



Case Report

A case of pseudomyxoma peritonei presenting as ventral and femoral hernia

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ABSTRACT

Pseudomyxoma peritonei (PMP) is a unique condition defined by a diffuse collection of mucinous gelatinous fluid in the abdominal cavity. The possible causes of PMP are mucin-producing cells from a ruptured cystadenoma or mucinous malignancies from appendix, colon, pancreas, stomach, and female adnexae. We report a case diagnosed as PMP that presented with multifocal hernias. An 89-year-old woman with history of appendiceal mucinous adenocarcinoma was found to have ventral and femoral hernia. Mucinous jelly-like fluid was found in the hernial sac intraoperatively and we confirmed the diagnosis of PMP via pathology. The symptoms and diagnosis, as well as special features of PMP, are reviewed. Current clinical concepts and treatment strategies are also discussed. The formation of a hernia is perhaps a presentation of PMP that clinicians may easily overlook.

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1. Introduction

Pseudomyxoma peritonei (PMP) is a unique condition defined by a diffuse collection of gelatinous fluid in abdominal cavity. The term PMP was originally applied to the spreading of mucin-producing cells from ruptured cystadenoma of ovary or appendix [1]. It was first described by Rokitsky in 1842 but Werth later named it “*pseudomyxoma peritonei*” in association with mucinous carcinoma of the ovary. The current concept indicates that not only dissemination of a ruptured cystadenoma but also mucinous malignancies from appendix, colon, lung, breast, pancreas, stomach, and female adnexae are also possible origins [2].

The most common presentations of PMP include increased abdominal girth, diffuse pain, and ascites [3]. However, these symptoms have limited diagnostic value and pathological evidence is critical for a definite diagnosis. Because the presentations are nonspecific and often appear unexpectedly, awareness of special symptoms and/or clinical settings have been described. One study indicated that up to 25% of male patients with PMP may present with recurrent inguinal hernias, whereas women present with an ovarian mass during pelvic examination [4]. We herein report

a female patient diagnosed with PMP who presented with multifocal hernias.

2. Case report

An 89-year-old retired female farmer visited our General Surgery clinic because of right lower quadrant and inguinal pain in recent 2 years. About 4 years ago, she presented with an acute abdomen and received a right hemicolectomy for ruptured appendiceal mucinous cystadenocarcinoma. The appendiceal tumor, measuring about $2.8 \times 2.4 \times 1.8$ cm, had invaded the mesoappendix and mesocolon as well as contagiously involving the muscular layer of transverse colon. The staging according to pathology and clinical image studies were T4N0M0. An incisional ventral hernia was found 1 year later and repaired primarily. The patient was otherwise healthy and this time she complained of pain and sensation of something “pushing out” over the umbilicus. The symptom were aggravated by bending forward and coughing but were usually reversible. In addition, she also complained of a small tender mass in the right inguinal area. She had been followed up at our clinic for her previous appendiceal cancer and the level of the tumor marker carcinoembryonic antigen had shown no elevation over this period. A series of abdominal computed tomography (CT) investigations showed a ventral hernia about 3.0×2.0 cm with omentum content (Fig. 1) that was accompanied by a right inguinal hernia (Fig. 3). The hernia content became heterogeneous when it was contrast enhanced showing small nodular changes in the upper part of the hernia (Fig. 2). No

Conflict of interest: none.

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Fig. 1. Abdominal CT showed one protruding hernia sac with hypodense homogenous soft tissue. This is compatible with a ventral hernia with reducible omentum content. No sign of cancer recurrence or abnormal fluid within the abdomen were found. CT = computed tomography.

tumor recurrence or abnormal fluid accumulation within abdomen was found.

Because our initial impression was a recurrent ventral hernia accompanied by a right inguinal hernia, herniorrhaphy was arranged. However, during the surgery, the ventral hernia sac appeared to be nodular and to have gelatinous fluid inside. While resecting the sac, a collection of pink, sticky, and jelly-like fluid of about 10 mL was found. The same finding, except that the amount was lesser of about 5 mL was also noted for her right inguinal lesion, which was later confirmed to be a femoral hernial sac. The sticky fluids in both hernias were confined within the sac as a cystic component and were without direct connection to the peritoneal cavity. Moreover, a highly strained femoral hernia sac with a very narrow neck was noted. PMP was confirmed by frozen section. We then performed laparoscopic exploration in detail of the whole abdomen but the results were negative for intraabdominal jelly-belly appearance and no suspicious tumors were discovered at the previous operative site. Based on the advanced age of the patient and the absence of any strong evidence of recurrence of the cancer, we did not carry out any other advanced treatment, such as intraperitoneal hyperthermic chemotherapy, after discussing the situation with her family. We drained the fluid, closed the fascial defects, and reinforced them with mesh.

The pathological report indicated profuse mucin pooling over the focal peritoneum and invasion of underlying hernial wall



Fig. 2. In the upper part of the ventral hernia sac, the presence of some heterogeneous contrast-enhanced substances with nodular lesions was noted.



Fig. 3. A pelvic CT showing one small hernial sac in the right inguinal area. No bowel involvement or incarceration was found. CT = computed tomography.

(Fig. 4). These findings suggested the spreading of well-differentiated mucin-producing cells and the morphological picture was that of metastatic mucinous adenocarcinoma, rpT4N0M1 (Fig. 5). By definition, the spreading of gelatinous fluid confirmed the diagnosis of PMP and its presentation as multifocal hernias was unexpected. Our patient recovered well without active complaints and was discharged on postoperative Day 5. She is currently being followed up at our General Surgery clinic for supportive care and undergoing further evaluation.

3. Discussion

PMP in clinical practice is often found incidentally and has a reported frequency of being present in 2/10,000 laparotomies [6]. The most common presentations have been diffuse abdominal pain, distension, or ascites [4,5]. Abdominal computed tomography is recognized as the standard for clinical diagnosis and features mucinous fluid accumulation in abdomen and displacement/scalloping of the adjacent organs [7]. Sugarbaker et al [11] described that the presence of a tumor greater than 5 cm on the jejunum, proximal ileum, or mesentery is associated with malignancy. Tumor markers, such as carcinoembryonic antigen, carbohydrate antigen 19-9, or carbohydrate antigen 12-5 may be elevated but share poor diagnostic values and are often saved for postoperative follow-up when there is advanced disease [8].

In addition to the common symptoms listed above, several studies have reported different settings. Baker and Goldman [9]

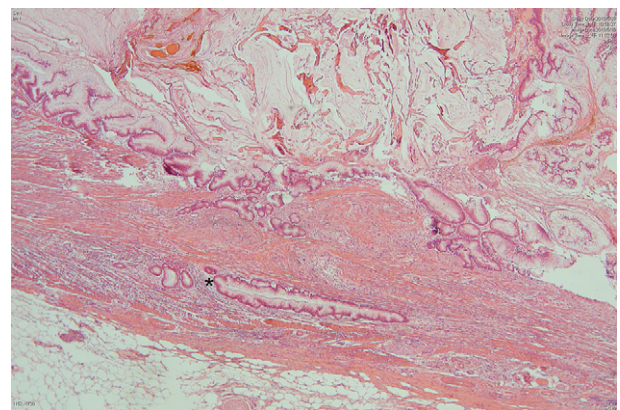


Fig. 4. A pathological specimen from the resected hernial sac. Diffuse mucinous jelly-like fluid and glandular structure were found. *Tumor invasion into the nearby peritoneum was also noted (Hematoxylin & Eosin 100 \times).

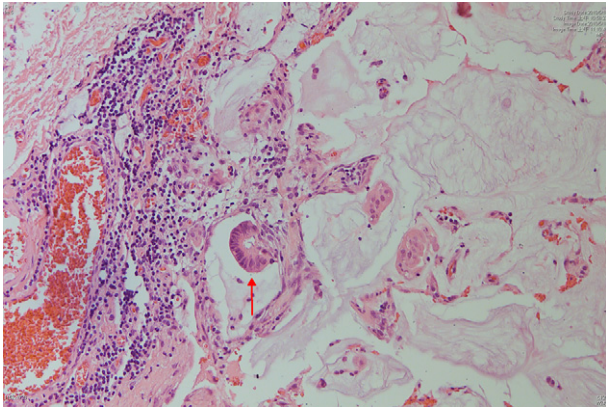


Fig. 5. One well-differentiated glandular tumor nest was found. Tumor seeding was confirmed (Hematoxylin & Eosin 400 \times).

described a case diagnosed with PMP who presented with a scrotal mass and urinary symptoms. Another case, reported by Lee et al [10], showed a right inguinal hernia from a ruptured cystadenoma of appendix. A case series demonstrated that up to 25% of male patients may show inguinal hernia together with the pathological finding of a mucinous tumor in the sac [4]. In our case, we found an elderly female patient with PMP who presented with ventral and femoral hernias at the same time and who had a history of appendiceal cancer. To the best of our knowledge, no similar case had been reported previously.

Sugarbaker et al [4,11] at the Washington Cancer Institute have published two series of treatment modalities for PMP. Aggressive cytoreduction surgery and intraperitoneal hyperthermic chemotherapy had been widely used with curative intent. However, these protocols are aimed at a different spectrum of the disease, one more closely associated with the concept of disseminated mucinous adenocarcinoma. In fact, PMP is considered a heterogeneous group of diseases that range across a spectrum from an indolent form of disseminated peritoneal adenomucinosis through an intermediate form to a malignant form with peritoneal mucinous carcinomatosis [12]. Ronnett et al [13] indicated the distinctive age-adjusted 5-year survival for the three categories to be 84%, 38%, and 7%, respectively. The debate on PMP is still open and currently the standard approach when treating PMP (defined as the indolent form, disseminated peritoneal adenomucinosis) remains symptomatic control and repeated debulking [14].

Our patient complained of tenderness over the ventral and femoral hernias. Originally, we thought that pain may be caused by an incarcerated intraabdominal organ, such as omentum. However, we found that the hernia sacs were filled with mucious fluid. The wall of femoral hernia had become highly strained because the fluid was confined within the sac because of its very narrow neck. To some extent, the pain in both areas could be explained by the

stretched peritoneum, which was caused by increasing non-releasable mucin within sac. A laparoscopic exploration was carried out during operation because we want to identify the presence of any residual mucious fluid or if there was a recurrent tumor within abdomen. To our surprise, the results were negative. Some residual PMP may have remained hidden in an interloop of the intestine or some other abdomen recess, this being easy to overlook during a laparoscopic examination. However, because the patient was elderly, we did not convert the operation into an open laparotomy to proceed with further advanced treatment.

In conclusion, we report a case of PMP because of appendiceal carcinoma that presented as ventral and femoral hernias without the presence of any common symptoms, such as distended abdomen or ascites. PMP was not diagnosed until the operation because of a lack of clinical clues from the initial laboratory findings and imaging study. We want to remind clinicians that hernia may be a presentation of PMP that is easy to overlook.

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