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Case Report

Intestinal Obstruction Secondary to Pseudomyxoma Peritonei: A Case Report

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Abstract:

Introduction

Pseudomyxoma Peritonei (PMP) is a rare neoplastic syndrome characterized by disseminated cancerous mucinous lesions and/or mucinous ascites in the intraperitoneal area, peritoneal surface and omentum, principally related to appendiceal neoplasm. Patients are usually asymptomatic or paucisymptomatic, complaining nonspecific symptoms, including abdominal pain and distension, anorexia and nausea. PMP slow and uninterrupted growth leads to significant abdominal distension and complications such as intestinal obstruction and dyspnea. Preoperative diagnosis is based on ultrasonography and computed tomography; nonetheless, explorative laparotomy and histopathological exam provide the ultimate diagnosis. Cytoreductive surgery (CRS) associated with heated intraperitoneal chemotherapy (HIPEC) is considered a potentially curative treatment in selected patients.

Case presentation

We present an unusual case of a male patient, aged 58 years-old, hospitalized in the Surgery Department of "Jazzolino" Hospital of Vibo Valentia (Italy) for intestinal obstruction secondary to intraperitoneal dissemination of PMP. The patient, who did not present common risk factors for abdominal district tumors, had been complaining of dyspespisa and important abdominal distension for some months, until he arrived at the emergency room with widespread abdominal pain, closed hive, batracian abdomen and recurrence of a previous umbilical hernia. His blood test resulted normal; CEA = 10,2 ng/dL (range 0,00 to 5,10); Ca 19.9 within limits. Abdominal ultrasound and CT scans suggested PMP on an appendicular base. The patient underwent explorative laparotomy with findings of ascites, abundant gelatinous material in the subhepatic site and in the Douglas fissure, disintegration of the epiplons, covered by the same gelatinous material, and of the wall of the ileal loops, incorporated and with a gelatinous-like consistency. Samples of the ascitic effusion and gelatinous material were taken for cytological exam, while biopsies of the omentum were sent for histological exam, which confirmed the suspected diagnosis. Once stabilised, the patient was referred to a specialised centre for further treatment.

Conclusions

PMP is rare and difficult to detect. Imaging guides preoperative diagnosis, although the final diagnosis is reached through explorative laparotomy and histopathology exam.

Complete CRS associated with HIPEC is considered to be the most suitable approach in selected patients. As an advanced disease and/or inadequate treatment lead to poor prognosis, patients diagnosed with PMP should be referred to specialised centres for further care.

Key words: psychodermatology; dermatopsychology; trichotillomania; mental Health; treatments in psychodermatology

Introduction

Pseudomyxoma Peritonei (PMP) is a rare neoplastic syndrome with a reported incidence of 1 in 1.000.000 cases/year or 2 in 10.000 laparotomies/year, with a ratio of men to women equal to 1 to 2/3 [1,2,3].

PMP is characterized by disseminated cancerous mucinous lesions and/or mucinous ascites in the intraperitoneal area, peritoneal surface and omentum, with or without detection of neoplastic cells [4,10]. It is principally related to appendiceal neoplasm, though it may rarely be a consequence of ovaric, colonic or biliary mucinous tumors [4,6]. Primary neoplasms are present in less than 2% of surgical appendectomy specimens [8].

Patients are usually asymptomatic or paucisymptomatic, complaining nonspecific symptoms, including abdominal pain and distension, anorexia and nausea [3,4,8]. As a result of its indolent nature, PMP grows uninterrupted until the slow accumulation of mucin into the peritoneal cavity leads to significant abdominal distension and common complications, such as intestinal obstruction and dyspnea [6,8].

Preoperative diagnosis is based on ultrasonography and computed tomography; nonetheless, the final diagnosis is established intraoperatively, thorough an explorative laparotomy, and confirmed by the histopathological exam [6].

The association of cytoreductive surgery (CRS) and heated intraperitoneal chemotherapy (HIPEC) is considered a potentially curative treatment in selected patients, despite clear evidence is lacking [7,9,10,11]. This combined approach has been reported to allow a 15-years survival rate of 59% and a progression-free survival of 8,2 years [5,10].

Case Presentation

We present an unusual case of a male patient, aged 58 years-old, hospitalized in the Surgery Department of "Jazzolino" Hospital of Vibo Valentia (Italy) for intestinal obstruction secondary to intraperitoneal dissemination of PMP.

The patient, with a family history of lung cancer, was previously subjected to radiometabolic therapy for hypertiroidism, had a personal past pathological history negative for other noteworthy disease and did not present common risk factors for abdominal district tumors. For some months he had been complaining of dyspespisa and important abdominal distension, without, however, undergoing a medical examination and clinical test.

The patient arrived at the emergency room with widespread abdominal pain, closed hive, batracian abdomen and recurrence of a previous umbilical hernia.

His blood test resulted normal; CEA = 10,2 ng/dL (range 0,00 to 5,10); Ca 19.9 within limits.

Abdominal ultrasound suggested the presence of mucin, while abdominal CT with contrast revealed fluid with superfluid density, fine internal septations and some calcifications [Figure. 1], thickening and cross-linking of the omental fat (omental cake) [Figure.2], collapsed jejuno-ileal loops in the central abdominal site [Figure. 3] and a tubular image, inseparable from the caecal fundus, with superfluid content and calcifications in the right iliac fossa (RIF) [Figure. 4], suggesting PMP on an appendicular base.

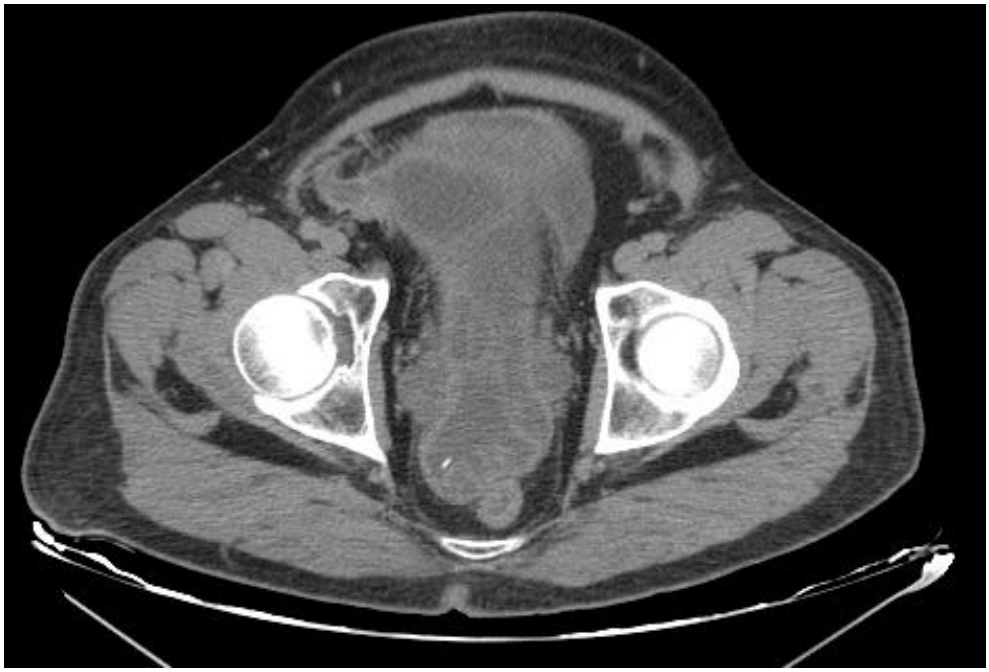

Figure 1: Pelvic axial CT scan showing evidence of fluid with superfluid density, fine internal septations and some calcifications

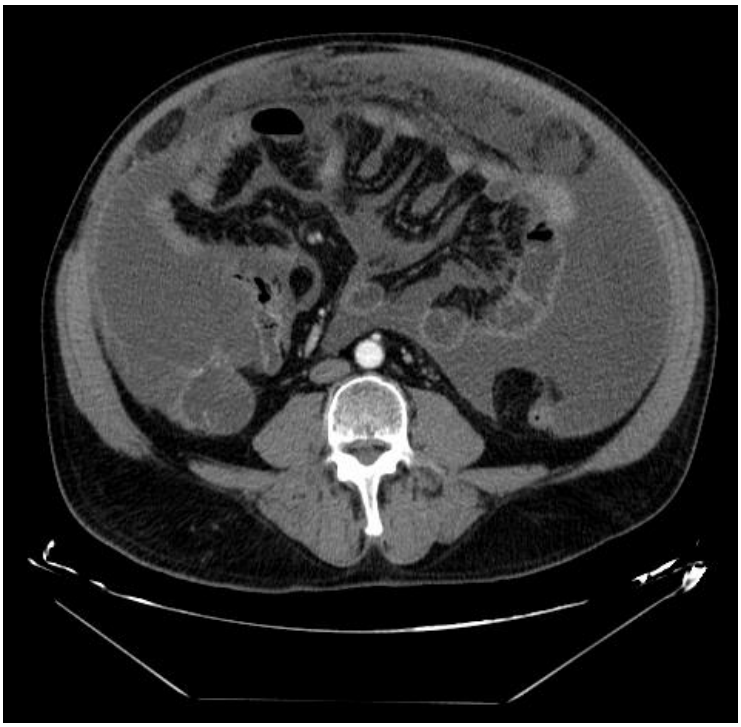

Figure 2: Axial CT scan showing thickening and cross-linking of omental fat (omental cake)

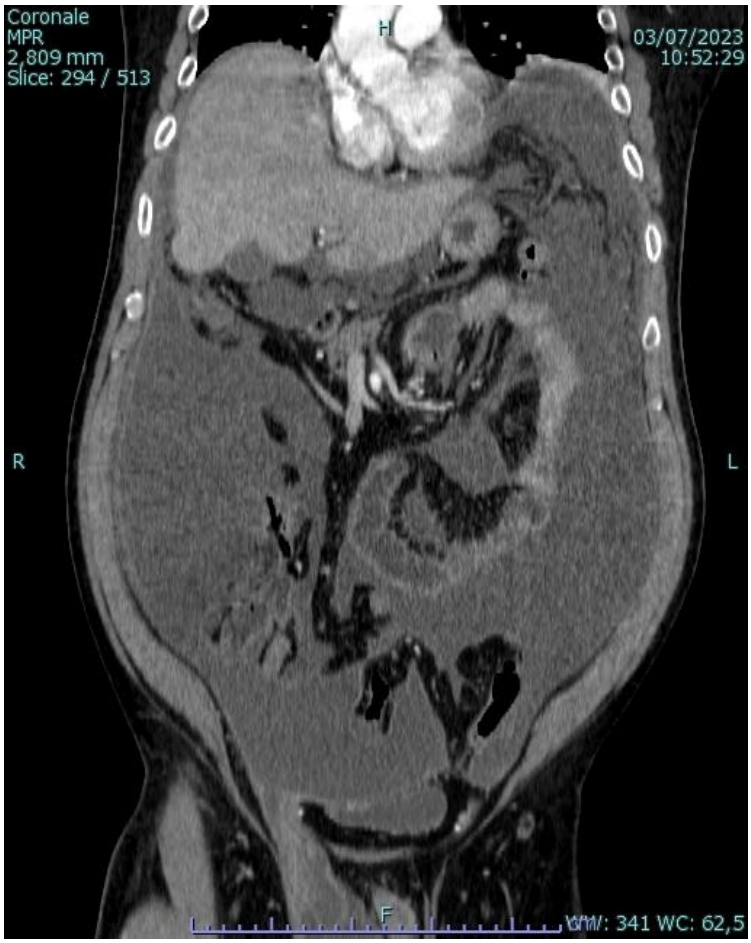

Figure 3: Coronal CT scan with collapsed jejuno-ileal loops in the central abdominal site

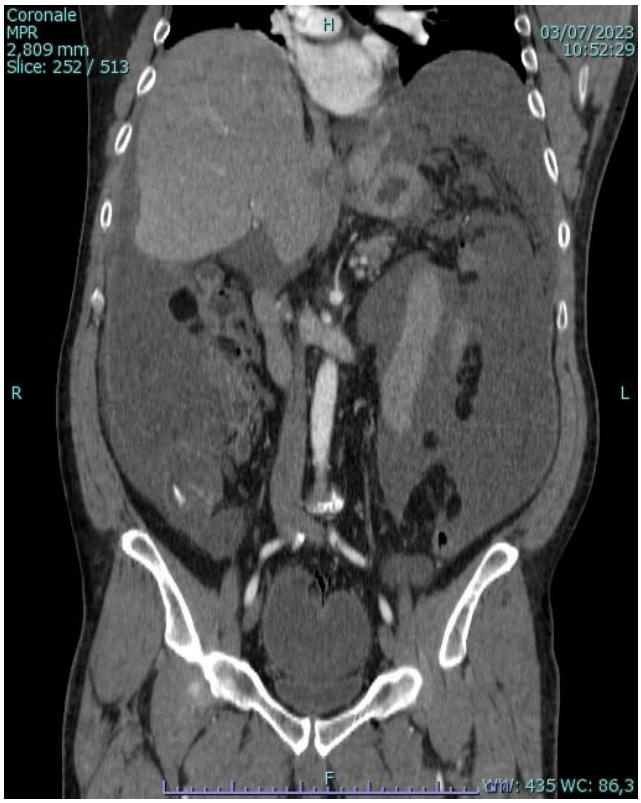

Figure 4: Coronal CT scan showing a tubular image, inseparable from the caecal fundus, with superfluid content and calcifications in RIF

The patient underwent explorative laparotomy with findings of ascites, abundant gelatinous material in the subhepatic site and in the Douglas fissure, disintegration of the epiplons, which appeared covered by the same gelatinous material (Figure. 3,4), and of the wall of the ileal loops, incorporated and with a gelatinous-like consistency.

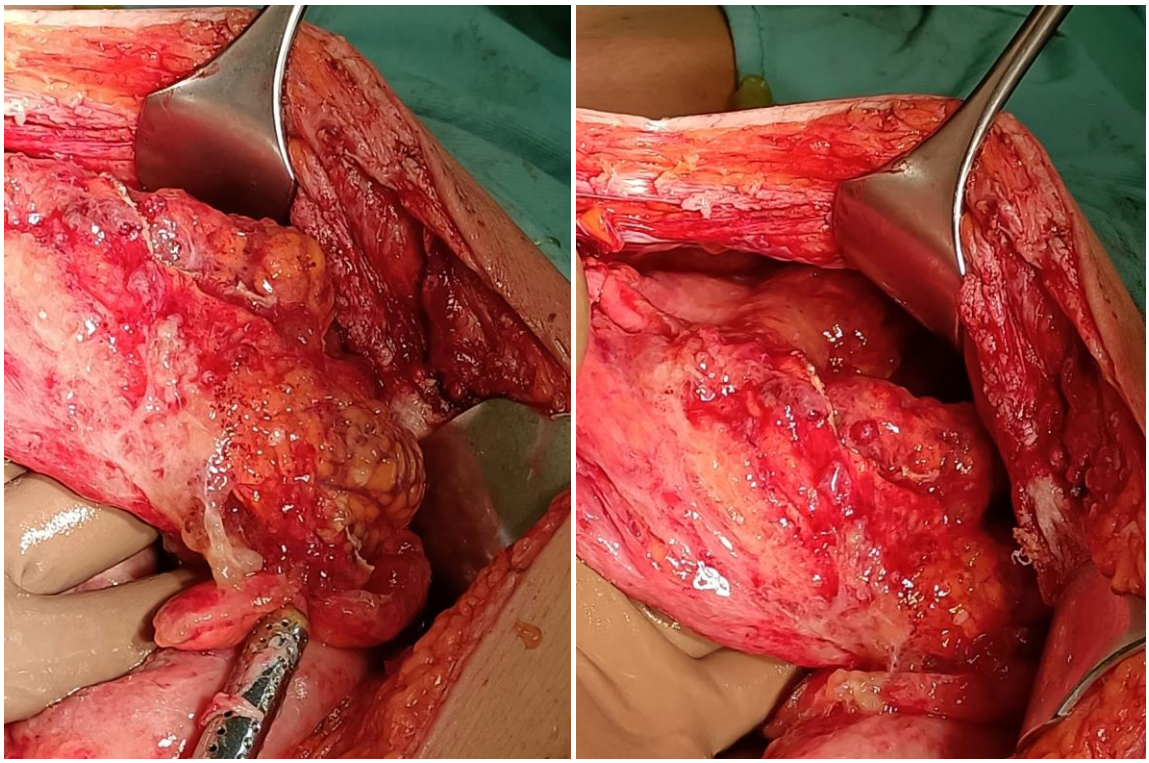

Figure 3, 4: Subverted omentum, covered by mucoid secretion

Samples of the ascitic effusion and gelatinous material (Figure. 5) were taken for cytological exam, while biopsies of the omentum (Figure. 6) were sent for histological exam.

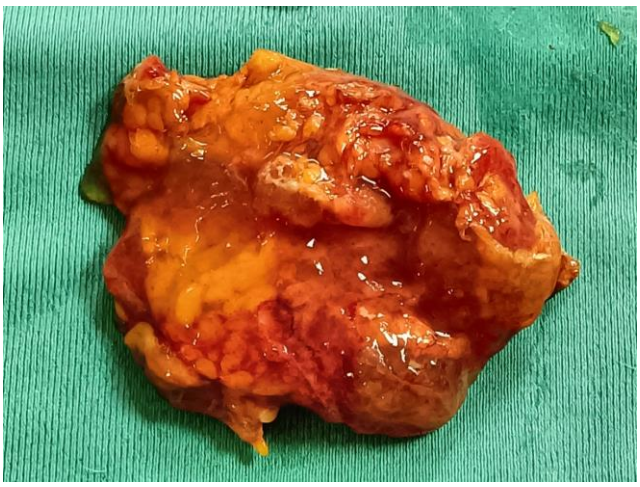

Figure 5: Mucus

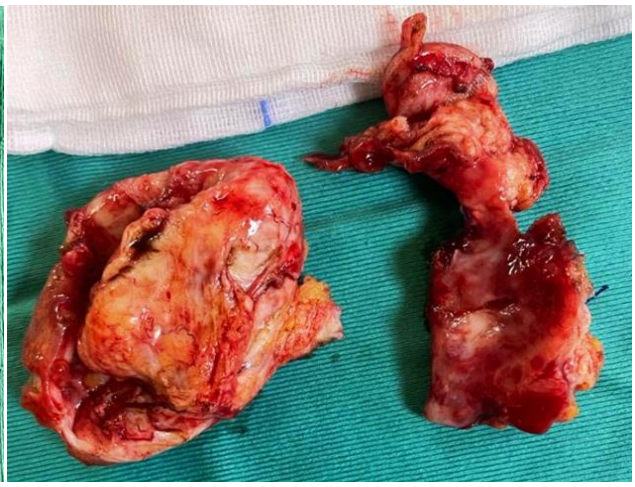

Figure 6: Peritoneal biopsies

The cytological exam conducted on the ascitic liquid revealed mucus without neoplastic cells (Figure. 7), while the histologic diagnosis was of low grade appendiceal mucinous neoplasm (staging pT4a, WHO Digestive System Tumor 2019) (Figure. 8, 9, 10, 11).

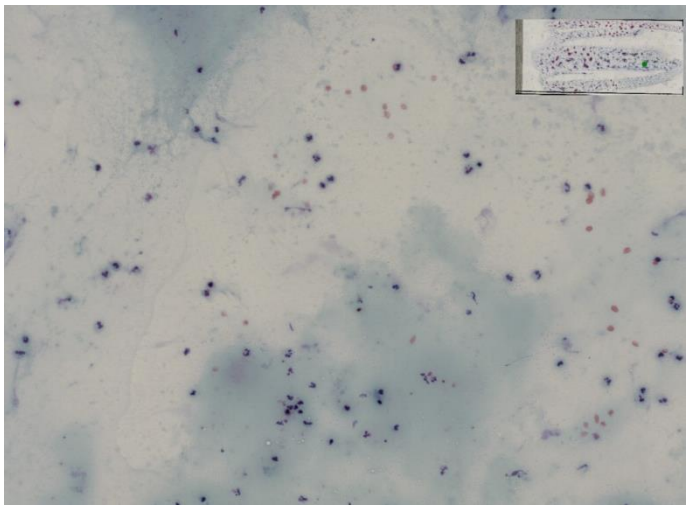

Figure 7: Mucin, neutrophils and rare red blood cells (Papanicolau-10X - Leica - Aperio LV1)

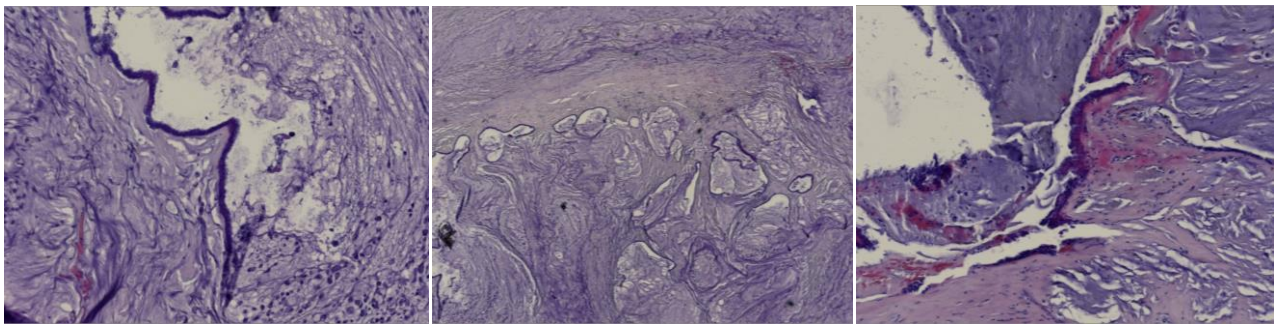

Figure 8,9,10: Fibrous tissue lined by neoplastic glands of low grade atypia admixed with abundant mucoid secretion devoid of neoplastic cells. (EE-2.5X - 10X Leica - Aperio LV1)

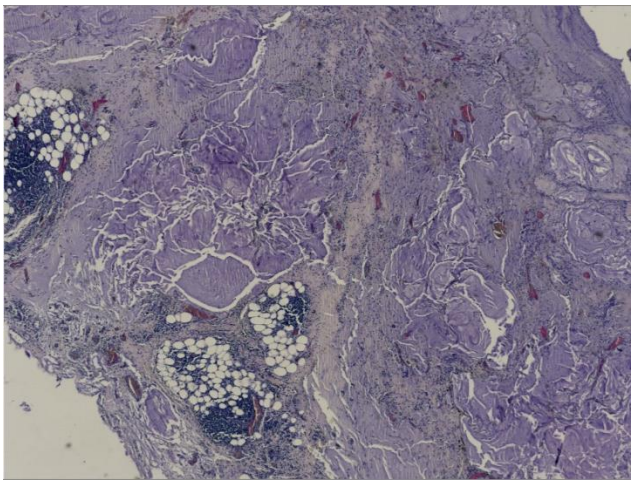

Figure 11: Peritoneal fibroadipose tissue with abundant mucin lake devoid of neoplastic cells. (EE-10X Leica - Aperio LV1)

Once stabilised, the patient was referred to a specialised centre for further treatment.

Discussion

PMP is an uncommon disease determined by a slow and continuous intraperitoneal dissemination of cancerous mucinous cells secondary to a rare adenomucinous tumor originating from the appendix, ovaries, colon, rectum or bile duct [1,4,6,10]. Recent immunohistochemistry and molecular genetics studies demonstrated both the appendiceal and ovarian etiology involvement in the determinism of this pathology [6]. As for adenomucinous neoplasm of the appendix, its cells progressively secrete abundant intraluminal mucus, causing an obstruction of the appendiceal lumen, an increase in intraluminal pressure and thus the appendiceal mucocele that results in a slow leak of mucus, containing mucinous epithelial cells, into the peritoneal cavity[1,2,8]. As in our case, the perforation of the appendix may reseal, while, over the course of months/years, free epithelial cells in the peritoneal cavity continue to proliferate and produce mucinous ascites, spreading the cells from the ruptured appendiceal neoplasm throughout the peritoneal cavity [1,2,8]. Tumor deposits are found especially in the subhepatic region, under the right diaphragm, in the omentum, in Douglas pouch, around the rectum and sigmoid, and, in women, on the ovarian surface; invasion of the peritoneal surface is usually absent [1,2,8]. The spread of mucous and epithelial cells into the peritoneum caused by the possible rupture of the mucocele is more likely to happen during laparoscopy, due to manipulation, grasping and pneumoperitoneum, and associates with a poorer prognosis [6].

Preoperative diagnosis is based on ultrasonography and computed tomography; nonetheless, explorative laparotomy and histopathological exam provide the ultimate diagnosis [6]. Tumor markers CEA and CA 19.9 have diagnostic and prognostic value [6].

The lack of more concrete data on the survival of patients with PMP cause uncertanty about treatment options, although most authors agree in considering the association of CRS and HIPEC (cisplatinum) a potentially curative treatment in selected patients [7,9,10,11].

Conclusions

PMP is an infrequent clinical condition presenting with scarce, ambiguous and often late symptoms. Radiological studies guide to preoperative diagnosis, though it is surgery that provides the final diagnosis, confirmed by the histopathology exam.

Complete CRS associated with HIPEC has been proven to be a valid approach in selected patients. As an advanced disease and/or inadequate treatment lead to poor prognosis, patients should always be referred to specialised centres for further care.

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