Mucinous Tumours of the Appendix and the PMP syndrome

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A rose by any other name...

• ‘Peritonitis myxomatosa chronica’ (Virchow)
• ‘Maladie gélatineuse du péritoine’ (Péan)
• ‘Pseudomyxoma peritonei’ (Werth, 1884)

Richard Werth 1850-1918
Origin of PMP

• Originally: ovary thought to be primary site
• Accumulating body of evidence points to appendiceal primary in most cases
• Rarely: gallbladder, stomach, colorectum, pancreas, fallopian tube, urachus, lung, and breast
Histological classification

- Confusing
- Various systems used historically
- 2015 consensus:
  - PMP with low grade features
  - PMP with high grade features
  - PMP with signet ring cells
Symptoms

• Increasing abdominal girth
• Umbilical or inguinal hernia
• Ovarian mass
• Often incidental finding
Staging

- **Imaging**
  - CT scan
    - Mucinous ascites, omental ‘cake’, thickened peritoneum
    - ‘Scalloping’ of the liver and spleen
  - Diffusion weighted whole body MRI
  - Important: extent (PCI), small bowel involvement, porta hepatis involvement
  - $^{18}$F PET-CT
- **Serum tumour markers:** CEA, CA19-9, and CA125
- **Colonoscopy**
  - mucus from appendiceal lumen
  - Exclude synchronous colon lesions
- **Diagnostic laparoscopy**
Treatment

• Repeat surgery to alleviate symptoms
• Complete cytoreductive surgery
• Hyperthermic intraperitoneal chemoperfusion (HIPEC)
• Early postop IP chemotherapy (EPIC)
• Systemic (IV) chemotherapy
  – Before surgery (preop, neoadjuvant)
  – After surgery
  – Both
Cytoreductive surgery

• Combination of:
  – Organ resection
    • Common: ileocaecal, colon, rectum
    • Sometimes: spleen, pancreas tail, liver, ureter...
  – Peritonectomy: stripping of peritoneal lining from underlying muscle

• Time consuming (8-12 hours)
Fig. 6. Left subphrenic peritonectomy.
HIPEC

• After completion of surgery
• Variety of drugs and methods used worldwide
  – Duration: 30-120 minutes
  – Chemotherapy used: mitomycin C, oxaliplatin,…
  – Temperature used: 38-42°C
  – Open, semi-closed, or closed abdomen
Systemic (IV) chemotherapy

• To be considered in patients with high risk of recurrence
  – High grade or signet ring cell type
  – Elevated CEA
  – Adverse histological features
• Agents used as in colorectal cancer
  – Oxaliplatin, irinotecan, 5-fluorouracil
  – Biologicals: bevacizumab (Avastin), cetuximab
• Duration: 3-6 months
• Usually very well tolerated
Quality of life after CRS and HIPEC

• Significantly affected by:
  – Postoperative complications
  – Extensive small bowel resection; presence of ileostomy (‘bag’)
  – Removal of rectum → ‘low anterior syndrome’
  – Removal of (part of) stomach
• Usually returns to normal after 3-6 months
• Most common persisting symptoms:
  – Loss of energy, feeling tired
  – Delayed gastric emptying
QoL after CRS and HIPEC

• Does not always receive due attention in clinical studies
• Importance of multidisciplinary support:
  – Stoma nurse
  – Dietician
  – Oncology nurse
  – Psychologist
  – etc