RELAPSE OF THE PSEUDOMYXOMA PERITONEI AFTER A CYTOREDUCTIVE SURGERY WITH PERITONECTOMY AND HIPEC, 10-YEAR FOLLOW-UP - CASE REPORT WITH A LITERATURE REVIEW

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ABSTRACT

Background: Pseudomyxoma peritonei is a rare clinical condition characterized by mucin-secreting epithelial cells which lead to formation of jelly-like structures within the peritoneal cavity and the accumulation of mucinous ascites. Most commonly it arises from the intra-abdominal spread of appendiceal mucinous tumors. Few therapeutic options exist, but a combination of cytoreductive surgery and hyperthermic intraperitoneal chemotherapy, as described by Sugarbaker, stands for the treatment of choice in many tertiary centers nowadays.

Case study: We present a 62-year-old female patient who was initially presented as acute appendicitis. Later pathohistological diagnosis of Pseudomyxoma peritonei just confirmed intraoperative suspicions. Definitive diagnosis was followed with a complete cytoreductive surgery and hyperthermic intraperitoneal chemotherapy. After a 10 year relapse-free follow up, she presented with colon adenocarcinoma and a recurrence of jelly-like incapsulated structures within the abdominal cavity.

Conclusion: In patients diagnosed with pseudomyxoma peritonei, according to current findings, best results are achieved using complete cytoreductive surgery and hyperthermic intraperitoneal chemotherapy. However, recurrences still do occur and there is no real consensus regarding their optimal treatment.

Keywords: Pseudomyxoma peritonei, cytoreductive surgical procedures, hyperthermic intraperitoneal chemotherapy

Introduction

Pseudomyxoma peritonei is a rare clinical condition with unknown cause and estimated overall incidence of 1-2 per million [1]. The primary tumor usually originates from the appendix, but can also appear in ovarium, urinary bladder or any other part of the gastrointestinal system [2]. Female patients are more likely affected. Synchronous tumors of appendix and ovaries were described, but recent immunohistochemical studies suggest that „synchronous“ ovarian tumor is actually a metastatic process of primary appendiceal tumor [3]. Pseudomyxoma peritonei begins as an appendiceal adenoma which starts to produce mucus intraluminally and consequently leads to obstruction and rupture of the appendix. Therefore, tumor cells are spread intraperitoneally, where they continue with their mucus production. Pseudomyxomal tumorous cells tend to characteristically accumulate around the liver, omentum and in the lower pelvic region, unlike some other gastrointestinal tumor metastases which usually incorporate close to their perforation site. As the tumor progresses, metastatic cells spread to entire abdomen, but there is always absence of tumor masses on the free portion of the small intestine, which is described as a pathognomonic sign [4, 5]. An intraabdominal accumulation of jelly-like masses causes unspecific symptoms like abdominal or pelvic pain, distension of abdomen, and infertility. After a long period of accumulation, eventually, mechanical obstruction of the gastrointestinal tract occurs. Because of that, early diagnosis is difficult to make and it is often misdiagnosed. Differential diagnosis in female patients is even harder as it includes ovarian tumors, which can also produce mucus and cause distension of the abdomen with similar symptoms. Therapy of pseudomyxoma peritonei depends on the tumor size and a stage of the disease. In the majority of patients, long term prognosis is poor and depends upon the histological type of tumor with 50% of 5-year survival [6]. When possible, cytoreductive surgery combined with intraperitoneal chemotherapy shows best long-term results [7]. Other therapy options include „debulking“ surgery, classic intravenous chemotherapy or expectative follow-up. We report a case of a female patient diagnosed with pseudomyxoma peritonei who was treated by complete cytoreductive surgery and peritonectomy accompanied by hyperthermic intraperitoneal chemotherapy (HIPEC). The patient had uneventful recovery in a 10-year relapse-free follow up but then presented with colon adenocarcinoma and a recurrence of jelly-like incapsulated structures within the abdominal cavity.
Case study
A 62-year old female patient presented to the emergency department with a seven-day history of constant periumbilical pain, localized to the right lower quadrant. 8 months prior to arrival, she experienced the same symptoms which regressed spontaneously. Her medical history revealed hypertension and uterine myoma. By the time of examination, she did not feel nausea, anorexia, neither had diarrhea. Physical examination revealed an obese female patient with rebound tenderness over the right lower abdominal quadrant. Vital signs were within normal limits (RR 140/90mmHg, BPM 78/min). Laboratory findings showed an elevated white blood count (11,21±109/L) and CRP (80mg/L). The ultrasound study showed a collection of dense fluid at the ileocecal region, which was described as an abscess formation. The plain abdominal radiograph showed no signs of pneumoperitoneum. Pararectal laparotomy revealed the intraabdominal cavity fulfilled with gelatinous masses and numerous cystic lesions, which covered peritoneum and liver. Appendix was thickened >3 cm in the distal part but showed no signs of inflammation or rupture. Appendectomy was done with pathohistological diagnosis of carcinoïd tumor of the appendix and peritoneal carcinomatosis. For the next 34 months she was treated conservatively with an obvious deterioration of her clinical condition and progression of the radiological findings (Figure 1.). Her main complaint was distension of the abdomen accompanied with anorexia and constipation (Figure 2.).

Laparotomy was done, intraabdominal jelly-like masses (Figure 3.) were evacuated and complete peritoneectomy together with cytoreductive surgery followed by hyperthermic intraperitoneal chemotherapy (mitomycin/cisplatin) was undertaken. The pathohistological diagnosis was pseudomyxoma peritonei.

Postoperative period passed uneventfully and the patient was in regular surgical and oncologic follow up for the next 10 years. During that period she was asymptomatic and felt occasional nausea and bloating. Because of sideropenic anemia, a diagnostic colonoscopy was undertaken, which revealed a stricture of the ileocecal anastomosis. Laparotomy showed infiltrating process at the location of the previously formed anastomosis. The new anastomosis was formed. Tumorous process was resected in a series of less aggressive palliative procedures, because of intraabdominal adhesions formation with high-grade tumors. The overall 10-year survival was 23% [8]. On the other hand, Sugarbaker et al. analyzed a similar group of patients, who were treated by series of less aggressive cytoreductions (average 2.2 surgeries per patient) and with selective usage of intraperitoneal chemotherapy with a median survival of 12.8 years for patients with low-grade tumor and only 4-year survival for patients with high-grade tumors. The overall 10-year survival was 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19].

The pathohistological diagnosis was mucinous and lipoma.

Discussion
In the past, patients diagnosed with pseudomyxoma peritonei were treated by surgical reduction of the tumor and repeated drainages of mucinous ascites, with poor results and common relapses, which resulted in low quality of life. [8]. In 1980, Spratt et al. introduced hyperthermic intraperitoneal chemotherapy as a new therapy option [9]. A few years later, Sugarbaker proposed treatment of pseudomyxoma peritonei with the combined procedure, which included cytoreductive surgery accompanied by hyperthermic intraperitoneal chemotherapy [10]. Despite a lack of multicentric prospective randomized studies, literature published during the last two decades confirms the efficiency of the combined procedure, which became the treatment of choice in many tertiary centers worldwide nowadays [11-16]. It is usually used for appendiceal low-grade tumors with peritoneal dissemination [17], with overall 10-year survival of 63% [18]. Miner et al. questioned the efficiency of complete cytoreductive surgery combined with HIPEC because of its radical approach and variety of perioperative complications. The study included a group of patients treated by series of less aggressive cytoreductions (average 2.2 surgeries per patient) and with selective usage of intraperitoneal chemotherapy with high-grade tumors. The overall 10-year survival was 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19]. The success of combined therapy lies in primary aggressive cytoreduction which tends to totally eradicate tumorous mass in just one radical surgical procedure which is followed by intraperitoneal chemotherapy and showed 20-year survival of 70% [19].
therapeutic problem. Delhorme et al. analyzed a tumor recurrence in a group of patients treated by primary cytoreductive surgery and HIPEC. During an 85 months follow-up 26% of patients had a tumor relapse, with a median of 25 relapse free months. In the majority of cases (76%), primary relapse was localized intraperitoneally, as it is described in the reported case [20]. Even though the treatment of intraperitoneal recurrences is still debatable, redo surgery for isolated intraperitoneal recurrence is proposed by most of the authors [21-25]. There is an absence of consensus regarding redo HIPEC, considering its failure to prevent relapse of the disease. With no doubt, large randomized studies are necessary to define which groups of patients would have a benefit and should be treated by cytoreductive surgery and HIPEC. Described patient fits in a group of successfully treated patients with the combined method, despite intraabdominal status worsening and disease relapse after a 10-year follow-up.

Conclusion

There is a lack of evidence based guidelines which would define the best treatment options for patients diagnosed with pseudomyxoma peritonei. According to current findings, complete cytoreductive surgery and hyperthermic intraperitoneal chemotherapy is a treatment of choice nowadays. A fact that pseudomyxoma peritonei is a rare clinical condition shouldn't lessen a need for further investigations which will lead to clear guidelines based on strong evidence.

The authors declare that there is no conflict of interest.

REFERENCES

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Figure 1.
A) Preoperative MSCT of abdomen and pelvis showing diffuse collections of gelatinous material and mucus around liver, spleen, and intestines. B) Postoperative MCST of abdomen and pelvis after a complete cytoreductive surgery with HIPEC.

Figure 2.
On the left preoperative picture of the patient with massive distension of the abdomen. On the right postoperative picture of the patient with uneventful recovery.
Figure 3.
Intraoperative picture showing evacuated gelatinous material and mucus.

Figure 4.
Abdominal CT scan of well defined, lobulated cyst in omental bursa which is in close contact with the pancreas.

Figure 5.
A) Intraoperative photo of the cyst located in retrogastric area protruding through the hepatic recess. B) Photo of the surgical specimen after extirpation and opening which revealed mucinous material.