Primary Peritoneal Carcinoma in Complete Remission: a Case Report

JORGE RAMOS1*, KRISHNA S. GUNTURU1*, SARAVANAN KRISHNAMOORTHY2, BARTON KENNEY3 and M. WASIF SAIF4

Departments of 1Medicine, 2Radiology and 3Pathology, Yale University, School of Medicine, New Haven, CT, U.S.A.; 4Departments of Hematology/Oncology, Pancreas Center, Columbia University College of Physicians and Surgeons, New York, NY, U.S.A.

Abstract. Primary peritoneal carcinoma (PPCa) is a relatively uncommonly diagnosed tumor. It has a similar presentation to ovarian cancer. PPCa has a poor prognosis with survival ranging from 12-18 months. PPCa spreads mainly transperitoneally, but lymphatic and hematological metastases have also been reported. It is a diagnosis of exclusion made after pathological report. Here, a case of a 71-year-old female who presented with early satiety, fatigue, weight loss and left cervical lymphadenopathy and was diagnosed with metastatic PPCa, is reported. The patient was treated with chemotherapy and achieved a complete remission. The management of this rare tumor is discussed herein.

Primary peritoneal carcinoma (PPCa) is a rare tumor and was historically classified as “carcinoma of unknown primary”. This entity was mostly reported in women arising from extra ovarian peritoneum with Mullerian potential (1, 2). The mean age at the time of clinical presentation is 61 years of age. Patients typically have advanced disease at the time of presentation. Symptoms are similar to patients with ovarian carcinoma with peritoneal metastasis, including abdominal pain and distension with ascites being the most common complaints. Diagnostic criteria to distinguish PPCa from primary ovarian cancer include normal sized ovaries, extra-ovarian site involvement greater than surface involvement of the ovary, the ovarian component, at most, must be less than 5x5 mm within the ovary and otherwise confined to the surface of the ovary, and finally, histological characteristics must be predominantly of the serous type (3). It is also important to distinguish PPCa from primary peritoneal mesothelioma via asbestos exposure history and immunohistochemistry. First line chemotherapy is typically a platinum-based combination with a taxane (4). The median survival in patients with PPCa is typically 12-18 months.

Case Report

A 71-year-old Hispanic female with a past medical history significant for diabetes-mellitus presented to her primary care physician with a two month history of fatigue, weight loss, early satiety, and new onset of left neck swelling. A CT scan of the neck and chest showed left-sided cervical and mediastinal lymphadenopathy. In addition, bulky lymphadenopathy of the upper abdomen surrounding the aorta and inferior vena cava was noted. A MRI of the abdomen showed a mass on the splenic hilum obstructing the splenic vein and bilateral adrenal masses. Subsequently, a PET scan showed multiple masses, including a 10x5.6x7.3 cm mass between the spleen and stomach, a 7.2x5.2 cm mass near the ascending colon and a 5.8x4.5 cm right adrenal mass. The patient was also noted to have a lesion on the pancreatic head and widespread lymphadenopathy. Follow-up MRI and PET scan at our institution again showed extensive lymphadenopathy, including the left supraclavicular chain (Figure 1), chest, abdomen and pelvis (Figure 2). Her cancer antigen (CA) 125 level at the time of presentation was 24,800 U/ml (reference value 0-35 U/ml). The patient also had an elevated CA 15-3 of 65.7 U/ml (reference value 0-31.3 U/ml) and normal CA 19-9 value of 3.3 U/ml (reference value 0-37 U/ml) and carcino embryonic antigen (CEA) at normal levels of 1.2 ng/ml (reference value 0-3 ng/ml). The patient had had a normal mammogram and a colonoscopy two years prior without any evidence of disease.
The patient had had a total abdominal hysterectomy and bilateral salpingo-oophorectomy 13 years prior to presentation. A fine needle aspiration of one of the abdominal masses was positive for adenocarcinoma. Immunohistochemistry was positive for cytokeratin (CK)19, CK7, BerEP4, Wilm’s tumor suppression gene (WT-1) and negative for calretinin, cluster of differentiation (CD-20), CDK2, thyroid transcription factor (TTF-1), and estrogen/progesterone receptor (ER/PR) (Figure 3-4). These findings, in addition to the clinical features, laboratory data and radiographic imaging, were consistent with primary peritoneal carcinoma.

The patient was started on carboplatin, docetaxel and erlotinib and received 11 cycles of this regimen. After 3 months since start of this regimen a follow-up CT scan showed a 41% reduction in disease burden per RECIST criteria. This regimen was discontinued because of docetaxel induced fluid retention syndrome and switched to bevacizumab because of radiographic progression of disease in the lungs. The patient received 5 cycles of bevacizumab before discontinuing this regimen secondary to symptomatic uncontrolled hypertension.

Gemcitabine was given briefly, but it was discontinued secondary to fluid retention. The patient was transitioned to single agent vinorelbine every 2 weeks. CT/PET scan performed after 19 cycles of vinorelbine and 27 months after the time of diagnosis, demonstrated that the patient had a complete metabolic response without evidence of mediastinal, hilar or abdominal lymphadenopathy (Figure 5). At this point the patient was taken off vinorelbine and continued to be disease free 3 years after her initial diagnosis and 8 months after her last chemotherapy with normal tumor marker values.

Discussion

PPCa has a similar presentation, histology and response to chemotherapy as that of ovarian carcinoma (5). Eltabbakh et al. reported that women with extra ovarian primary peritoneal carcinoma (EOPPC) were older than women with ovarian cancer and had later menarche (6).

Immunohistochemical stain for PPCa is positive for CK-7, WT-1, and CA-125 (7). Several reports have suggested that
Figure 3. Biopsy material from the abdominal mass. A. Rare clusters of malignant cells admixed with blood (H&E, 20X). B. Malignant cells with high grade nuclear features and mitotic activity (H&E, ×40).

serum CA-125 is a useful tumor marker for monitoring the course of PPCa (8, 9). In the present patient, the initial CA 125 was elevated at 24,800 U/mL and at the end of the chemotherapy CA-125 was 19.7 U/mL.

Platinum-based combination chemotherapy, similar to the regimens used for ovarian cancer is widely recommended in PPCa(10). A combination of platinum and taxane based regimen either including paclitaxel or docetaxel has been the first-line treatment standard (11). The current patient was initially treated with a docetaxel, carboplatin and erlotinib combination with which the patient had a good response (12). Gemcitabine and vinorelbine have also shown activity in platinum-resistant ovarian cancer (13, 14). Targeted therapy with biological agents such as bevacizumab have shown 15% response in platinum resistant ovarian cancer or peritoneal serous cancer(15).

References


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