, appendix or right colon in 39 cases, and the greater omentum in all 46 cases. Disease recurrence had occurred in all cases with an average disease-free period of 9 months (range: 6–18 months) (Figs. 3 and 4).

Repeat debulking surgery for recurrence had been performed for 12 cases with a higher rate of postoperative respiratory, cardiac, small bowel fistula, and wound complications. Complications were seen in 11 patients (91%).

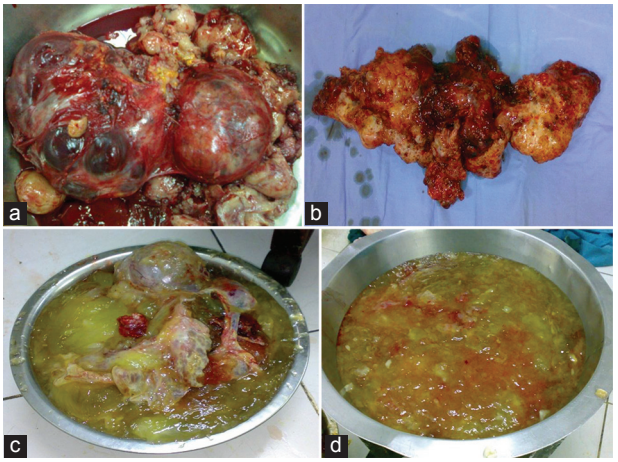
A 65-year-old man presented with a huge abdominal mass (Fig. 4). On exploration, a large omental cake was found with multiple tumor deposits at the mesentery of the intestine. Complete excision of the mass and debulking of the abdominal deposits was done. No apparent primary cause was found, and the postoperative pathology was PMP. This patient came back after 2 years with a recurrent abdominal mass. On second exploration, debulking of the mass with appendectomy was performed.

(c) CRS as in (b) combined with total parietal peritonectomy had been performed as a primary operation in only 12 cases and as a second operation in 10 cases: a total of 22 out of 62 cases of PMP (35%). Peritonectomy procedures involved stripping of the parietal peritoneum and resecting structures at fixed sites that contained adenomucinosis tumor tissue (Fig. 5).

The CRS, consisting of five procedures: omentectomy, splenectomy, left subdiaphragmatic peritonectomy, right subdiaphragmatic peritonectomy, pelvic peritonectomy — sleeve resection of sigmoid colon and cholecystectomy — lesser omentectomy three of our cases had been followed by postoperative intraperitoneal chemotherapy with mitomycin C, heated to 44°C, followed by intraperitoneal 5-FU for 5 days through abdominal drains.

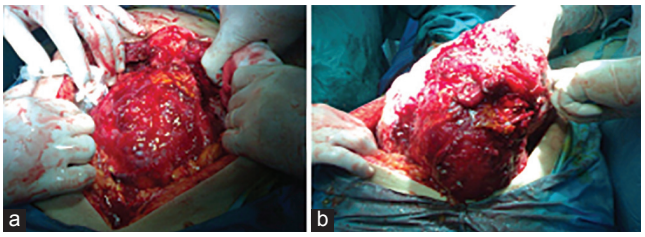
(d) Systemic chemotherapy: First indication included nine patients with large tumors or with inoperable or recurrent PMP in our series (15%) had been offered a combination of primary chemotherapeutic agents as a primary method of treatment. The patients were given six cycles of adjuvant chemotherapy. For inoperable or recurrent cases, the response was assessed after three cycles. These patients were then reconsidered for surgery or for continuation of palliative treatment. Three patients (33%) had partial benefit from this treatment with partial response.

Figure 3



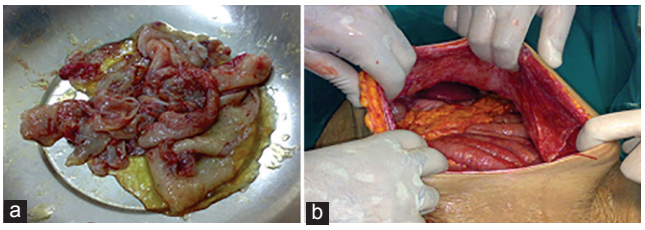
(a) Total abdominal hysterectomy. (b) Omentectomy of omental cake. (c) Total evacuation of the abdomen. (d) Large amount of gelatinous ascites (jelly like).

Figure 4



65 years old male patient presented with huge abdominal mass, on exploration, large omental cake was found with multiple tumor deposits at mesentery of the intestine, complete excision of the mass, debulking of the abdominal deposits was done. No apparent primary cause was found, the pathology postoperative was PMP. This patient came back after 2 year with recurrent abdominal mass, on second exploration, debulking of the mass with appendectomy was done.

Figure 5



(a) Total omentectomy and parietal peritonectomy. (b) Abdomen ready for closure.

Another 18 patients (30%) had undergone chemotherapy as adjuvant treatment: after surgery in 10 cases that had undergone primary reduction surgery or after excision of recurrence in eight cases. It was generally well tolerated with little side effects. The most commonly used agents include cisplatin, carboplatin, paclitaxel, 5-flourouracil, capcitapine, and mitomycin C given either intravenously or

intraperitoneally (using a postoperative intraperitoneal catheter). The intraperitoneal drain catheter had been used for injection of local intraperitoneal chemotherapy comprising three cycles in only three cases. Patients had good tolerance to local peritoneal chemotherapy with no major complications. For PMP of ovarian origin, a combination of cisplatin or carboplatin and paclitaxel was administered to 25 patients, 5-flourouracil or capcitapine, mitomycin C with or without cisplatin for gastrointestinal origin cases, and gemcitabine for pancreatic tumors (two cases).

A total of 69 surgical procedures had been performed in 46 cases, including 46 primary operations, 10 secondary operations, one tertiary cytoreduction and peritonectomy, and 12 second debulking procedures for recurrence. Postoperative mortality and morbidity results are shown in Table 2.

The 1-, 3-, and 5-year survival rates for all patients with PMP were 25.5, 9.3, and 0%, respectively. The median OS was 21 months. Details of disease-free survival and OS for all cases are shown in Table 3. Also disease-free survival and OS of cases of recurrence are shown in Table 4.

Discussion

PMP is a rare condition, which is known for its high mortality when not treated properly. The first step toward improving the prognosis of these patients is to recognize this clinical syndrome at an early stage. The main symptoms of our patient were nonspecific and misleading, and led to delayed diagnosis.

The most important symptom cited in the literature is increasing abdominal girth (50%), whereas in our patients abdominal pain was the chief complaint (46%), which characterizes the progressive stage of disease with peritoneal dissemination. Patients present late with a typical 'jelly belly' and complain of intestinal obstruction caused by the progressive amount of mucinous tumor and ascites.

The mean interval between the existence of a primary tumor and established PMP is described to be ~21 months [17]. The time interval between onset of symptoms and definitive diagnosis in our series ranged from 2 to 8 months with a mean of 4 months.

Histopathological examination of the appendix usually reveals a mucinous epithelial neoplasm. In 30% of female patients, the first symptom is an ovarian mass. Often these patients first consult the gynecologist with a pelvic mass. The correct diagnosis is then awaited for until after surgery, when examination of the mucinous ovarian tumor and the appendix reveals PMP metastases from an appendiceal primary or less frequently from an ovarian primary. In some cases, during laparoscopy or laparotomy for whatever reason, or during hernia repair, the surgeon or gynecologist unexpectedly encounters mucus [18].

In our patients, various methods for investigations were used, including abdominal ultrasound, CT, MRI, and gastrointestinal contrast film. PMP was diagnosed in 18 of the afore-mentioned 35 CT scans. CT had a sensitivity of 51% in recognizing PMP. Suspected ovarian tumor was the most common cause of PMP, comprising 57% of initial surgeries.

The origin of mucin production was not detected in most of the cases [21 cases (46%)]. Immunoreactivity for CK20 and CK7 is a good method for defining the original tumor. Unfortunately, these methods had not been applied to most of our cases.

Table 2 Different complication rates according to type of surgical interference

Type of operation	Debulking	Peritonectomy	Cytoreduction, pertonectomy	Recurrence debulking	Total [*n* (%)]
Number	46	10	1	12	69
Operative mortality	2	1	0	4	7 (10)
Cardio-respiratory complications	6	2	1	6	15 (22)
Deep vein thrombosis	10	1	0	4	15 (22)
Intestinal leak	2	2	0	5	9 (13)
Wound dehiscence	8	2	0	4	14 (20)
Wound infection	14	2	1	4	21 (30)
Total					69 (100)

Table 3 DFS and OS of all cases according to method of treatment

Type of operation	Primary debulking	Cytoreduction and pretoneictomy	Systemic chemotherapy	Palliative supports	*P* value
Number	35	11	2	4	
Disease-free survival	10	18	0	0	NA
Overall survival	27	34	10	5	0.03

DFS, disease-free survival; NA, not applicable; OS, overall survival.

Table 4 DFS and OS of recurrent cases after recurrence

Type of operation	Debulking of recurrence	Chemotherapy for recurrence	Palliative for recurrence	*P* value
Number	12	14	20	
Disease-free survival	8	0	0	NA
Overall survival	10	4	2	0.09

DFS, disease-free survival; NA, not applicable; OS, overall survival.

There has recently been a global interest in the management of PMP, particularly in the removal of the tumor through complex surgical techniques combined with total parietal peritoneal excision with or without HIPEC [19].

CRS with intraoperative HIPEC is a treatment strategy with encouraging survival results for selected PMP patients. The efficacy and superiority of this treatment compared with serial debulking have been established by many studies across the world. The outcome of this treatment has shown survival rates of 85% in 20 years, according to the latest follow-up studies. At least two randomized trials and many multicentric studies support this claim [20,21].

After treatment, patients should be monitored for recurrent or progressive disease. A CT scan is a very important tool for detecting progressive disease and can be performed 3 months after treatment as the basis for further follow-up. After that, a CT scan should be performed every 6 months in the first year and once a year or when progression is suspected in subsequent years. Other useful tools for detecting disease in the postsurgical period are the tumor markers CEA and CA 19.9, which also act as prognostic factors for survival [17].

Surgical excision of the tumor without rupture is important because rupture of the lesion causes PMP. Even in case of benign disease such as cystadenoma, dissemination of mucin-producing cells into the peritoneal cavity can cause PMP [22].

On the basis of the Sugarbaker peritonectomy procedure, a study by Deraco and colleagues showed that CRS with intraperitoneal hyperthermic perfusion permitted complete tumor removal, and this study confirmed the efficacy of this combined treatment in terms of improved long-term survival and better local control of the disease [23,24]. A recent study by Faris and Ryan concludes that the treatment of the low-grade variants of PMP includes serial cytoreduction surgery, with data indicating possible, but unproven, benefit from HIPEC, whereas there is no consensus so far on the role of cytoreduction and HIPEC in the management of the more aggressive histological variants and peritoneal carcinomatosis [25].

As a result, they support that systemic chemotherapy should be the standard of care for patients with high-grade variants and peritoneal carcinomatosis, as in our cases. Recent studies with promising results have shown that fluorouracil-based adjuvant chemotherapy can be used for PMP of appendiceal origin [9]. However, one must know that most of these studies do not focus on cases of aggressive PMP. Finally, PMP may recur following CRS, as seen in our case, especially when the disease is diagnosed at an advanced stage.

In recent times the laparoscopic approach has enabled avoidance of a large incision for exploration of the peritoneal cavity, conferring the advantage of minimally invasive surgery. Laparoscopic access and visualization may be compromised by disease extent, in particular a large omental 'cake', rendering accurate laparoscopic assessment impossible.

There are no randomized data on the role of adjuvant chemotherapy. Results from phase II trials and retrospective reviews are conflicting. The most widely used chemotherapeutic agents are 5-FU, cyclophosphamide, mitomycin C, and cisplatinum. The retrospective analysis from the Mayo Clinic series showed that survival rates were significantly better in patients treated with intraperitoneal infusions than in those treated with systemic chemotherapy.

The use of heated intraoperative intraperitoneal chemotherapy after complete dissection of adhesions and before anastomoses are completed allows optimal perfusion of the chemotherapy to the peritoneal surfaces and organs. There has recently been a global interest in the management of PMP, particularly in removal of the tumor by complex surgical techniques combined with total parietal peritoneal excision with or without HIPEC [19]. We hope that by introducing this technique at our institute we can improve local and overall outcomes.

Conclusion

PMP is a rare disease with poor prognosis when not treated properly and is characterized by mucinous ascites and peritoneal implants. The first step in improving the prognosis is to recognize the clinical syndrome at an early stage. CT imaging should be the choice for radiological assistance in the diagnosis of PMP.

Surgical debulking is the standard treatment for PMP in primary and recurrent tumors. Combined modality treatment, consisting of CRS in combination with total parietal peritonectomy, could be appropriate for

aggressive peritoneal mucinous carcinomatosis. Adding intraoperative HIPEC is the standard approach for improving survival after surgical debulking of tumors with favorable histology.

Referring of patients to specialized centers that treat these patients on a regular basis is essential to prevent high morbidity and mortality. Improvement of survival can be achieved by a combination of surgical experience and adequate patient selection. Recurrence is common and requires reoperation with or without adjuvant chemotherapy.

Acknowledgements

Conflicts of interest

None declared.

References

1 Bevan KE, Mohamed F, Moran BJ. Pseudomyxoma peritonei. World J Gastrointest Oncol. 2010; 2: 44–50.

2 Werth R, Pseudomyxoma peritonei Arch Gynecol Obstet, 24, 100–118, 1884.

3 Frankel E Uber das sogenannte pseudomyxoma peritonei. Med Wochenschr. 1901;48:965–970.

4 Buell-Gutbrod R, Gwin K, Pathologic diagnosis, origin, and natural history of pseudomyxoma peritonei. Proceedings of the ASCO Annual Meeting, May 31 - June 4, 2013; Chicago, Illinois.

5 Winder T, Lenz HJ Mucinous adenocarcinomas with intra-abdominal dissemination: a review of current therapy. Oncologist 2010;15:836–844.

6 Ronnett BM, Yan H, Kurman RJ, Shmookler BM, Wu L, Sugarbaker PH Patients with pseudomyxoma peritonei associated with disseminated peritoneal adenomucinosis have a significantly more favorable prognosis than patients with peritoneal mucinous carcinomatosis. Cancer. 2001;92:85–91.

7 Li C, Kanthan R, Kanthan SC. Pseudomyxoma peritonei – a revisit: report of 2 cases and literature review. World J Surg Oncol 2006; 4:60.

8 Ferreira CR, Carvalho JP, Soares FA, Siqueira SA, Carvalho FM Mucinous ovarian tumors associated with pseudomyxoma peritonei of adenomucinosis type: immunohistochemical evidence that they are secondary tumors. Int J Gynecol Cancer. 2008;18:59–65.

9 Touloumis Z, Galyfos G, Kavouras N, Menis M, Lavant L Aggressive pseudomyxoma peritonei: a case report with an unusual clinical presentation. Case Rep Oncol Med; 2013 (2013), 926963.

10 De Abreu Filho JG; EF De Lira Mucocele of the appendix: appendectomy or colectomy? J Coloproctol. (Rio J.) 2011;31. Available on http://www.scielo.br/scielo.php?pid=S2237-93632011000300008&script=sci_arttext.

11 Harshen R, Jyothirmayi R, N Mithal Pseudomyxoma Peritonei. Clin Oncol (2003) 15: 73–77.

12 de Oliveira AM, Rodrigues CG, Borges A. Pseudomyxoma peritonei: a clinical case of this poorly understood condition. Int J Gen Med. 2014; 7: 137–141.

13 Renehan AG, Stern PL, O'Dwyer ST, Oliveira AM, Rodrigues CG, Reis Borges A, *et al.* Pseudomyxoma peritonei. Encyclopedia Cancer; 2003; 27:1089–1103.

14 Misdraji J, Yantiss RK, Graeme-Cook FM, Balis UJ, Young RH, Appendiceal mucinous neoplasms: a clinicopathologic analysis of 107 cases Am J Surg Pathol 2003; 27: 8:1089–1103.

15 Losa F, Barrios P, Salazar R, Torres-Melero J, Benavides M, Massuti T, *et al.* Cytoreductive surgery and intraperitoneal chemotherapy for treatment of peritoneal carcinomatosis from colorectal origin. Clin Transl Oncol 2014;16:128–140.

16 Levesque, R SPSS programming and data management: a guide for SPSS and SAS users, 4th ed. (2007), SPSS Inc., Chicago IL.

17 Yalcin S, Ergül E, Korukluoglu B, Yalcin B Pseudomyxoma peritonei: what do we have?. Int J Surg. 2007;15. Available on http://ispub.com/IJS/15/1/8098#.

18 Smeenk RM, Verwaal VJ, Zoetmulder FAN Pseudomyxoma peritonei. Cancer Treat Rev 2007; 33:138–145.

19 Mohamed F, Cecil T, Moran B, Sugarbaker P A new standard of care for the management of peritoneal surface malignancy. Curr Oncol 2011;18.

20 Hayes-Jordan A, Anderson P, Curley S, Herzog C, Lally KP, Green HL, *et al.* Cancer Treatment Reviews 2007;33:138–145.

21 Blackham AU, Levine EA Cytoreductive surgery with hyperthermic intraperitoneal chemotherapy for malignant peritoneal mesothelioma. Available at: http://www.science.gov/topicpages/c/continuous+hyperthermic+peritoneal.html. [Accessed 1 January 2013].

22 Idjuski S, Turkalj I, Petrovic K, *et al.* Pseudomyxoma peritonei due to mucinous adenocarcinoma of the appendix. JBR-BTR 2013; 96:184.

23 Sugarbaker PH Pseudomyxoma peritonei: a cancer whose biology is characterized by a redistribution phenomenon. Ann Surg 1994; 219:109–111.

24 Deraco M, Baratti D, Inglese MG, Idjuski S, Turkalj I, Petrovic K, Vanhoenacker FM. Peritonectomy and intraperitoneal hyperthermic perfusion (IPHP): a strategy that has confirmed its efficacy in patients with pseudomyxoma peritonei. Ann Surg Oncol 2004; 11:393–398.

25 Faris JE, Ryan DP, Marcello D, Dario B, Maria G, Biagino A, *et al.* Controversy and consensus on the management of patients with pseudomyxoma peritonei Curr Treat Options Oncol 2013;14:365–373.