

# An Unusual Cause of Abdominal Distension

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## Case Presentation

Madam Wong was a 61-year-old woman who had medical history of hypertension, chronic hepatitis B infection, cholecystectomy and THBSO more than 15 years ago. She presented to the surgical unit because of progressive abdominal distension with significant weight loss in May 2000. CEA was markedly elevated. CT scan showed tense ascites with internal septations within ascitic fluid collection and omental cake suspicious of omental metastases. Bedside paracentesis was difficult as the ascitic fluid was viscous and jelly-like. The clinical diagnosis was Pseudomyxoma Peritonei. She initially declined laparotomy and was referred to palliative care clinic for follow up. However, because of the lack of tissue diagnosis as well as the worsening of symptoms, she finally underwent operation in December 2000. The operation included peritonectomy, omentectomy, total colectomy, total splenectomy together with intraoperative hyperthermic intraperitoneal chemotherapy. Histology confirmed well-differentiated adenocarcinoma with mesenteric lymph node metastases and pseudomyxoma peritonei. She had been symptom free and enjoyed good quality of life for one year after the surgery although there was recent gradual reaccumulation of ascitic fluid.

## Literature review

The term Pseudomyxoma Peritonei was first used by Werth in 1884<sup>1</sup>. It means diffuse collection of gelatinous fluid associated with mucinous implants on the peritoneal surfaces and omentum. It is 2 to 3 times more common in females than males and is present in 2 of every 10000 laparotomies<sup>2</sup>. The mean age of occurrence is 53 but the eldest reported age is 88<sup>3</sup>. However, the term is so non-specific and it has been so broadly applied that controversies regarding definition, pathology, site of origin, treatment and prognosis exist amongst pathologists, surgeons and gynecologists. The recent development of immunohistochemical techniques has led to a better understanding of the aetiology and pathogenesis of this disease<sup>4</sup>. It is now generally believed that Pseudomyxoma

Peritonei is a result of neoplastic mucus secreting cells within the peritoneal cavity that have low grade cytological features and are sparsely distributed within the extracellular mucinous collections. In most cases, these cells are appendix in origin, either from a ruptured appendiceal cyst, benign adenoma or a low grade appendiceal adenocarcinoma. Common clinical presentation includes progressive abdominal distension, fatigue, nausea and vomiting, abdominal mass (ovarian mass 30% in woman). Rarely, it may present as scrotal mass, hernia with mucinous tumor (<20% in man and <10% in woman) and appendicitis (<5%). Sometimes, this is an unexpected diagnosis made at laparotomy. The most commonly used means to diagnose pseudomyxoma peritonei pre-operatively is CT scan, which shows features such as mucinous ascites which is of fat density and with a heterogenous appearance. Also, scalloping of the liver, spleen and mesentery together with central displacement of bowel loops, ascitic septations and loculi, curvilinear calcification and omental thickening are also highly suggestive of Pseudomyxoma Peritonei in CT scan. For many years , conservative approach has been used. In recent decade, extensive surgical cytoreduction in combination with intraoperative hyperthermic intraperitoneal chemotherapy using 5-fluorouracil and mitomycin C has become the mainstay of treatment <sup>5, 6</sup> . The 5 year survival rates ranges from 53% to 75% <sup>6, 7, 8</sup> , but the outcomes vary widely between relatively benign and malignant subgroups <sup>9</sup> . However, recurrence nearly always occurs and may occasionally require re-operation. Other palliative method such as the use of mucolytic agents e.g dextrose solution <sup>10</sup> , was attempted with disappointing results. Repeated laparoscopic aspiration of the mucinous ascites as a palliative procedure was also reported <sup>11</sup> . But more studies are required before it can be widely adopted.

## **Conclusion**

Aggressive surgical approach with resection of the bulk of disease +/- intraperitoneal chemotherapy offers the optimal palliation and prognosis.

## **References**

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