



PSEUDOMYXOMA PERITONEI – when cancers play disguise

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Background

Pseudomyxoma peritonei (PMP) is a rare disease of the abdominal or peritoneal cavity.¹ The majority of cases result from a ruptured mucus-secreting tumour of the appendix.² More rarely, the condition arises in other parts of the bowel; in the ovary; and even more rarely, in the bladder.

The disease results in the presence of a large amount of mucinous fluid produced by cells that have spread and grown on the peritoneum lining the abdominal cavity. Left untreated, it will cause compression of the abdominal organs, making normal nutrition impossible.

Pseudomyxoma peritonei that starts in the appendix is said to have an incidence of around three to four per million per year.¹

Middle-aged women presenting with abdominal distension, ascites and ovarian mass are often presumed to harbour ovarian malignancy. Non gynaecological conditions such as PMP can mimic this presentation closely. E rare instances, a high index of suspicion prevents a delay in diagnosis and can potentially allow curative treatment.

Case Report

A 59 year old woman who had previously been well, presented at University Hospital Lewisham with progressively worsening abdomen distention, pain and weight loss.

She was initially under the lower GI team who were unable to complete a colonoscopy due to extrinsic compression. She was referred to gynaecology when an ovarian mass and ascites were seen on CT abdomen.

Gynaecology proceeded to a CT chest and MRI scan. Imaging demonstrated a large adnexal mass, mucinous loculated ascites and omental disease. An ultrasound guided drainage of ascites was attempted but only a "jelly-like" substance aspirated, there was no free drainage.

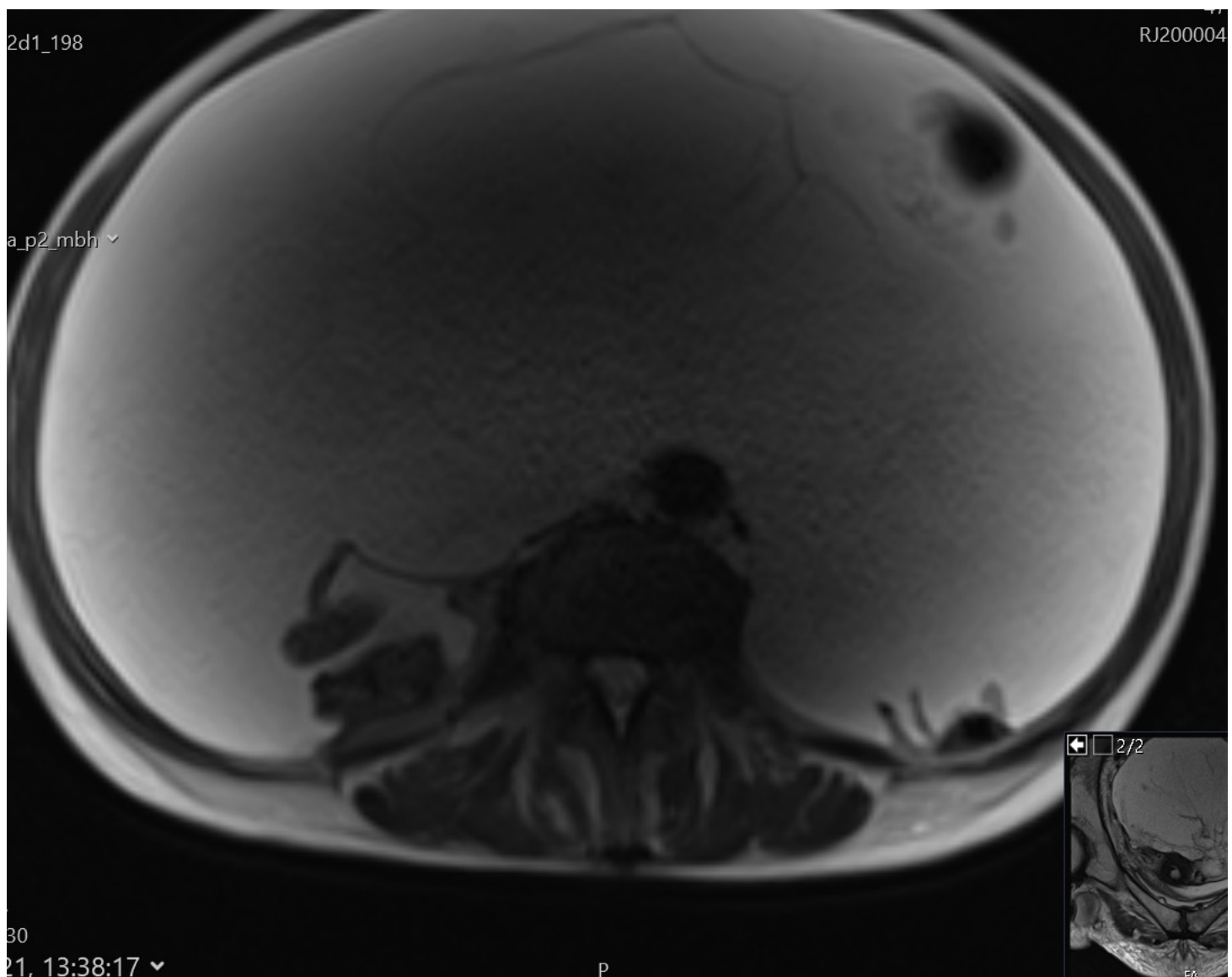
Tumour markers were raised.
Ca19-9 – 326
CEA – 21
Ca125 – 72.5

Case Report

After a multidisciplinary discussion, disseminated pseudomyxoma of gastrointestinal origin was suspected and she was referred to Peritoneal Malignancy Institute, Basingstoke for further management.⁴

She was in discomfort due to distended abdomen, had to be given parenteral nutrition and operated as emergency.

A laparotomy revealed a very large right Krukenberg with a large amount of mucinous ascites. This was arising from an appendix tumour. Otherwise she had classical PMP with disease on the surface of the liver, spleen, both paracolic gutters, and pelvis. There was mucin on the small bowel that wiped free with good small bowel. Complete cytoreduction was done for her. She was then given an hour of heated intraperitoneal chemotherapy with Mitomycin C at 41°C.



**MRI
abdomen
showing
mucinous
ascites**

Her histology was in keeping with low grade mucinous carcinoma peritonei. There were no lymph nodes involved. There was an incidental well differentiated neuroendocrine tumour in the caecum and she has been referred for a multidisciplinary discussion to determine whether this needs any further management.

She is recovering remarkably well from her procedure and has a follow up plan in place for imaging and blood markers to be repeated in a year.

Discussion

Mucinous ascites in women can originate from myriad cancers, the commonest being the appendix and ovary. Enhanced cognizance of the disease coupled with advances in investigative methods have allowed the organ of origin to be accurately identified, particularly, when there is doubt on initial investigation. Our patient benefitted from an expedited review at a dedicated centre and could receive relevant care in line with national timelines for cancer care.

Conclusion

All gynaecologists should be familiar with the possibility of rare presentation of non gynaecological malignancy masquerading as gynaecological one. High index of suspicion, use of centralised referral pathways and centres will allow timely and appropriate cancer care for these patients.

TRIVIA

The prefix 'myxo' arises from Latin 'muxe' for mucus

Often called 'Jelly Belly' in medical parlance for the mucus filled abdomen

Normal peristalsis and poor adherence of epithelial cells results in classical 'bowel sparing'

References

1. Smeenk RM, van Velthuysen ML, Verwaal VJ, Zoetmulder FA. Appendiceal neoplasms and pseudomyxoma peritonei: a population based study. Eur J Surg Oncol. 2008;34:196–201. doi: 10.1016/j.ejso.2007.04.002. [PubMed] [CrossRef] [Google Scholar]
2. Mittal R, Chandramohan A, Moran B. Pseudomyxoma peritonei: natural history and treatment. Int J Hyperthermia. 2017;33:511–519. doi: 10.1080/02656736.2017.1310938.
3. Esquivel J, Sugarbaker PH. Clinical presentation of the Pseudomyxoma peritonei syndrome. Br J Surg. 2000;87:1414–1418
4. Sulkin TV, O'Neill H, Amin AI, Moran B. CT in pseudomyxoma peritonei: a review of 17 cases. Clin Radiol. 2002;57:608–613.