

# Urachal mucinous adenocarcinoma with pseudomyxoma peritonei

## A case report

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### Abstract

**Rationale:** Pseudomyxoma peritonei is an unusual clinical condition, and the appendix and ovaries are reported as the primary sites.

**Patient concerns:** A 44-year-old man who was reported a 3-month history of lower abdominal pain and distention, along with increased abdominal girth, was admitted with a palpable tender mass in the central lower abdomen.

**Diagnosis:** Ultrasonography showed a large well-circumscribed cystic-solid mass with lobulated margin, extending from the anterosuperior dome of the urinary bladder to the anterior abdominal wall. A computed tomography (CT) scan revealed a midline heterogeneous, hypodense, irregular polycystic-solid mass adjacent to the anterior wall of the abdomen and anterior to the dome of the urinary bladder. fluorodeoxyglucose positron-emission tomography/CT showed intense fluorodeoxyglucose uptake in the thickened wall of the mass. Intraoperative laparoscopic exploration also revealed a midline abdominal mass adjacent to the dome of the urinary bladder. Laparotomy showed that the mass originated from the dome of the urinary bladder and was disconnected with the urinary bladder lumen. The final histopathological diagnosis was urachal mucinous adenocarcinoma associated with high-grade pseudomyxoma peritonei.

**Interventions:** The patient underwent surgical cytoreductive procedure and the perioperative intraperitoneal chemotherapy.

**Outcomes:** The patient made an uneventful recovery, and 7 months later had no recurrence.

**Lessons:** The urachus is a tubular structure, which extends medially from the apex of the bladder to the allantoid during fetal development, and it usually obliterates after birth. Urachal remnants can cause urachal carcinoma or bladder cancers. Pseudomyxoma peritonei originating from mucinous neoplasm of the urachus is extremely rare.

**Abbreviations:** CT = computed tomography, FDG-PET/CT = fluorodeoxyglucose positron-emission tomography/computed tomography, PMP = pseudomyxoma peritonei.

**Keywords:** pseudomyxoma peritonei, urachal adenocarcinoma, urachus

## 1. Introduction

Pseudomyxoma peritonei (PMP) is an unusual clinical condition, and the appendix and ovaries are reported as the primary sites.

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The authors declare that they have no actual or potential conflicts of interest.

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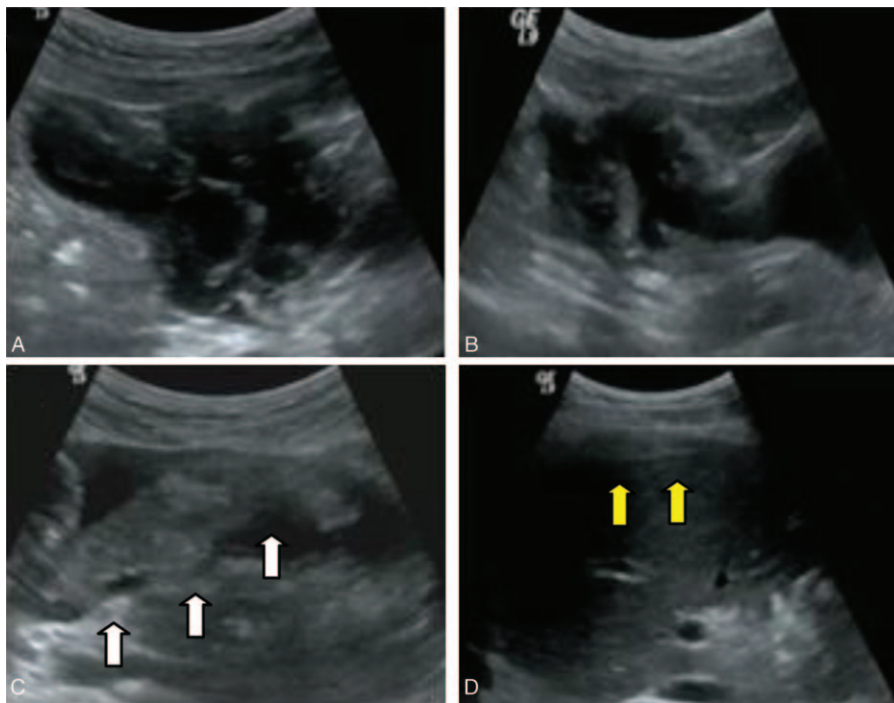
Although there is increasing knowledge of PMP, it remains a diagnostic challenge because of its rarity and lack of differential diagnostic symptoms. PMP should be considered in the differential diagnosis for abdominal and retroperitoneal tumors, especially with abdominal cystic-solid mass.

The urachus is a tubular structure, which extends medially from the apex of the bladder to the allantoid during fetal development. It usually obliterates after birth, and is converted into the median umbilical ligament. Its lumen is lined with a transitional epithelium that has the potential for focal glandular metaplasia, possibly followed by malignant transformation. Urachal remnants can cause diverse abnormalities.<sup>[1]</sup> Urachal carcinoma accounts for 0.01% of all malignancies,<sup>[2]</sup> and 0.17% to 1% of all bladder cancers.<sup>[1,3]</sup>

## 2. Case report

A 44-year-old man with no significant past medical history was admitted in August 2016 with a mass in the inguinal region. The patient reported a 3-month history of lower abdominal pain and distention, along with increased abdominal girth.

His physical examination showed a palpable tender mass in the central lower abdomen. Tumor markers were positive: carcinoembryonic antigen was 25.12 ng/mL, and carbohydrate antigen 19-9



**Figure 1.** (A) Ultrasonography showed a large well-circumscribed cystic-solid mass with lobulated margin, measuring  $42 \times 44 \times 78$  mm. The inside of the mass was heterogeneously hyperechoic, with some calcific foci within the wall. (B) The mass extended from the anterosuperior dome of the urinary bladder to the anterior abdominal wall. (C) A small volume of ascites was detected in the peritoneal cavity surrounding the mass, with localized thickened omentum (white arrow) inside. (D) Substantial pressure trace was detected around the liver (yellow arrow).

was  $73.7$  U/mL. Laboratory investigations revealed white cell count of  $14.26 \times 10^9$ /L with 5.9% lymphocytes. Other renal and liver function tests, urine analyses, and chest radiography before surgery showed no abnormalities.

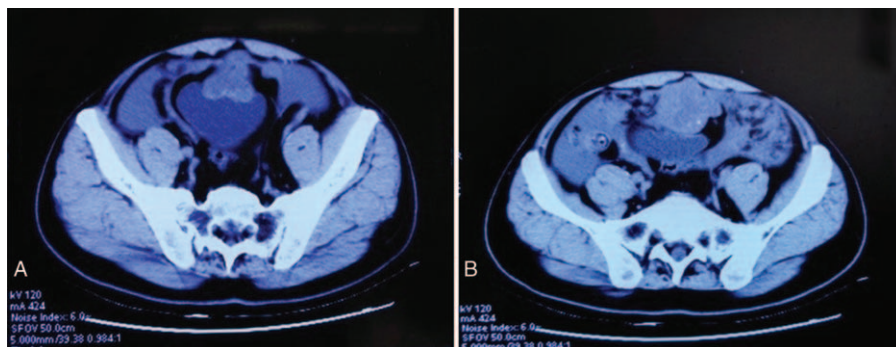
### 2.1. Imaging examinations

Ultrasonography showed a large well-circumscribed cystic-solid mass with lobulated margin, measuring  $10.2 \times 4.4 \times 7.8$  cm, extending from the anterosuperior dome of the urinary bladder to the anterior abdominal wall. The inside of the mass was heterogeneously hyperechoic, with some calcific foci within the wall. A small volume of ascites was detected in the peritoneal cavity surrounding the mass, with localized thickened

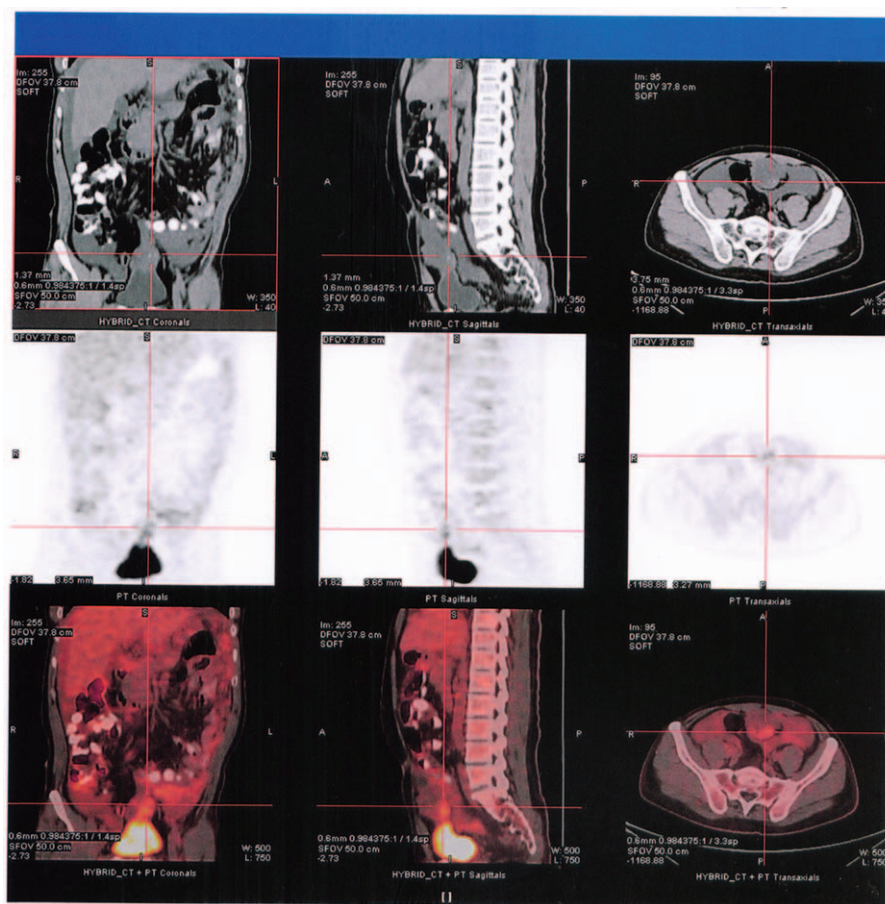
omentum. Substantial pressure trace was detected around the liver (Fig. 1).

A computed tomography (CT) scan of his abdomen and pelvis revealed a midline heterogeneous, hypodense, irregular polycystic-solid mass adjacent to the anterior wall of the abdomen and anterior to the dome of the urinary bladder (Fig. 2). Amorphous calcifications within the wall were also detected. The substantial wall of the mass was well-enhanced on contrast-enhanced images.

$^{18}$ F-fluorodeoxyglucose positron-emission tomography/CT (FDG-PET/CT) showed intense FDG uptake in the thickened wall of the mass with  $SUV_{max}$  of 2.9 (Fig. 3). The radioactivity uptake was  $SUV_{max}$  3.6, increased by 24%, after delayed scanning. Mucinous tumors were considered.



**Figure 2.** (A) A computed tomography (CT) scan revealed a midline heterogeneous, hypodense, irregular polycystic-solid mass adjacent to the anterior wall of the abdomen and anterior to the dome of the urinary bladder. (B) Amorphous calcifications within the wall were also detected. The substantial wall of the mass was well-enhanced on contrast-enhanced images.



**Figure 3.** FDG-PET/CT showed intense FDG uptake in the thickened wall of the mass with  $SUV_{max}$  of 2.9, and the radioactivity uptake was  $SUV_{max}$  3.6, increased by 24%, after delayed scanning. FDG-PET/CT=fluorodeoxyglucose positron-emission tomography/computed tomography.

## 2.2. Surgical findings

Intraperitoneal laparoscopic exploration revealed a midline abdominal mass adjacent to the dome of the urinary bladder. The peritoneal and omentum surfaces were involved by disseminated gelatinous tumor implants. Yellowish ascitic fluid was identified in the abdominal cavity, including the vesicorectal space.

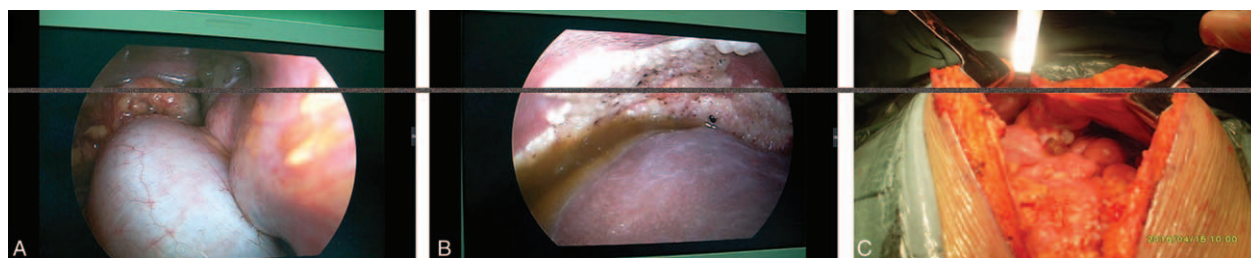
Then laparotomy was performed, which showed that the mass originated from the dome of the urinary bladder and was disconnected with the urinary bladder lumen (Fig. 4).

Peritoneal Cancer Index (PCI)<sup>[4]</sup> before cytoreductive surgery was 21, and the PCI after cytoreductive surgery was 0.

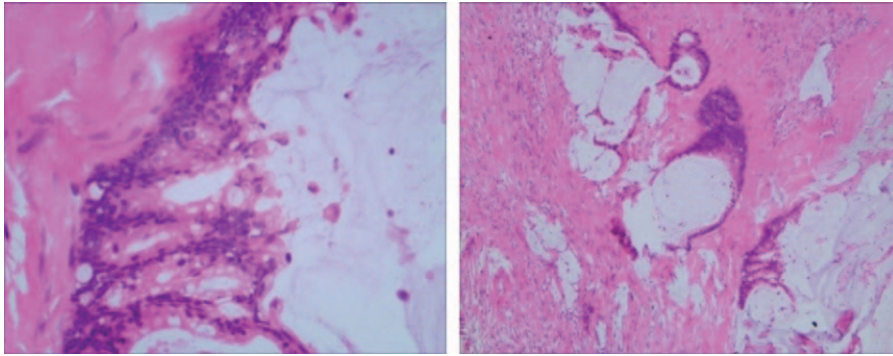
## 2.3. Pathological findings

Gross examination revealed a cystic-solid mass with abundant yellowish mucoid fluid, measuring 11.0 × 5.0 × 8.0 cm, connected to a small cuff of seromuscular layers of the bladder wall.

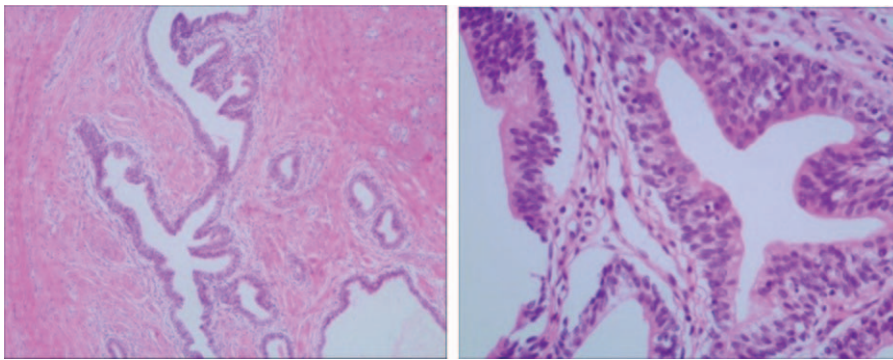
Histologically and cytologically, the cyst contained abundant mucoid fluid within the cavity. Parts of the cystic cavity were lined by tall columnar epithelium, which had a papillary and villiform structure. Areas of dysplasia with high-grade nuclear atypia, cellular crowding, and stratification were observed (Fig. 5). Some urachal remnants were observed near the cyst (Fig. 6). The final histopathological diagnosis was urachal mucinous adenocarcinoma associated with high-grade pseudomyxoma peritonei.



**Figure 4.** (A) Intraperitoneal laparoscopic exploration revealed a midline abdominal mass adjacent to the dome of the urinary bladder. (B) The peritoneal and omentum surfaces showed disseminated gelatinous tumor implants during laparoscopy. (C) Laparotomy was performed, which showed that the mass originated from the dome of the urinary bladder and was disconnected with the urinary bladder lumen.

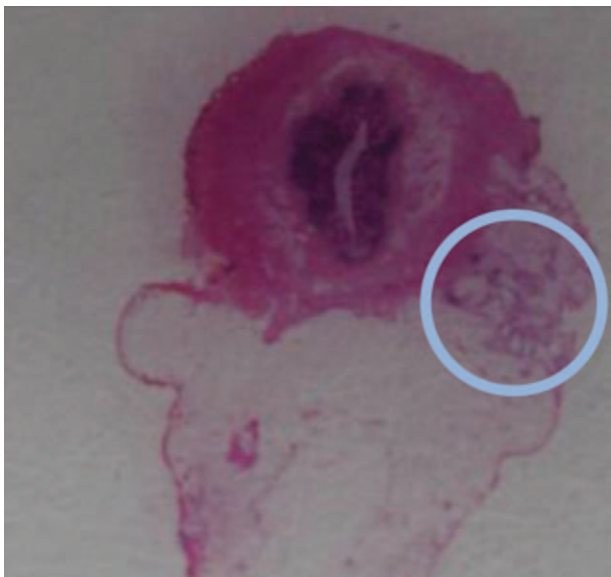


**Figure 5.** The cyst contained abundant mucoid fluid within the cavity, and parts of the cystic cavity was lined by tall columnar epithelium, which had a papillary and villiform structure. Areas of dysplasia with high-grade nuclear atypia, cellular crowding, and stratification were observed.



**Figure 6.** Urachal remnants were observed near the cyst.

Complete appendix sampling was conducted, which showed no neoplastic finding in the mucosal layers of the appendix (Fig. 7). Only parietal serosal layers were involved from outside to inside, which indicated that the appendix was not the primary focus.



**Figure 7.** No neoplastic finding was observed in the mucosal layers of the appendix. Only parietal serosal layers were involved from outside to inside (blue circle).

#### 2.4. Therapeutic intervention

The patient underwent surgical cytoreductive procedure. The urachal tumor, umbilicus, and disseminated gelatinous tumor implants on the peritoneum were resected, and partial resection of the bladder, appendectomy, and omentectomy was also performed. All omentum, diaphragmatic peritoneum, lesser omentum, round ligaments, part of falciform ligament, and all the nodules on the small intestine and mesentery of small intestine were carried out. Cholecystectomy with skeletonization of the hepatoduodenal ligament was also carried out. This procedure including all peritoneums which were infiltrated by cancer, resulted in a macroscopically complete cytoreduction (Sugarbaker completeness of cytoreduction score<sup>[5]</sup> of 0). Then the perioperative intraperitoneal chemotherapy was performed.

#### 2.5. Follow-up and outcomes

The patient made an uneventful recovery, and 7 months later had no recurrence through CT scan and ultrasound examination.

### 3. Discussion

Pseudomyxoma peritonei is a rare clinical condition characterized by extensive intraperitoneal spread of mucinous ascites, eventually leading to gross abdominal distension.<sup>[6–9]</sup>

The etiology is a mucinous neoplasm, most often arising in the appendix. PMP originating from mucinous neoplasm of the urachus is extremely rare. We identified about 28 previous cases in the literature.

Although there is increasing knowledge of PMP, it remains a diagnostic challenge because of its rarity and lack of differential diagnostic symptoms. In this case, a substantial cystic-solid mass was detected, extending from the anterosuperior dome of the urinary bladder to the anterior abdominal wall, which indicated urachal tumor. Additionally, ascites, localized thickened omentum, and substantial pressure trace around the liver were also detected, which indicated PMP.

As symptoms of this disease are nonspecific, it presents a major diagnostic challenge to clinicians. Clinical presentation is late and patients usually undergo prolonged health deterioration before an accurate diagnosis is made. PMP should be considered in the differential diagnosis for abdominal and retroperitoneal tumors, especially with abdominal cystic-solid mass.

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