OMCR

Oxford Medical Case Reports, 2018;10, 318–322

doi: 10.1093/omcr/omy057
Case Report

CASE REPORT

Pseudomyxoma pleurii and peritonei secondary to sigmoid colon adenocarcinoma: a rare clinico-pathologico-radiological presentation

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Abstract

Pseudomyxoma peritonei (PMP) is a rare condition resulting from mucin-producing tumors that have disseminated into intraperitoneal implants and mucinous ascites. The extra-abdominal spread of PMP is exceptionally rare, with few reported cases in the medical literature. Pseudomyxoma pleurii is an infrequently encountered clinical syndrome characterized by transdiaphragmatic pleural extension and spread of PMP. The disease is highly fatal. We hereby report a case of 58 years old woman who presented with an abdominal distension and shortness of breath of 2 months duration. Histopathology confirmed the diagnosis of large mucin-producing rectosigmoid adenomatous polypoid lesion with malignant transformation and PMP that had spread to the right pleural space. PMP from colon tumor is uncommon and its transdiaphragmatic pleural extension is very unusual complicated by management challenge and high mortality rate.

INTRODUCTION

First case of Pseudomyxoma peritonei (PMP) was described by Rokitansky in 1842 [1]. In 1884, Werth found its associated with the ovarian cancer. In 1901, Franckel *et al.* found its association with appendiceal cystic tumors [2]. PMP is a rare clinical condition resulting from accumulation of copious mucinous material from mucin-producing tumors that disseminate into abdomen, peritoneal and pelvic cavities as ascites and implants. It is characterized by chronic, relapsing, disease [3]. The presence of neoplastic and inflammatory cells in the mucinous secretions distinguishes this condition from simple ascites. Patients who have PMP and have disseminated peritoneal adenomucinosis have more favorable prognosis than patients who have peritoneal mucinous carcinomatosis [4, 5]. This tumor is mainly superficially invasive but still is a fatal disease. Because of this unusual pattern of dissemination, it draws significant attention and interest from the surgical oncologists as it pertains to pursue aggressive loco-regional therapy in order to improve survival and possible cure. Pseudomyxoma pleurii is a condition where the pleural cavity is filled with mucinous material. It is caused by the transdiaphragmatic spread of PMP from abdominal origin [6]. Massive pleural disease is a life threatening condition that impairs cardiopulmonary functions. PMP mostly arises from appendiceal adenomas or mucinous cyst, adenocarcinoma or ruptured primary ovarian cancer, but can also arise from an indeterminate site [7, 8].

CASE PRESENTATION

A 58-year-old lady was admitted to our hospital with the complaints of shortness of breath, abdominal distension and loss of

Received: February 8, 2018. **Revised:** May 21, 2018. **Accepted:** June 29, 2018

appetite for 2-month duration. She was diagnosed with liver cirrhosis in another hospital 2 weeks prior to this presentation. She was referred to us for the management of large ascites, which was thought to be from decompensated chronic liver disease. She was treated for toxic goiter some years back with radioactive iodine therapy and was lost to follow-up.

She had a history of drinking a bottle of wine and spirits daily for 30 years. She denied any smoking history. On physical examination she was alert and oriented, afebrile, had pallor but there was no evidence of jaundice or edema. Systemic examination revealed tachypnea (32b/min), tachycardia 128/b/min, desaturating on room air (SpO_2 88%). Chest percussion revealed stony dullness on the right side. Cardiovascular examination was normal apart from tachycardia. Abdomen was grossly distended with mild tenderness and shifting dullness but no organomegaly or masses. Rectal examination was normal.

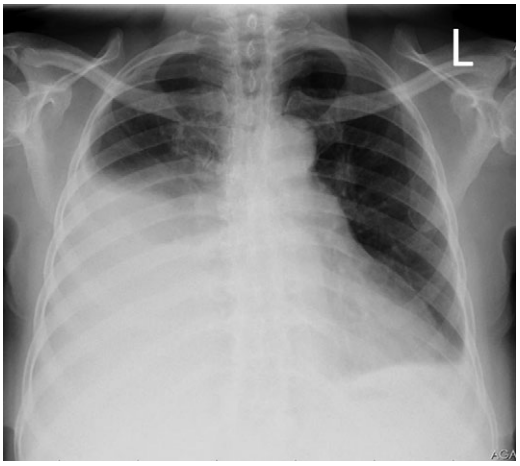

Figure 1: X-ray chest AP view showing large right pleural effusion with loss of right costo-phrenic and cardio-phrenic angle.

Complete blood count showed iron deficient anemia with low hemoglobin of 8.7 g/dl and mild neutrophilia and monocytosis. Liver function tests were normal. Renal function tests were normal. Viral hepatitis B and C, HIV I/II tests were all negative. Erythrocyte sedimentation rate (ESR) was high (70 mm/h). C-reactive protein (CRP) was slightly high (34.4 mg/l). Serum alpha fetoprotein (AFP) was normal (4.38 ng/ml) and serum CA-125, CA 19-9, and carcinoembryonic antigen (CEA) and lactate dehydrogenase (LDH) were all elevated (166.70 U/ml, 354.40 U/ml, 300.20 ng/ml, 329.30 U/l, respectively). Serum thyroid stimulating hormone (TSH) was high (9.77 uIU/ml), T3 and T4 were critically low (0.686 and 5.07 pmol/l, respectively). Chest X-ray revealed large right pleural effusion with possible underlying collapse/consolidation (Fig. 1). Computed tomography scan of the abdomen revealed large ascites, scalloping of the liver margins with multiple subcapular lesions (largest measuring $12.8 \times 6.4\ cm^2$), peritoneal deposits and large mixed echogenicity solid lesion seen all over abdomen and pelvis, involving omentum and mesentery extending into poach of Douglas. Gall bladder, urinary bladder, uterus and ovaries were not seen separately from this lesion (Fig. 2). This was interpreted to be large intraperitoneal tumor deposit. Visualized bowel loops appeared normal. Common bile duct (CBD), spleen and both kidneys were normal. Intraperitoneal, omental and mesentery fluid loculations were noted. The findings were suggestive of PMP. Ascites fluid tap revealed neutrophils 2600/µl, was exudative but negative for adenosine deaminase (ADA), Zeil Nelson stain, Indian ink, gram stain and culture. Pleural fluid was exudative and had cell count 1000/µl and the rest of tests were negative in the ascites fluid sampling. Cytology smear showed predominantly 80% lymphocytes and 20% polymorphs with proteinaceous

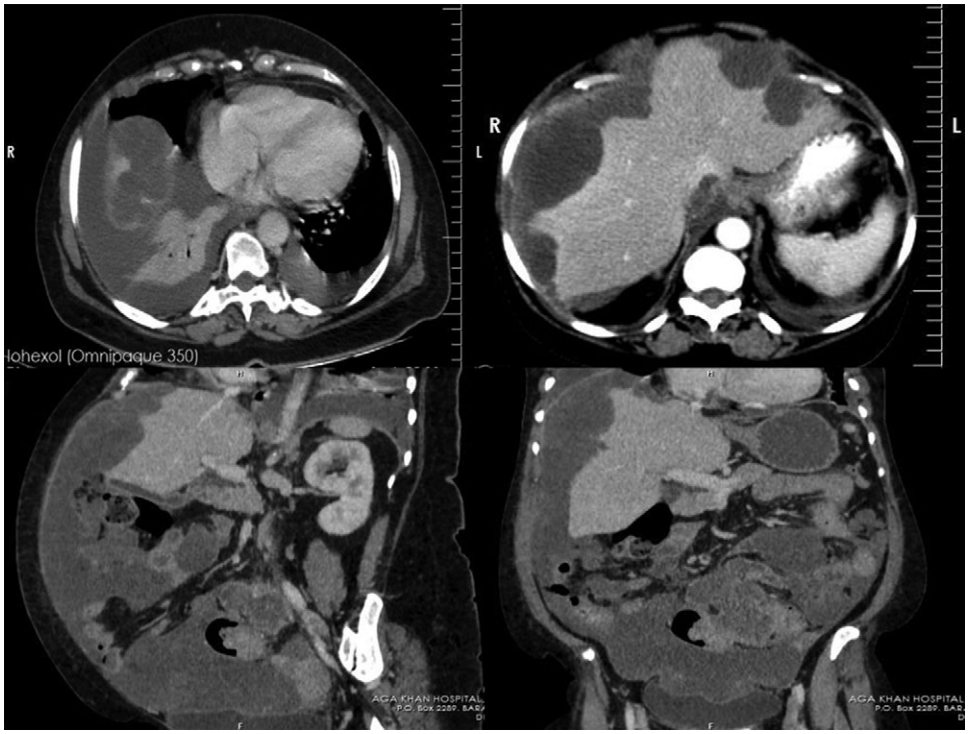

Figure 2: CT scan of the abdomen revealed multiple subcapsular lesion, peritoneal deposit and large ascites (shown via arrow).

background showing few RBCs. Colonoscopy revealed a very large polypoid but soft lesion with features of a villous adenoma in the rectosigmoid (20–30 cm from anal verge) with possible malignant transformation (Fig. 3). Colonoscopic biopsies revealed villous adenoma with moderate dysplasia and focal invasion (Fig. 4A and B). In view of multiple tumor metastasis and PMP, features were consistent with malignant transformation. Biopsies from omental masses revealed extracellular mucin pools with floating clusters.

Computerized tomography-positronic emissions tomography (CT-PET scan) of the head, chest and abdomen was done which showed uptake in peritoneal cavity, subdiaphragmatic, inferior surface of the liver, omentum, pelvis with mucinous and in the sigmoid colon. These findings confirmed the diagnosis of PMP and sigmoid primary neoplastic pathology (Fig. 5). Also seen was uptake along bowel loops and right pleura.

DISCUSSION

PMP is uncommon clinical condition caused by dissemination in the abdominal cavity of mucinous adenocarcinoma cells from mucin-producing tumors that results in mucinous or gelatinous ascites [9]. It rarely spreads via the lymphatic system or through the blood stream. PMP is characterized by having mucin and scattered cancer cells in the abdominal cavity (also referred as gelatinous collections or jelly belly) with mucinous implants on the omental and the peritoneal surfaces [10]. In the absence of treatment, mucin will ultimately build up and will compress the vital structures as the stomach, kidneys, colon, liver, pancreas and spleen. This is also known as local and regional distribution or loco-regional progression [11], which as a result of raised intra-abdominal pressure and abdominal compartment syndrome can results in impairment of intestinal function and lead to malnutrition, fistula formation and infections [12]. This leads to significantly increased mortality and morbidity. PMP in the old literature was said to occur from a variety of primary tumors but mainly from appendicular tumors [8, 11, 13]. Recently, PMP from colorectal tumors have been reported, which is very rare [14]. Pseudomyxoma pleurii is an even uncommon pathological form which is characterized by the presence of malignant mucinous implants due to transdiaphragmatic spread of the PMP [6]. In the index patient the primary tumor was an adenocarcinoma in the sigmoid colon causing PMP with extension to the right pleura causing pseudomyxoma pleurii, making it a very rare presentation

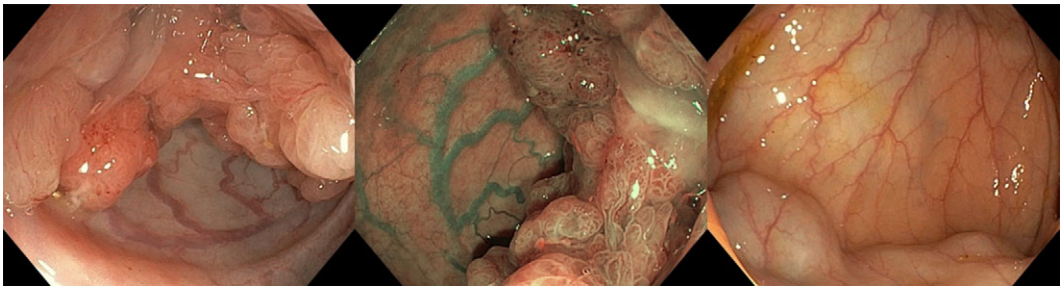

Figure 3: Colonoscopy revealed large polypoid but soft lesions with features of villous adenoma in the rectosigmoid colon (shown via arrow).

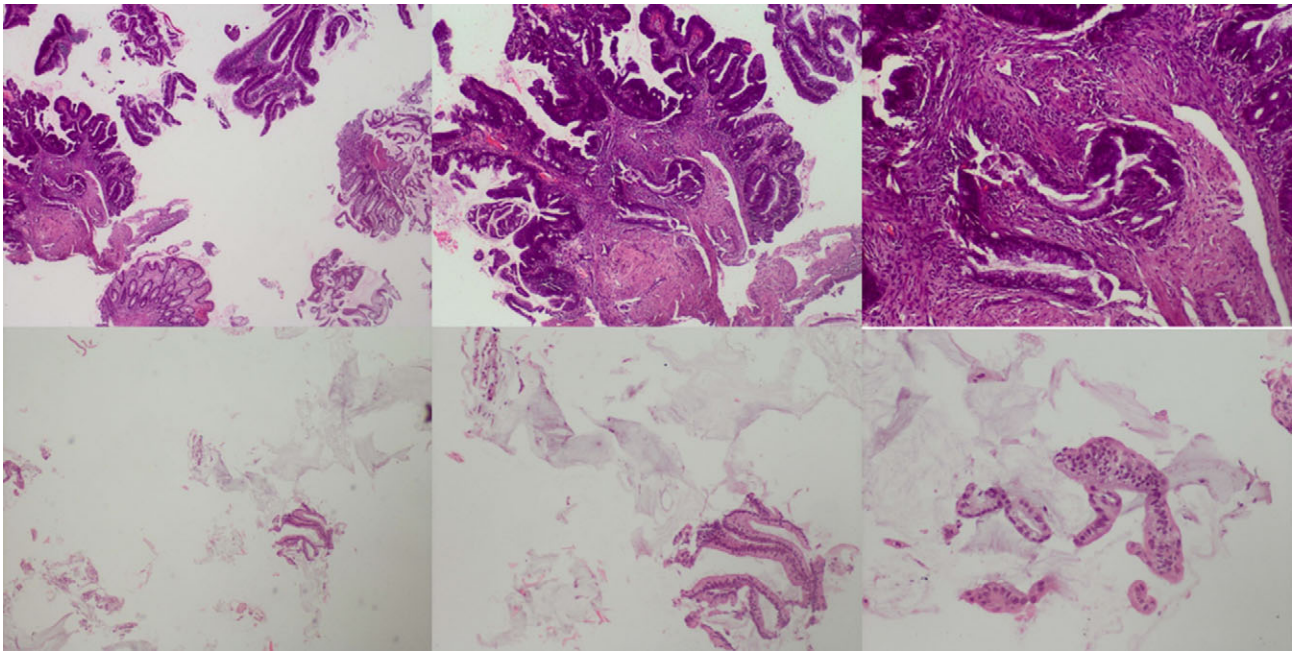

Figure 4: (**A**) Low power and (**B**) high power images of colonic biopsy showing villous adenoma with moderate dysplasia and focal invasion.

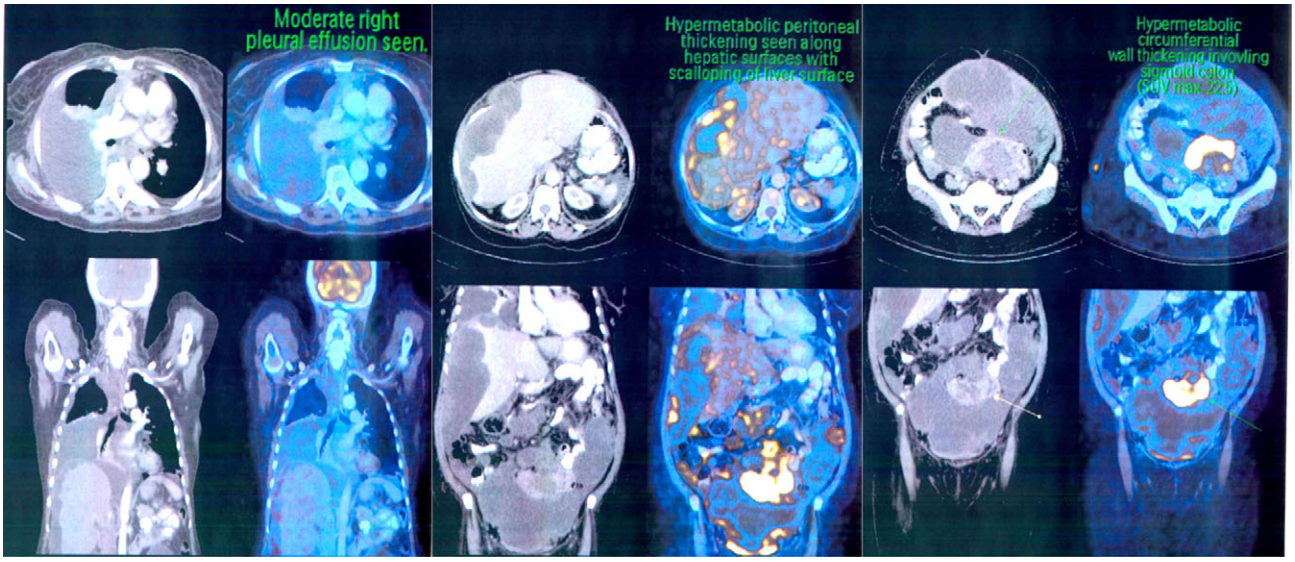

Figure 5: PET-CT scan showing uptake in peritoneal, sub-diaphragmatic, inferior surface of liver, omental area and pelvic area.

among only few cases reported in the literature. This condition is often misdiagnosed in the early part of the disease process as decompensated chronic liver disease with massive ascites due to patient having similar non-specific symptoms in the early stages of the disease. This patient had presented very late with extensive disease dissemination. As it was also observed in this patient, routine laboratory studies are seldom helpful in making the diagnosis. Radiology imaging with computed tomography helps in suggesting the diagnosis but histopathology is the gold standard and provides the confirmatory diagnosis [3, 4]. Patients with PMP with adenomucinosis have much favorable prognosis when compared to patients with peritoneal mucinous carcinomatosis [2, 5]. CT-PET scan is useful to detect extent of the disease and potential for treatment [15]. Even with the better understanding and recent advances in the management of this disease, PMP creates a diagnostic challenge in the early stage of the disease. PMP patients who have elevated pre-operative tumor markers such as CEA cancer antigen 125 (CA 125) and carbohydrate antigen 19-9 (CA 19-9) are at increased risk of developing recurrent disease despite aggressive treatment [3]. PMP patients who have normal tumor marker levels have a better overall survival overall [16, 17]. Our patient has presented late in the course of the disease, making it challenging. The main therapeutic goal is prevention of local and regional recurrence. Surgery and complete cytoreduction treatment combination followed by intraperitoneal chemotherapy, rather than intravenous chemotherapy have been suggested as standard treatment option. Cytoreductive (debulking) surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) has been also suggested as an effective treatment options for treatment of patients who have peritoneal surface metastatic cancers such as stage IV colon cancer and PMP. HIPEC therapy produces hyperthermia which improves the penetration of tumor by the chemotherapeutic agents. Systemic effects are much less due to the peritoneum–blood barrier [6]. This technique is a two-step process: first phase entails surgically removing any visible tumor or cancer, and in phase 2 delivering heated chemotherapy drugs into the affected area [18]. Systemic chemotherapy is primarily recommended for patients with extensive peritoneal disease and high-grade cystadenocarcinoma [19].

Prognosis in patients with PMP is closely related to the tumor burden, pre-operative tumor volume success of tumor resection by cytoreductive surgery and pathological classification [20]. Our patient was started on combination systemic chemotherapy with FOLFOX (folinic acid (leucovorin), 5-fluorouracil (5FU) and oxaliplatin) for 4 months before she underwent HIPEC surgery when CEA was <50 ng/ml.

CONCLUSION

PMP is a rare and frequently misdiagnosed condition. Pseudomyxoma pleurii (involvement of the pleural cavity) via transdiaphragmatic spread of PMP is very rare with an unfavorable prognosis. Local and or systemic chemotherapy with extensive cytoreduction procedures and treatment should be considered. In this patient systemic chemotherapy was given as palliative therapy. Clinical suspicion should be high for early diagnosis of this rare entity.

ACKNOWLEDGEMENTS

None.

CONFLICT OF INTEREST STATEMENT

None of the authors have any conflict to disclose.

FUNDING

None.

ETHICAL APPROVAL

The case report did not require the ethical board approval.

CONSENT

Consent has been obtained from the patient

GUARANTOR

Casmir Wambura, MD and Salim Surani, MD.

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