Two Steps to "Jelly Belly" Appendiceal Mucinous Neoplasms and Pseudoymyxoma Peritonei

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Introduction

Pseudomyxoma Peritonei (PMP) is a complex disease characterised by an accumulation of mucinous ascites and peritoneal implants which generally arise from a perforated mucinous tumour of the appendix. A descriptive term used to describe this disease is "Jelly Belly".

The history and classification of PMP's and associated appendiceal mucinous neoplasms were somewhat contentious in the past. With the implementation of the Peritoneal Surface Oncology Group International Consensus Classification, PMP's and appendiceal neoplasms were categorised into three groups. These three groups are:

1. Low grade,
2. High grade,
3. High grade with signet cells.

The role of Anatomical Pathology and PMP

In our diagnostic laboratory, typically we receive 5-10 specimens from patients with PMP. These include right or extended colectomy, paracolic gutters, pelvic peritoneum, uterus, tubes and ovaries, gall bladder, falciform ligament and diaphragmatic peritoneum to name a few. One of the main objectives for our pathologists when dissecting these specimens is to identify the appendix. If the appendix is present they determine if the appendix is intact or has an obvious perforation. Adequate samples are taken from all specimens to identify cells in the mucin and to grade the tumour accordingly.

These two cases illustrate the type of specimens received by the pathology laboratory and the application of the diagnostic terminology.

Case Report 1.

The clinical history was of a 36 year old male with a mucocoele of the appendix. Macroscopically the specimen consisted of a distended appendix 68mm in length and up to 53mm in width at its greatest diameter. On opening the appendix numerous cream soft spherical particles were present measuring 1-2mm in diameter. (Fig. 1) No mass was evident in the appendiceal wall and no nodes were identified.

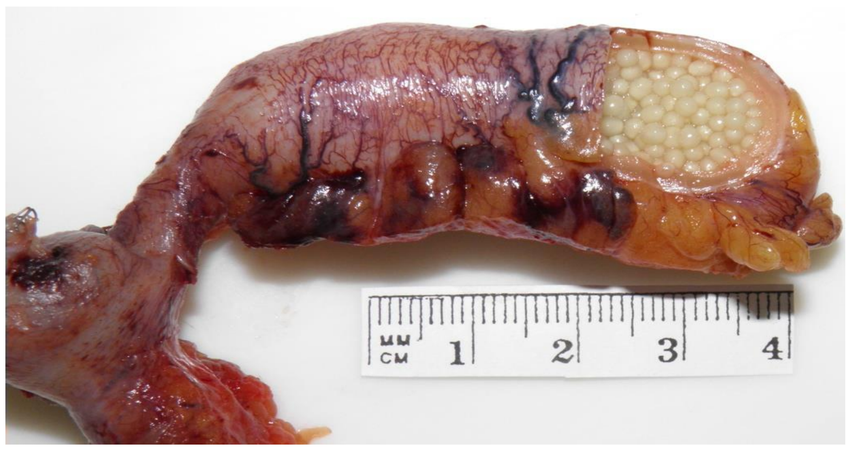

Figure 1. Macroscopic appearance of appendix

Results

Histologic sections show distension of the distal appendix with intraluminal accumulation of mucin spherules (a rare phenomenon). The lining is glandular with low grade dysplasia (Fig 2). There is no evidence of malignancy. Local excision is complete. The final diagnosis was low grade appendiceal mucinous neoplasm.

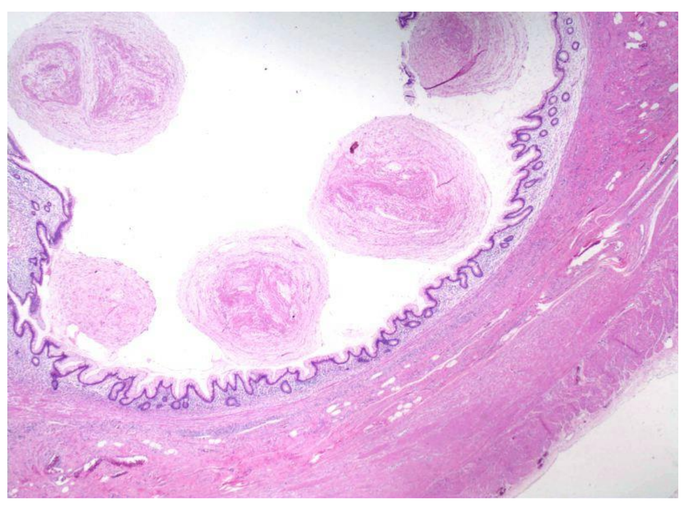

Figure 2. Intraluminal mucin spherules and low grade mucinous epithelium

Case Report 2.

The clinical history was of a 31 year old male with multivisceral mucinous deposits.

Multiple separate specimens were received including distal gastrectomy and omentum, right diaphragm, colon, spleen, stomach, rectum, pancreas, liver, ileocolic node and gallbladder (Fig 3).

On initial macroscopic examination the specimens were found to be caked with gelatinous tumour (Fig 4). The appendix was identified and noted to be dilated, filled with mucin and showed an apparent distal rupture.

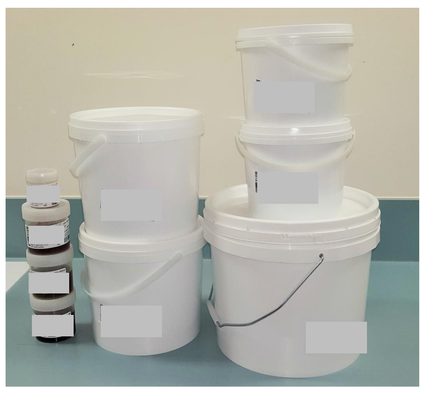

Figure 3. Multiple separately submitted specimens

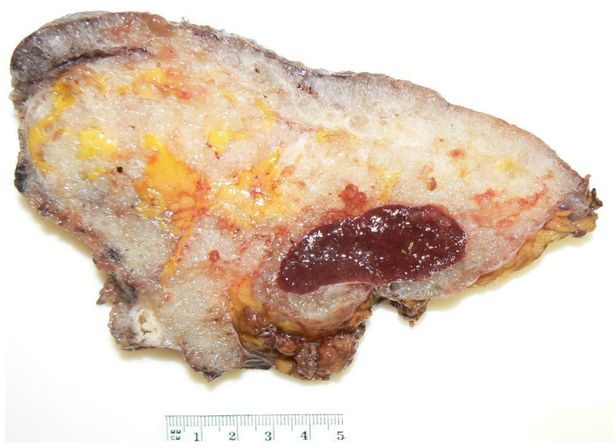

Figure 4. Spleen surrounded by mucinous tumour

Results

Histological sections show a mucin-filled dilated appendix, lined by high grade dysplastic papillary epithelium (Fig. 5). There is infiltration of the ileal wall and other sites by tumour epithelium (Fig. 6). Single signet ring type cells are seen. The final diagnosis was high grade appendiceal mucinous neoplasm and high grade pseudomyxoma peritonei.

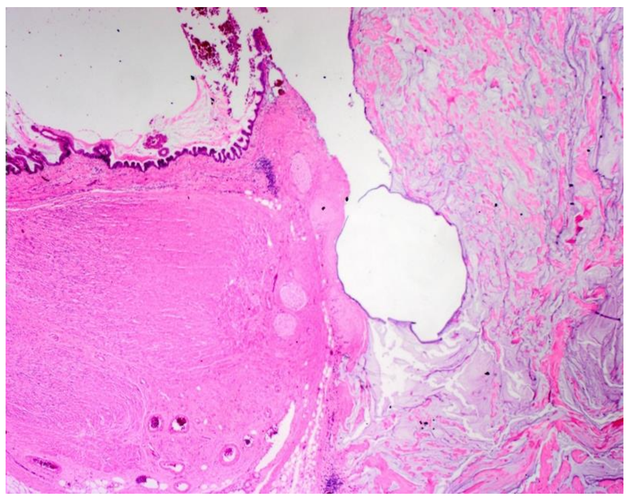

Figure 5. High grade dysplastic epithelium lining appendix

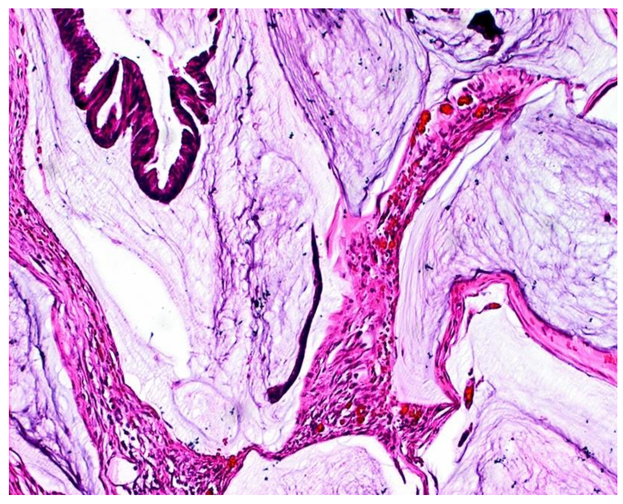

Figure 6. High grade PMP

Conclusion

PMP demonstrates the re-distribution phenomenon where there is accumulation of mucin and mucinous epithelium in various sites where re-uptake of peritoneal fluid occurs, such as omentum, paracolic gutters, large bowel, liver and sub-diaphragm. The treatment of patients with PMP is typically with aggressive cytoreductive surgery (CRS) to obtain complete macroscopic clearance of the disease. This can be coupled with perioperative intraperitoneal chemotherapy. A study of 2298 patients with PMP, whom all underwent CRS saw a median survival rate of 16.3 years as was quoted in a journal article of clinical oncology.

References

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