Pseudomyxoma Retroperitonei: 
A Rare Cause of Retroperitoneal Cystic Mass

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ABSTRACT
Pseudomyxoma retroperitonei is a rare condition, characterized by accumulation of mucinous material in the retroperitoneal space, originating predominantly from the appendiceal mucinous neoplasms. A 72-year-old male patient presented with a history of progressive right side abdominal pain for 5 months with a palpable abdominal mass. Ultrasound, computerized tomography, and magnetic resonance imaging showed large right abdominal multiloculated cystic lesion with heterogeneous echoic contents. Colonoscopy revealed normal mucosa with extramural pressure on the right colon and cecum. He underwent complete excision of the mass along with right hemicolectomy. The cystic mass was containing gelatinous material. Histopathology revealed low grade mucinous neoplasm. Pseudomyxoma retroperitonei should be considered in the differential diagnosis of patient presenting with progressive right side abdominal pain and retroperitoneal cystic mass.

Key Words: Pseudomyxoma retroperitonei. Retroperitoneal cystic mass. Appendiceal mucocele.

INTRODUCTION
Pseudomyxoma peritonei is a rare clinical condition that is characterized by the presence of mucinous ascites in which gelatinous material spreads and accumulates throughout the peritoneal cavity and pelvis.¹,² Pseudomyxoma retroperitonei is extremely rare condition which is also called pseudomyxoma extraperitonei.¹,² We present a rare case of pseudomyxoma retroperitonei, extending from the posterior area to the hepatic flexure of the colon and anterior to the right kidney to the right iliac fossa, in which the treatment was complete excision of the mass along with right hemicolectomy.

Our objective in presenting this case is to increase the awareness among clinicians about this rare cause of retroperitoneal cystic mass for adequate management. The case is discussed in the context of other reported cases in the literature.

CASE REPORT
A 72-year-old male patient, presented to the emergency department (ED) with a history of progressive right side abdominal pain for 5 months. He had no history of change in bowel habit, urinary symptoms, and loss of weight or appetite. On examination, he was conscious and oriented, with blood pressure of 130/70 mmHg, pulse rate of 78 beats/minute, temperature of 36.8°C, and non-tender huge mass in the right side of the abdomen, extending from the right upper quadrant to the right iliac fossa. Laboratory values were as follows: white blood cell count 5.4 x 10⁹/L (normal value [NV]: 4 - 10.5 x 10⁹/L); hemoglobin 11.6 g/dl (NV: 14 - 18 g/dl); urea 4.4 mmol/L (NV: 3.6 - 7.1 mmol/L); creatinine 108 mmol/L (NV: < 133 mmol/L); and normal coagulation profile. He then underwent abdominal ultrasound (US) which showed large right abdominal multiloculated cystic lesion with heterogeneous echoic contents, extending from right upper quadrant to the right iliac fossa with normal right kidney (Figure 1). Abdominal computed tomography (CT) revealed right retroperitoneal multilocular, septated, non-enhancing, large cystic mass, measuring 19 x 10 x 11 cm with 25 HU radiodensity, and linear and clumpy calcifications, extending anterior to the right kidney, down into the right iliac fossa. Magnetic resonance imaging (MRI) showed large, well defined and lobulated, T1 low, T2 high signal intensity cystic mass with multiple enhancing septations in the right retroperitoneal space, displacing the right colon and small bowel loops anteriorly and medially. The imaging criteria and location were those of cystic retroperitoneal neoplasm, probably sarcoma. Complete colonoscopy revealed normal large bowel mucosa with evidence of extramural pressure on the right colon and cecum. He underwent exploratory laparotomy through midline incision with medial mobilization of the right colon which revealed large retroperitoneal cystic mass encroaching the confluence of the taenia coli in the cecum (the anatomical site of the base of the appendix), but the appendix was not found. This cystic mass is tightly adherent to the cecum and right colon. During mobilization of the cystic mass, there was small perforation, which leaked gelatinous material, and it was repaired immediately. Complete excision of the mass...
along with the right hemicolectomy was performed with uneventful postoperative recovery. The cystic mass was containing gelatinous material like pseudomyxoma peritonei. Thus, we diagnosed the mass as pseudo-myxoma retroperitonei. Histopathology revealed low grade mucinous neoplasm with mucinous contents. Immunohistochemistry was positive for CK20, MUC-2 and MUC-1 but negative for CA125 and CK7. Histopathology of the colon and cecum was normal with no malignancy. At present, the patient is under the care of oncology for chemotherapy.

DISCUSSION

Pseudomyxoma peritonei has been reported frequently in the literature since long time. However, retroperitoneally confined pseudomyxoma (pseudomyxoma retroperitonei) as in this case, is extremely rare with only 36 cases reported in the international literature including this case. Pseudomyxoma peritonei and retroperitonei are believed to originate predominantly from appendiceal or ovarian mucinous neoplasm that release mucinous neoplastics cells into the peritoneal cavity or retroperitoneal space, respectively.1,2 The majority of reported cases of pseudomyxoma retroperitonei originate predominantly from the appendix due to rupture of retrocaecal appendiceal mucocoele, and rarely from the ovary, colon and other sites.3,4 In this case, the retroperitoneal mass was connected to the base of the appendix. The clinical presentation of pseudomyxoma retroperitonei is usually nonspecific, with a history of slowly progressive abdominal or lumbar pain and palpable mass in the abdominal examination.3 Abscess formation with high fever, spontaneous skin fistula, discharges and weight loss have also been reported.4,5 Our patient presented with slowly progressive abdominal pain over 5 months, associated with palpable abdominal mass which was obvious even by the inspection of the abdomen. Laboratory values may reveal elevated serum level of tumor markers including carcinoembryonic antigen (CEA) and CA 19 - 9,2 which has not been assessed in this case. Imaging tools such as US, CT scan aid the diagnosis. US would reveal the mucinous material as retroperitoneal fluid. Abdominal CT scan should be obtained with oral, rectal and intravenous contrast. The imaging findings of pseudomyxoma retroperitonei in CT scan are similar to those of pseudomyxoma peritonei.6 Pseudomyxoma retroperitonei appears as masses, usually multicystic, with thick walls and septae, which displace and distort the adjacent structures. It may also reveal punctate or curvilinear mural calcifications.6 In this patient, the imaging studies revealed a retroperitoneal, multicystic, septated mass with linear and clumpy calcifications and displacing the right colon and small bowel loops anteriorly and medially. Histopathologically, the pseudomyxoma has been classified into grade I (benign peritoneal adenomucinosis), grade II (intermediate type), and grade III (malignant mucinous carcinomatosis).1,2 Immunohistochemistry of these cells are generally positive for MUC-2 and CK20 but negative for MUC-1,
CA125 and CK7. Histopathology in this case revealed low grade mucinous neoplasm with mucinous contents, and the immunohistochemistry was positive for CK20, MUC-2 and MUC-1 but negative for CA125 and CK7. The treatment of pseudomyxoma retroperitonei is mainly surgical with debulking and resection of the involved viscera, followed sometimes by adjuvant systemic chemotherapy. However, the best prognosis can be expected with intact removal of the retroperitoneal mass. Although hyperthermic intraperitoneal chemotherapy (HIPEC) has been used extensively in the pseudomyxoma peritonei, it has not been reported in pseudomyxoma retroperitonei; however, it should be considered as an optional treatment modality due to the high recurrence rate. This patient underwent complete resection of the retroperitoneal mass along with right hemicolectomy. Follow-up is critical due to the high risk of recurrence or progression of the disease, and it consists of physical examination, tumor markers, and CT scan.

In conclusion, pseudomyxoma retroperitonei is a rare clinical condition, which can form a retroperitoneal cystic mass. It should be considered in the differential diagnosis in a patient presenting with progressive right side abdominal pain and retroperitoneal cystic mass. As a lethal entity, complete intact resection of pseudomyxoma retroperitonei along with the adherent viscera may give the highest chance of cure.

REFERENCES