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Case Report

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Treatment of pseudomyxoma peritonei: a case report

Abstract

Pseudomyxoma peritonei (PMP) is a rare disease and even after advances in its understanding and management it often has a protracted course and multiple recurrences despite aggressive surgery and chemotherapy. We report a case of previously healthy woman who present with acute abdomen arising from appendiceal tumor with appendectomy undergone before nine years. Also she underwent emergency surgeries followed by multiple recurrences.

Keywords: pseudomyxoma peritonei, appendiceal mucinous, chemotherapy

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Abbreviations: PMP, pseudomyxoma peritonei; CT, computed tomographic; 5-FU, 5-flourouracil; CEA, carcino embryonic antigen

Introduction

Pseudomyxoma peritonei (PMP) is a rare, chronic, relapsing, diagnostically challenging and poorly understood disease characterized by disseminated mucinous ascites and peritoneal implants usually originating from the appendix or ovaries. This is a rare condition in which cells have spread from the appendix or ovaries into the abdominal or peritoneal cavity resulting in mucinous tumor implants.[1-3] Pseudomyxoma peritonei (PMP) first described by Werth[4] in 1884 by absorbed the massive intraperitoneal accumulation of gelatinous pseudomucin due to the perforation of ovarian pseudomucinous cystomas. It is characterized by abundant extracellular mucin in the peritoneum. The "myxomatous" appearance is attributed to the associated fibroblastic and vascular proliferation that is probably incited by the mucin. This results in multifocal peritoneal, serosal and omental implants admixed with copious amounts of mucin accumulation within the abdomen and pelvis resulting in the belly full. Diagnosis of pseudomyxoma requires a high index of suspicion supplemented by various radiologic studies, including CT scan, followed by pathologic diagnosis[5] and the evaluation of tumor markers. Symptoms may include abdominal or pelvic pain and/or bloating, distension, digestive disorders, weight changes, increased girth and infertility. Treatment for PMP is variable, both due to its rarity and to its frequently slow growing nature. Treatment ranges from watchful waiting to debulking and cytoreductive surgery.[6]

Case report

A 60 years old female with a history of a cramp-like abdominal pain and ruptured appendiceal mucinous that was removed with the omentum 8 years ago (2004) and histopathology of appendicular mucocele with pseudomyxoma peritonei. She was asymptomatic until the fall of 2006 when she admitted with acute abdomen. Initial exploratory laparotomy with total abdominal hysterectomy, bilateral salpingo-oophorectom and omentectomy was done without postoperative chemotherapy. On May 2009 she started complaining of recurrent abdominal pain with mobile nontender epigastric mass. Computed tomographic (CT) of abdomen and pelvis demonstrated a large epigastric mass, it was hypoechoic, laminated architecture, rounded in shape, regular smooth well define outline, located in epigastric region with displacement of the surrounded organs. This mass located between the liver and stomach and measured 10.4x8.9x7.1cm. volume of mass was 343ml. A debulking surgery was performed for remove the mass and tissue diagnosis of PMP was confirmed histopathology (Figure 1-3). Postoperatively, after intravenous administration of many antibiotics, her general condition slowly improved and she left the hospital without complication. Chemotherapy started and after 6 courses of 5-Flourouracil she went into remission. Two years after 5-FU she began complain of severe abdominal pain and diarrhea. CT scan of abdomen and pelvis revealed multiple epigastric masses extended into the pelvic, peritoneum, mesentery and both kidney with elevation of CEA and normal CA19-9. After one cycle of oral capecitabine, she went into partial remission with relieve of abdominal pain and diarrhea.

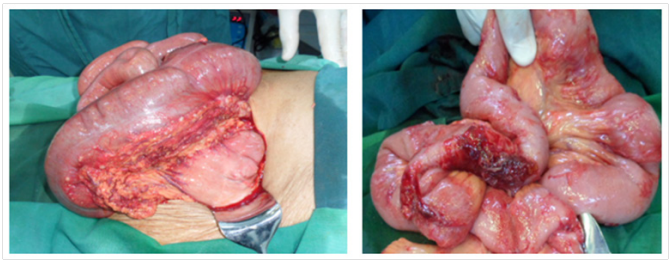

Figure 1 Operative view: Pseudomyxoma peritonea adherent to the intestine.

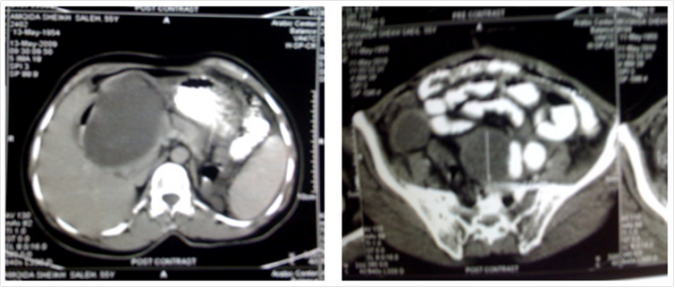

Figure 2 Abdominal CT scan shows collection fluid.

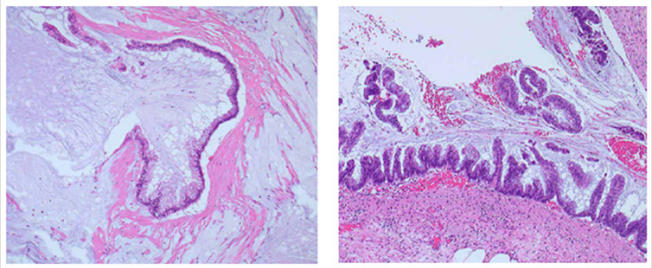

Figure 3A & 3B Fragments of gland in floating in a lake of mucin x10 (3A) and Tall columnar intestinal epithelium with mucin secretion x10 (3B).

Discussion

Pseudomyxoma Peritonei is an uncommon tumor known for its production of mucin in the abdominal cavity.[7] Unlike most cancers, this disease rarely spreads through the lymphatic system or through the blood stream. Therefore it is characterized by mucin and scattered cancer cells in the abdominal cavity. If left untreated, mucin will eventually build up to the point where it compresses vital structures: the colon, the liver, kidneys, stomach, spleen, and pancreas. In the past, pseudomyxoma peritonei was said to occur from a variety of primary tumors.[8,9] This may be true, but in the vast majority of cases, the patients have an appendiceal tumor giving rise to this clinical entity.[9] A precise diagnosis is difficult due to the lack of specific symptoms in the early stage of the disease. Routine laboratory studies are seldom helpful in making this diagnosis. An accurate preoperative diagnosis of pseudomyxoma peritonei can be aided by radiological imaging with computed tomography but remains the final diagnosis being confirmed by histopathology.[1,4]

Mucocele of the appendix was recognized as a pathologic entity by Rokitansky in 1842 and was formally named by Feren in 1876. Appendiceal mucoceles are uncommon entities arising from a variety of different pathologic processes, of which only a small subset are associated with development of pseudomyxoma peritonei.[10,11] Our patient had appendicitis-like syndrome, repeated surgeries and she received systemic chemotherapy, however, she alive with the recurrent mass extended in the pelvic, peritoneum, mesentery and both kidney. Even with a better understanding and recent advances in the management of these cases, PMP remains an enigmatic disease with a protracted clinical course characterized by multiple recurrences over months to years despite surgery and/or chemotherapy.[1] The main goal for treatment of this condition is prevention of locoregional recurrence, rather than systemic disease. Sugarbaker[6] has suggested that a combination of surgery and complete cytoreduction should be followed by intraperitoneal rather than intravenous chemotherapy. Systemic chemotherapy if beneficial is likely to have a transient response and is primarily recommended for patients with extensive peritoneal disease and high grade cystadenocarcinoma.[12,13] Prognosis in this disease is closely related to the bulk of the disease as evaluated by the tumor site, preoperative tumor volume and completeness of tumor removal by surgery and the microscopic degree of differentiation of the neoplastic epithelium as evaluated by the histopathological examination.[14]

In this context, PMP patients with pre-operative elevated tumor markers such as CEA (carcino-embryonic antigen) and CA 19-9 are at increased risk of developing recurrent disease despite aggressive therapy. Likewise, PMP patients with normal levels of these tumor markers have an overall improved prognosis.[15-17] Sugarbaker PH[16] has shown that repeated surgeries result in a median survival of approximately 2 years, with only a small percentage alive at 5 years. Patients who have repeated surgeries and extensive systemic chemotherapy show some improvement in survival, but no long-term, disease-free survival is expected. Fernandez et al.,[17] reported 5- and 10-year survival rates of 54% and 18% respectively.[17] That means that many patients die after the fifth year. Most studies are too recent to evaluate the real efficiency of the various modes of treatment. It is imperative that histological features are taken into account. This is not the case with most available studies; nevertheless, certain trends have been observed, and if debulking surgery with iterative operations are still widely performed, ultra radical surgery could be an attractive option. Another observation significance in this patient that she received systemic 5-fluorouracil-based chemotherapy alternative with capecitabine with objective response. This supports strongly the value of an aggressive locoregional surgical approach followed by systemic chemotherapy.

Conclusion

a. PMP is a condition characterized by mucinous ascites and peritoneal implants, now thought to be of appendiceal origin. Surgical debulking is the recommended treatment both for primary and recurrent tumours.

b. The surgeons should be alert to treating mucinous neoplasm of the appendix, with special care being directed towards adequate excision and through debridement at the initial diagnosis.

c. The periodic post operative abdominal sonography or CT scan and tumor marker laboratory test are used to monitor the disease for any tumor regrowth.

d. Chemotherapy with 5-Flourouracil or capecitabine clearly improves survival rates.

Acknowledgments

None.

Conflicts of interest

Authors declare that there is no conflict of interest.

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