

HIV patient with pseudomyxoma peritonei of extra-appendiceal origin is disease-free and alive 9 years after complete cytoreductive surgery

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Abstract. – **OBJECTIVE:** Pseudomyxoma peritonei (PMP) is a clinical syndrome that is mainly characterized by the presence of mucinous ascites that results from the rupture of a mucin-producing neoplasm. No reports exist so far regarding the management of this syndrome in HIV patients.

CASE REPORT: A 54-year old male patient with a diagnosed atypical colitis developed additionally over time a complicated diverticulitis which was initially treated conservatively with antibiotics. Due to septic complication, the patient received a Hartmann resection. Six months after the first surgery a Hartmann reversal was tried; it, however, revealed peritoneal implants and a significant stenosis of the colon stump. Resection of these lesions confirmed histopathologically the presence of a low-grade pseudomyxoma peritonei. Five months later, a second try for a Hartmann reversal was performed. In the view of the slightly enlarged paracolic lymph nodes a low anterior resection was performed with a, primary descendentostomy. Histopathological examination revealed no more signs of the tumor, which confirmed the completeness of the cytoreductive surgery by the first try for a Hartmann reversal.

CONCLUSIONS: Completeness of cytoreductive surgery and low grade histology seem to be the most important factors for the prognosis in pseudomyxoma peritonei in immunocompromised patients as suggested by the long overall and progression free survival of the present patient.

Key Words

Pseudomyxoma Peritonei, Diverticulitis, HIV, Hepatitis C.

Introduction

Pseudomyxoma peritonei (PMP) is a clinical syndrome that is mainly characterized by the presence of mucinous ascites that results from the rup-

ture of a mucin-producing neoplasm. Typical origin of PMP is the appendix¹ but mucinous tumors from other organs such as the ovary, colon, urachus and the pancreas have also been described².

Case Presentation

A 54-year old male patient was diagnosed with an atypical colitis in 2001; after a colonoscopy was performed, it revealed inflammatory alterations of the sigma and the rectum compatible with a healing process after possible traumatic injury or infection. The patient had already been diagnosed with a chronic HIV infection (Stage A2/CDC 1993), a hepatitis C infection and was also presenting a history of drug abuse. A planned colonoscopy in 06/2006 by persistent bowel movement disorders and mild pain of the lower abdomen revealed an acute diverticulitis with sigmoid stenosis. Colonoscopy was performed afterward until the ileocecal valve, however revealing no other abnormalities in the rest of the colon. The patient had a history of emergent appendectomy 13 years ago due to acute appendicitis with a negative for tumor final histological examination. The colonoscopy revealed no pathological findings regarding the proximal resection margins of the Appendix and finally the patient was treated for the acute diverticulitis as outpatient with oral antibiotics.

Despite the initial management, the patient presented again after 20 days to our emergency department with acute onset of pain on the lower left abdomen. A CT scan was performed that revealed a complicated diverticulitis of the sigma classified as Hinchey I-II with pathologic dilatation of the paracolic lymph nodes. Initially, the patient was treated with intravenous antibiotics as inpatient but he became septic and

therefore a Hartmann resection with the creation of an end colostomy was emergently performed. Clinical examination of the abdomen intraoperatively confirmed dilatation of the paracolic lymph nodes and the microbiological examination of the intrapelvic abscess confirmed the presence of *Proteus Mirabilis*. One month after the operation the patient was discharged. The histopathological examination confirmed inflammatory alterations of the colon in the background of a complicated diverticulitis without any signs of malignancy. In addition, the resected lymph nodes along with the colon specimen were negative for malignancy.

Six months after the primary operation, the patient was electively admitted to our hospital for a planned Hartmann reversal. Preoperative work-out with a contrast enema revealed a significant stenosis of the colon stump. Intraoperatively, the stenosis was hard in consistency and interestingly the inspection of the abdomen revealed the presence of peritoneal implants in the peritoneum of the colon stump, the small intestine and the omentum with a diameter maximum to 1.2 cm. The palpated stenosis was resected along with the peritoneal implants with adequate margins and in this situation no Hartmann reversal was performed. The histopathological examination confirmed the presence of a low-grade pseudomyxoma peritonei. Radiologic follow up with CT revealed no signs of a tumor but slightly dilated paracolic lymph nodes. Five months after surgery a second try for a Hartmann reversal was performed. However, clinical examination of the abdomen did not demonstrate suspected pathological findings but in the view of the slightly dilated lymph nodes a complete low anterior resection was performed with a primary descenderectostomy along with a loop ileostomy. The histopathological examination did not reveal any more signs of pseudomyxoma peritonei that subsequently confirmed that a complete cytoreductive surgery was performed in the previous operation.

Follow up of the patient suggested no occurrence of disease recurrence. The patient has a stable chronic HIV infection and remains disease free as far as the pseudomyxoma peritonei is concerned.

Discussion

PMP represents a rare clinical syndrome with an incidence of 1-2 patients per million per year. The vast majority of these tumors have an appendiceal origin³, an observation which makes PMP

of extra-appendiceal origin extremely rare. In a recent study from Baratti et al⁴, the criterion to confirm PMP of extra-appendiceal origin was the presented histologic confirmation of the non-neoplastic appendix. For patients with a history of appendectomy, an intraoperative sampling from the prior appendectomy sites is performed with the histopathological findings being considered as equivalent to the primary appendiceal lesions. Regarding the HIV patient, the characterization 'extra-appendiceal' was based on the history of non-neoplastic appendix occurrence after appendectomy, the absence of pathological findings in colonoscopy prior to Hartmann resection and the absence of macroscopic peritoneal implants in the prior appendectomy sites.

A unique feature of PMP is the redistribution phenomenon. The normal flow of peritoneal fluid follows the mucus and the neoplastic cells of the tumor, therefore being redistributed within the peritoneal cavity to sites of fluid absorption through the lymphatic flow. As a consequence, tumor cells accumulate more often in positions such as the paracolic gutters, omentum, pelvis and liver capsule².

Cytoreductive surgery for patients who are considered preoperatively resectable remains the standard of therapy. However, cytoreductive surgery requires the resection of the involved parietal peritoneum along with the involved viscera. Such an approach might entail the performance of greater omentectomy, splenectomy, stripping of the left and right diaphragm, cholecystectomy, lesser omentectomy, appendectomy or right/total colectomy, partial or total gastrectomy, pelvic peritonectomy with resection of the rectosigmoid, hysterectomy and bilateral salpingo-oophorectomy¹. Of note, the completeness of cytoreduction (CCR) is an important prognostic factor. In a study of Chua et al [5] regarding patients with PMP of appendiceal origin, the five-year overall survival was 85% when no tumor was macroscopically present after resection (CCR Score 0) and 80% when no tumor implant greater than 2.5 mm in diameter existed (CCR1). However, it was significantly lower that was up to 24% when a gross tumor implant remained. If a CCR0 is not doable, the aim is to leave only signs of microscopic disease and therefore heated intraperitoneal chemotherapy (HIPEC) might be used in patients with CCR0/1¹. In addition, the primary origin of the tumor (appendiceal versus extra-appendiceal) does not seem to correlate with poorer survival according to the study of Baratti et al³ (n=225 patients) that demonstrated a ten-year overall survival of 63.4% regarding the occurrence of appendiceal PMP and

62% concerning extra-appendiceal PMP. In the same study independent prognostic factors associated with poor survival were: high grade peritoneal histology, prior systemic chemotherapy, more than four visceral resections and incomplete cytoreduction. The patient of the current report would have been expected to be at a higher risk of recurrence due to his immunocompromised situation related to the chronic HIV infection. The CCRO score and the low-grade histology seem to have contributed to the good course of the patient's disease.

From a molecular aspect, PMP harbors a variable biologic behavior with up to 30% of patients dying due to disease progression. Recently, Pietrantonio et al⁴ demonstrated in their study (n=40) that the presence of KRAS mutations was independently associated with progression free survival. KRAS mutations were present in 72% of the tumors. However, the limitation of this study is mostly associated to the small sample size of the patients and the retrospective nature of the study.

Conflict of Interest

The Authors declare that they have no conflict of interests.

Ethics approval

Approval was obtained from the Research Ethics Committee of the National and Kapodistrian University, Athens Medical School, Laiko General Hospital.

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