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CASE REPORT

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Pseudomyxoma peritonei: a case report and current concepts in the literature

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Abstract This report evaluates the natural history and clinicopathologic variables of patients with pseudomyxoma peritonei (PMP), and reviews the authors' experience with different treatments in the current literature. PMP results from implantation of malignant tumors or peritoneal irritation caused from ruptured benign cysts. This entity is traditionally characterized by the accumulation of mucinous ascites and a relatively long survival period. Diffuse peritoneal spread occurs in most patients with PMP, and distant metastasis is infrequent. Disease progression is difficult to predict because of the spectrum of underlying pathologic processes with varying malignant potential. Debulking surgery, radiation therapy and chemotherapy have all been advocated for optimal patient management.

Key words Pseudomyxoma peritonei • Mucinous tumor Appendix

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Introduction

Pseudomyxoma peritonei (PMP) is a rare clinical condition characterized by the accumulation of mucinous ascites, a relatively long survival period and, uncommonly, extraperitoneal metastases [1, 2]. Generally, PMP is a slowly progressive and low-grade malignant process; aggressive surgical approach with resection of the bulk of diseased tissue offers optimal palliation and prognosis [3, 4]. This report presents a case of PMP with an enlarging mass with gelatinous material in the abdomen and a review of the current literature.

Case report

The patient was a 51-year-old male who presented with a history of fatigue, weight loss and abdominal distention for two years. He had been evaluated for suspected hepatic fibrosis and tuberculous peritonitis with unrewarding results. He was referred to our department from the State Hospital.

Physical examination revealed a pale and dehydrated patient with cachectic appearance. The abdomen was non tender, but was extremely distended with normoactive bowel sounds. Percussion revealed matity all over the abdomen. On palpation no organ was enlarged. On auscultation of the lungs, expiration was prolonged and diffuse rallies were heard at the base of the right lung.

Abnormal laboratory findings included elevated carcinoembryonic antigen (CEA) levels (28 ng/ml), and decreased levels of total protein (5.5 mg/dl) and albumin (2.4 mg/dl). Complete blood cell count and the other routine biochemical analyses were in the normal ranges. Chest radiography and abdominal direct X-ray revealed elevation of the diaphragm on the right. Abdominal ultrasonographic fidings of the patient were reported as hydatid disease filling the entire abdomen.

Computed tomography (CT) revealed massive ascites of fat density with scattered masses of soft tissue density within

it and areas of thickened peritoneum (Fig. 1). These were interpreted as tomographic findings of peritoneal carcinomatosis or tuberculous peritonitis. Abdominal paracentesis, performed to obtain ascites for microbiologic and cytochemical analyses, revealed semisolid gelatinous material without cells.

We decided to perform explorative laparotomy for definite diagnosis. At operation, the abdomen was found to be entirely filled with semiliquid gelatinous mucoid material. Many sites of peritoneum were thickened and implanted with mucoid material. There were also mucinous implants on the right diaphragm and adhesions between peritoneal thickenings and intra-abdominal and intrapelvic structures. Biopsies taken from these peritoneal thickenings, implants and mucoid material were sent for frozen-section pathologic analysis. On pathologic examination, glandular epithelial cells were found and the result was reported as pseudomyxoma peritonei (Fig. 2). In order to find the probable origin of the condition, we tried to explore the ileocecal region to identify the appendix: however, there were so many tight adhesions that it was not possible to reach the appendix without deteriorating the patient's condition and increasing the operative morbidity. As far as could be evaluated, the other intra-abdominal organs were normal. Following debulking of the semisolid mucoid intra-abdominal mass, the peritoneal cavity was irrigated and closed.

The patient recovered uneventfully. He was discharged on the seventh postoperative day and sent to the Department of Oncology for systemic chemotherapy with 5-fluorouracil. He is alive and free of tumor after 8 months. He will be re-explored to carry out appendectomy and, if necessary, a second debulking.

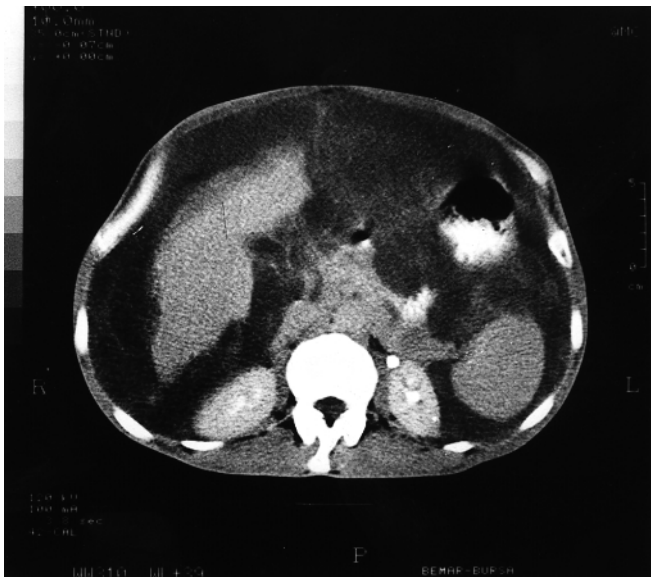

Fig. 1 Computed tomogram showing massive ascites of fat density with scattered masses of soft tissue density within it

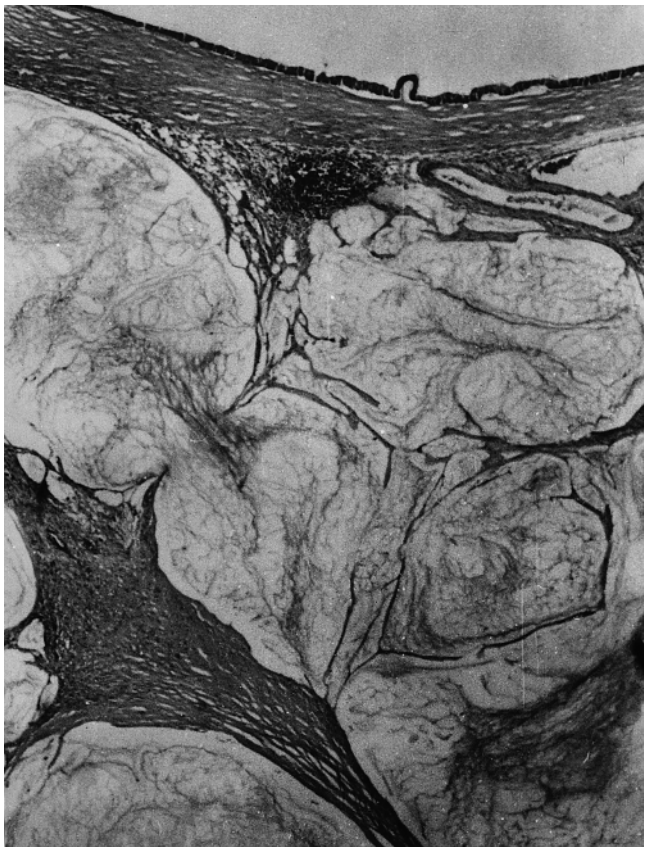

Fig. 2 Low-power view of mucin-containing loculations separated by fibrous bands. Some of them are lined by pseudostratified glandular epithelial cells

Discussion

The presence of gelatinous material in the peritoneal cavity from a benign ovarian cystadenoma was first described by Werth in 1884 [5]. In 1953, the cause of pseudomyxoma was postulated to be an appendiceal mucinous adenocarcinoma and not only obstruction of appendiceal lumen with resultant mucocele [6]. It is now generally agreed that the term PMP should be restricted to describe the condition of those patients in whom peritoneal mucus is found in the presence of an intraperitoneal adenocarcinoma [4-9]. PMP arises from mucin-producing adenocarcinomas of the appendix and the ovary in the majority of cases, but there are reports of PMP occurring in association with pancreatic carcinoma, carcinoma of the stomach, carcinoma of the breast, and bile duct cancer [1, 5].

According to two physiologic mechanisms, described by Sugarbaker [10], the tumor will progress by the production of mucus, exfoliation of tumor cells, and a redistribution of these cells around the abdomen. First, the abdominal surfaces that absorb peritoneal fluid, such as the greater omentum and underface of the diaphragm, are coated by large numbers of tumor cells as fluid is concentrated during

months and years. The other mechanism of tumor redistribution is simply gravity. Free-floating intraperitoneal tumor cells will puddle in large volumes within the pelvis, within the right subhepatic space, within the left abdominal gutter, and at the ligament of Treitz [10].

The incidence of PMP is approximately 2 in 10 000 laparotomies [11]. On the other hand, malignant tumors of the appendix are rare accounting for 0.2%- 0.5% of all tumors of the gastrointestinal tract [12]. The mean age of patients with PMP originating from the appendix is 46 years compared with a mean of 59 years in those with ovarian primary cancer [3]. The most common symptoms and findings are abdominal pain, distention, palpable mass and weight loss, which occurs during a period of months to years; abdominal distention occurs late in the disease course [1, 3, 5-6].

Laboratory tests are of little value diagnostically. Some degree of anemia may be present. The CEA level is sometimes elevated and may be useful in follow-up evaluations of the patient whose CEA level decreases after resection and tumor debulking [6]. Radiographically, CT and magnetic resonance imaging can be quite helpful and yield a diagnosis of PMP [13, 14]. Barium study of the gastrointestinal tract and ultrasound are less helpful but can be used as screening modalities [6]. The majority of cases are diagnosed surgically, although this situation is changing [5, 9].

Histologic examination shows most malignancies to be low-grade carcinoma [4, 5]. This may account for the indolent behavior, lack of distant metastases, and frequent long-term survival associated with PMP [5]. The tumor does not actively attach to an abdominal or pelvic surface; it does not generally invade neighboring organs [9, 10]. Thus, the low-grade nature of the tumor may limit distant spread but allow local invasion, albeit at a superficial level [5]. However, local spread of PMP by direct extension to the pleural or pericardial space has been reported in the literature, although this is uncommon [15].

For many years the clinical syndrome of PMP has been enigmatic. Particularly, in women with PMP, there has been considerable debate as to whether the ovarian tumors are secondary to the appendiceal tumor or are independent primary ovarian tumors. Recently, Ronnett et al. demonstrated that most ovarian mucinous tumors in PMP were secondary to the appendiceal tumor [16, 17]. Therefore, pseudomyxoma peritonei is currently defined as a grade I mucinous adenocarcinoma that arises from a primary appendiceal adenoma [18].

The surgical approach largely depends upon the extent of disease at exploration, with surgical resection potentially involving the small intestine or a segment of colon [2, 4]. Although the goal for operative treatment is total removal of the tumor, this has been achieved in only 20% in the study by Gough et al. [5]. However, in this study there was no significant difference in the rates of recurrence between those patients who had potentially curative resection of localized disease and those patients who had subtotal debulking of diffuse tumor. Therefore, it is suggested that aggressive surgical debulking followed by radiotherapy and/or chemotherapy should be considered because of the diffuse peritoneal involvement [4, 5, 9, 10].

In the early postoperative period, before adhesions form, intra-abdominal irrigation and chemotherapy are used to destroy the few remaining cancer cells. Adjuvant intraperitoneal chemotherapy with 5-fluorouracil may be effective in controlling disease on the peritoneal surfaces [4, 19]. Systemic chemotherapy has been used in the treatment of PMP for many years. But, currently, many investigators suggest that intraperitoneal chemotherapy is better than systemic chemotherapy on patient survival [4, 5, 10, 12, 19].

Radiotherapy also may be a useful adjunct. In the study by Fernandez and Daly [9], an improvement in the 5-year survival rate from 44% for patients receiving chemotherapy to 75% for patients receiving radiation has been noted. The study by Gough et al. [5] also suggested that patients with PMP should receive radioactive phosphorus or some form of intracavitary radiotherapy, particularly if mucin removal is incomplete after surgery. However, sometimes it may cause fibrosis and obstruction of the intestinal tract [1].

Long-term treatment is directed at complications of obstruction and fistula formation, and repeated laparotomy is often necessary for recurrent symptoms referable to accumulation of mucinous ascites or intestinal obstruction [2]. Recurrent tumor may grow within fibrous adhesions after a surgical procedure; tumor expanding within this fibrous matrix can be associated with infiltration of inflammatory cells and neovascularization and become densely adherent to intra-abdominal structures, such as small intestine, making surgical extirpation more difficult [4].

In four large, previously reported series of PMP, the survival rates at 5 years were 53%-80% [5, 9, 12, 20, 21]. The presence of abdominal distention, a history of weight loss, the presence of diffuse disease or invasion of other organs are associated with shorter survival times [5]. Death as a result of PMP is usually secondary to intestinal and biliary obstruction, fistula formation, peritonitis or pulmonary embolus but not to visceral invasion or metastatic disease [1, 6, 9, 21].

As each reoperation is associated with a more difficult dissection with higher postoperative morbidity and mortality rates, Sugarbaker et al. [12] advocated complete cytoreduction with the use of the required peritonectomy procedures and early postoperative intraperitoneal chemotherapy. The study by Smith et al. involved repeated surgical procedures and systemic chemotherapy [20]. As the Mayo Clinic's experience, Gough et al. used repeated surgical procedures and intraperitoneal radioisotopes or chemotherapy [5].

The disease, when treated by multiple surgical procedures, presents a median survival of approximately 2 years

[18]. Good results depend on early diagnosis and treatment before large volumes of diseased tissue accumulate. In modern therapy using peritonectomy procedures and intraperitoneal chemotherapy with mitomycin C and 5-fluorouracil, the long-term survival at 10 years approaches 80% [18]. As noted by Sugarbaker et al. [21], improvements in the cytoreductive approach await the development of surgical technologies to increase the total clearance of cancer from the abdominal cavity and chemotherapy treatments that are complete enough to sustain control of small-volume residual disease on all peritoneal surfaces. In summary, as a tenacious and indolent process, PMP is a treatable disease; extensive repeated surgical debulking with postoperative intraperitoneal chemotherapy appears to be the treatment of choice.

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