中文題目：一位表現典型果醬狀腹內腫瘤及巨大網膜蛋糕的腹膜偽黏液瘤病例
英文題目：A Pseudomyxoma peritonei case presented with classical jelly-like tumors in the abdomen, and a huge omentum cake.
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***Introduction**:*

Pseudomyxoma peritonei (PMP) is a rare and slowly progressive tumor, with an estimated incidence of 1–2 per million per year. It was first described by Rokitansky in 1842 as a complicated peritoneum reaction caused by an ovarian neoplasm, featuring accumulation of jelly-like material and an increase in abdominal girth, giving the disease its classic sign “jelly belly.” Etiologies of PMP originate most plausibly from peritoneal dissemination of ruptured mucinous neoplasm from the gastrointestinal tract, typically the appendix, and other pelvic organs such as the ovaries. We reported a case of a 51-year-old female with PMP who had a classical presentation of jelly-like tumors in the abdomen, and a huge omentum cake found during the operation.

***Case Report**:*

A previously healthy 51-year-old female came to our clinic because of post-prandial nausea and vomiting for one month. She also had very poor appetite and had a body weight loss of 8 kilograms in less than 6 months. Abdominal computed tomography (CT) showed massive ascites and suspected PMP caused by bilateral ovarian mucinous tumors. Thus, she was admitted for further examination.

After admission, general blood examination showed only normocytic anemias, and elevated tumor markers of cancer antigen 125 (101.71 U/ml), carbohydrate antigen 19-9 (167.24 U/ml) and carcinoembryonic antigen (57.28). Initially, she underwent abdominal paracentesis, and a total of 175ml of light pink jelly-like ascites were sent for further study, but the cytology revealed negative of malignant cells. She then received open laparotomy performed by a Gynecologist. During the operation, a huge omentum cake and massive jelly-like pelvic tumor with adhesions were found. Biopsy to the peritoneal omentum, and excision of the pelvic tumor were performed. All the biopsy specimens revealed high grade PMP. The immunohistochemical (IHC) studies showed positive of CK20 and CDX-2, and

negative of CK7 and PAX8. Based on the histological features and IHC profile, the gastrointestinal tract was favored as the origin of the PMP. We adopted a comprehensive treatment of cytoreductive surgery (CRS) with hyperthermic intraperitoneal chemotherapy (HIPEC) for the patient as it is the optimal management for PMP currently.

***Discussion**:*

PMP is an extremely rare clinical syndrome, typically characterized by gelatinous myxoperitoneal effusion, progressive abdominal distention, and obstruction. Following rupture of the mucinous tumor, it initiates the development of PMP, allowing it to spread into the peritoneal cavity in the form of gelatinous deposits which causes an increase in intraabdominal pressure. Due to its indolent nature and non-specific symptoms, most are found incidentally in patients undergoing laparotomy, laparoscopy, or imaging for other medical conditions, making it hard to detect at early stages.

The origin of the primary tumor or PMPs has been widely debated, including the appendix, ovary, fallopian tube, pancreas, urachal tube, colon, and rectum. Based on clinicopathology and immunohistochemistry, researchers have recently confirmed that the majority of PMPs arise from ruptured mucinous tumors of the appendix. Although immunohistochemical markers (eg, CK7, CK20, CDX2, and SATB2) may aid in distinguishing the origin of PMP, to clarify the accurate origin of complicated PMP still remains challenging. Our patient was initially suspected of PMP of ovarian origin, however, as the IHC markers showed CK7-negative/CK20-positive, which is often associated with carcinomas of colorectal origin.

CT is the primary mechanism of diagnosis for PMP, with an appendiceal mucocele visualized as a calcified mass near the ileocecal valve. Classic radiographic features include loculated collections of fluid accumluating along peritoneal surfaces, resulting in a scalloped appearance of coated abdominal organs and omental caking. However, MRI is more sensitive at identifying whether the mucocele is mucin- or fluid-filled.

CRS and HIPEC are currently accepted as first-line treatment for selected PMP

patients. Gupta et al found that CRS with HIPEC achieve 83% five-year overall survival if the tumor is low grade, and 68% if the tumor is high grade. Nonetheless, despite treatment with CRS combined with HIPEC, disease recurrence was found to be as high as 28%. For the patients with PMP invading critical positions, studies of the pre-operation evaluation score to predict resectability in PMP are still ongoing.

***Conclusion**:*

PMP is a rare clinical disease that is difficult to detect at early stages and has potential for recurrence. Our patient, a 51-year-old female presenting with non-specific gastrointestinal symptoms but a classic jelly belly appearance, had typical PMP radiographic findings on abdominal CT. The disease was further confirmed by gelatinous mucus and a huge omentum cake found intraoperatively, and by histological and IHC stains. An optimal treatment of CRS with HIPEC was adopted.

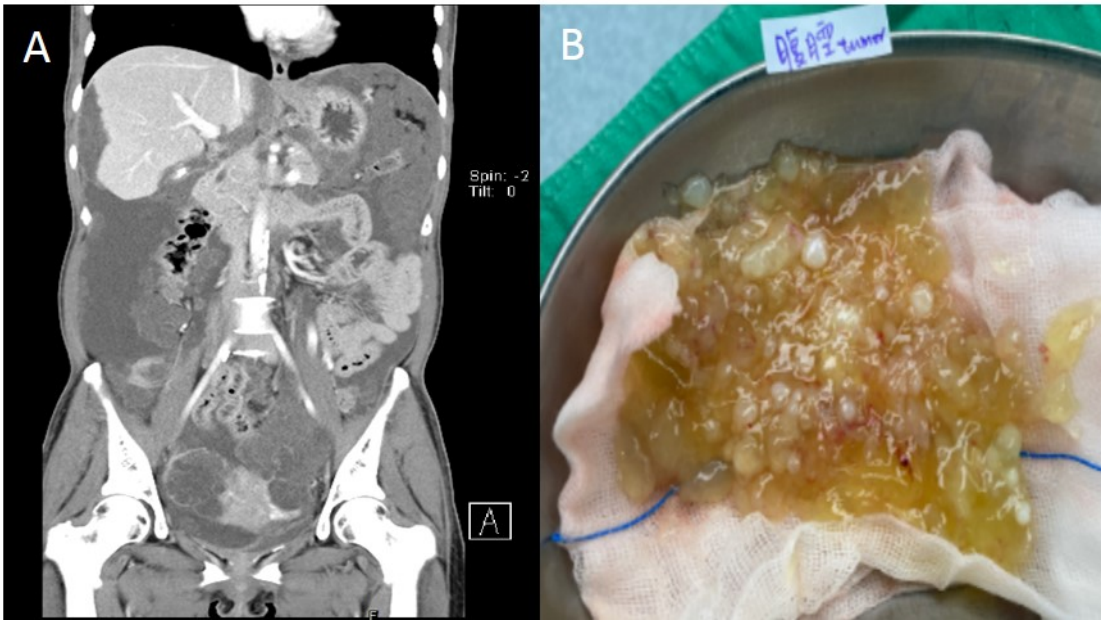

Figure 1. The typical Pseudomyxoma peritonei findings of the patient.

(A) the scalloped shape accumulation of the massive ascites on the abdominal CT scan; (B) the classic jelly-like appearance of the pelvic mucinous tumors

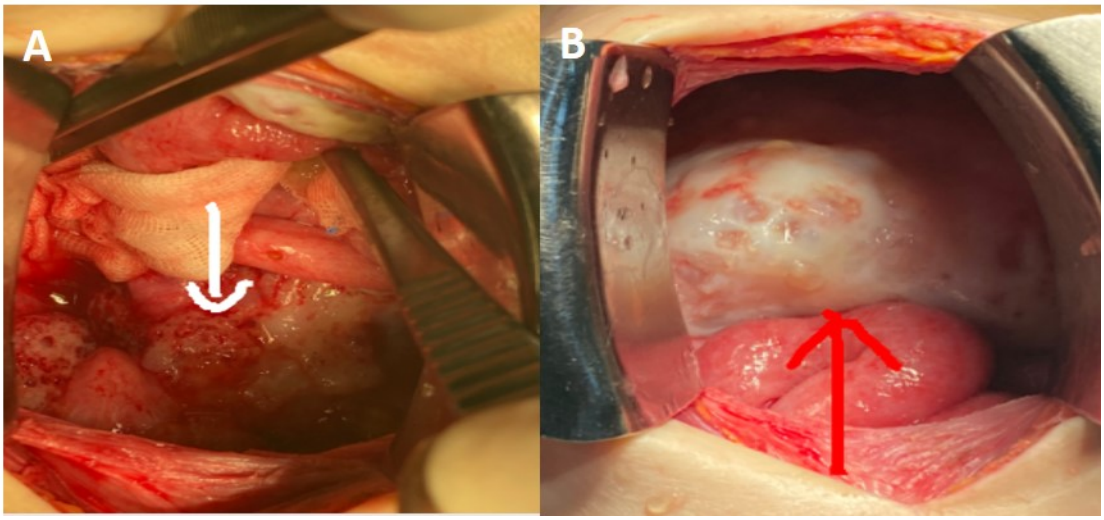

Figure 2. Intra-operative findings of the patient. (A) the gelatinous pelvic tumor with adhesions (white arrow); (B) a huge omentum cake (red arrow)