

## Pseudomyxoma peritonei presenting with inguinal hernia

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### Rezumat

#### *Hernie inghinală - manifestare a pseudomixomului peritoneal*

Pseudomixomul peritoneal (PMP) este o nosologie rară, caracterizată prin acumularea intraperitoneală de mucus produs de celule neoplazice de origine apendiculară. Semnele clinice fiind diverse, diagnosticul preoperator deseori este dificil. Descriem cazul clinic al unui pacient de 67 ani, internat la peste o lună de la herniotomie prezentând semne clinice de PMP. A fost efectuată citoreducerea chirurgicală, peritoneumectomie, apendicectomie, omentectomie și chimioterapie intraperitoneală perioperatorie. Pacientul a fost asimptomatic timp de 15 luni, când a decedat aparent în urma unui atac cardiac. În concluzie, în caz de prezență a lichidului gelatinos în timpul herniotomiei – pacientul trebuie investigat histologic și tomografic pentru a confirma PMP.

**Cuvinte cheie:** pseudomixom peritoneal, hernie inghinală

### Abstract

Pseudomyxoma peritonei (PMP) is rare being characterized by intraperitoneal accumulation of mucinous ascites produced by neoplastic cells, mostly originating from a perforated

appendiceal adenoma. The clinical signs of the disease are variable, and preoperative diagnosis is often difficult. We describe the clinical case of a 67-year-old patient referred to our unit one month after a left inguinal hernia repair, presenting clinical signs compliant with PMP. Surgical cytoreduction, peritonectomy, appendectomy, and greater omentectomy with perioperative intraperitoneal chemotherapy were performed. The patient was disease free for a 15 month period when he died apparently due to a cardiac event. We advocate that in all cases of gelatinous fluid in a hernia sac PMP must be suspected, thus histological investigation is mandatory as well as abdominal computed tomography (CT) in order to confirm the diagnosis.

**Key words:** pseudomyxoma peritonei, hernia

### Introduction

Pseudomyxoma peritonei is a rare disease. It refers to a progressive disease process within the peritoneum which originates from the appendix or ovaries and is characterized by the production of copious amount of mucinous fluid resulting in a "jelly belly" (1,2). The symptoms of the disease vary, preoperative diagnosis is often difficult, and, in many cases, the diagnosis is established during surgery (3). There are several reports regarding the diagnosis of PMP during hernia repair (3-6). Recently, our surgical team treated a patient with PMP arising from mucin-producing appendiceal tumor, who presented initially with a new-onset inguinal hernia.

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## Case report

A 67-year-old male was referred to our hospital with lower abdominal pain and gradual abdomen distension. One month earlier the patient was operated in a local hospital with a left inguinal hernia and during herniorrhaphy, a large amount of gelatinous mucinous material was found in the indirect-hernia sac and a diagnosis of PMP was established on cytological grounds – pseudomyxoma peritonei. On physical examination, abdominal ascites was noted. All routine laboratory parameters were within normal limits. The serum concentration of CEA and CA 19-9 tumor markers was elevated. A CT scan of the abdomen revealed a huge mass consisting of a low attenuation mucinous material in the abdominal (Fig.1A) and pelvic cavity (Fig.1B).

At laparotomy, a massive gelatinous ascites (Fig.1C), multiple peritoneal and greater omentum implants (Fig.1D) and a distended appendix were found. Aggressive surgical cytoreduction, peritonectomy, appendectomy, and greater omentectomy were performed. We considered right-sided hemicolonectomy unnecessary since our patient was diagnosed with non-aggressive peritoneal mucinous lesion. The postoperative completeness of cytoreduction (CCR) score was CCR-2 (7). During surgery about 4000 ml of mucinous ascites was evacuated using 5% dextrose solution as mucolytic agent. Finally, abdominal cavity lavage with a tumoricidal agent (5% povidone-iodine) was performed. Surgery was finalized with hyperthermic (42°C) intraperitoneal chemotherapy. Immediate postoperative intraperitoneal infusion of 5-fluorouracil (5-FU)

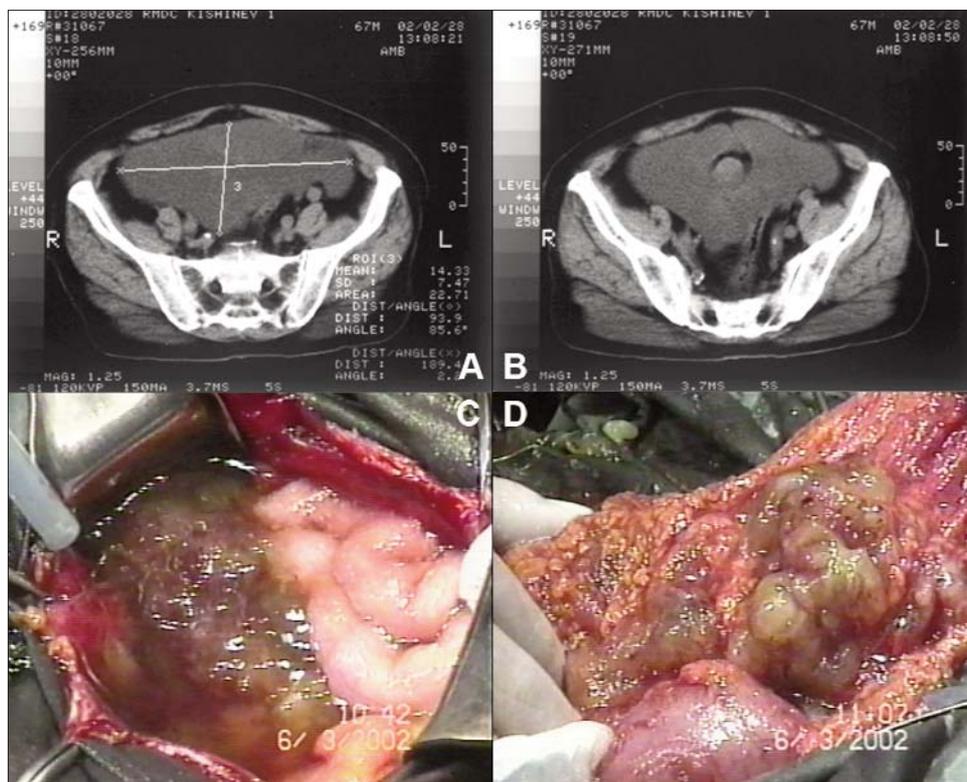
prepared in 5% dextrose peritoneal dialysis solution was initiated in a daily dose of 20 mg/kg during next 5 days.

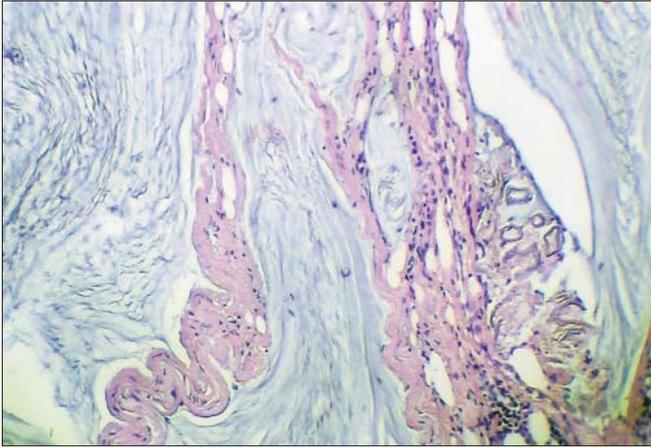
The pathologic examination of the surgical specimen demonstrated mucinous cystadenoma of the appendix and DPAM (Fig. 2) according to Ronnett's histopathological classification (8). The postoperative course was uneventful. Tumor markers, CEA and CA19-9, reduced to the normal range postoperatively. A follow-up CT scan revealed complete mucinous ascites disappearance, unfortunately, the patient died, apparently due to a cardiac event 15 month later, being free from PMP recurrence.

## Discussion

Pseudomyxoma peritonei first described by Werth is an uncommon and poorly understood disease characterized by abundant extracellular mucin in the peritoneum. The incidence of PMP is approximately two in 10.000 laparotomies, and about 75% of patients are female with an average age of 53 years (2). Recent progress in immunohistochemical techniques has showed that PMP is associated with mucin-producing appendiceal tumor (8). Although other sites of PMP, such as ovary, gallbladder, stomach, pancreas, colon, uterus, fallopian tubes, urinary bladder, breast and lung, have been reported (2). Initial symptoms differ greatly and depend on the localization of the disease. Patients present with the following symptoms: ovarian mass (39% of women), suspected acute appendicitis (27%), increasing abdominal girth (23%), inguinal hernia (25% of male patients, 4% of female patients),

**Figure 1.** Preoperative CT showing fluid collection in the peritoneal (A), and pelvic cavity (B). PMP in abdominal cavity (C). Mucinous implants on the surface of the greater omentum (D).





**Figure 2.** Disseminated peritoneal adenomucinosis. (H&E, 80)

ascites (4%) and non-specific complaints (9%) (3). The possible trigger mechanism for new onset of an inguinal hernia is thought to be that free-floating intraperitoneal tumor cells penetrate into the hernia sac via the persisting processus vaginalis (3). The CT scan is now widely employed to establish the diagnosis and extent of PMP (3,9).

Up to date the optimal treatment of patients with PMP remains poorly defined. The treatment of pseudomyxoma peritonei syndrome traditionally consisted of repeated operative procedures focusing on evacuating all free mucus and debulking as much tumor as possible (4). Although some authors have argued that surgical debulking of PMP should be performed on a selective basis, most agree that patients with PMP are best treated, at least initially, with aggressive local therapy (2). Sugarbaker group introduced cytoreductive surgery (CRS), which includes greater and lesser omentectomy, splenectomy, bilateral subphrenic peritonectomy, pelvic peritonectomy, rectosigmoid resection, and distal or total gastrectomy (10,11). According to González-Moreno S and Sugarbaker PH., right hemicolectomy does not confer a survival advantage in patients with mucinous appendiceal tumours with peritoneal dissemination unless metastatic involvement of the appendiceal or distal ileocolic lymph nodes is documented by biopsy, or the resection margin is inadequate (12).

Histologically, three main diagnostic categories are described: the benign form of disseminated peritoneal adenomucinosis (DPAM), the malignant form of peritoneal mucinous carcinomatosis (PMCA), and an intermediate form with features between DPAM and PMCA (8). This classification has important prognostic significance, because patients

with DPAM have a significantly better prognosis compared to those with PMCA (4). The majority of patients will eventually suffer recurrence. The 5-year survival rate ranges from 53 to 75 per cent, but outcomes vary widely between relatively benign and malignant subgroups (2, 10).

In conclusion, PMP is a rare, slowly progressive disease that produces extensive mucus accumulation within the abdomen and pelvis. Furthermore, our case, along with others, highlights that inguinal hernias can sometimes be the initial presentation of PMP.

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