Retroperitoneal fibrosis
Inflammatory fibrotic process in the retroperitoneum causing compression of retroperitoneal structures, including ureters.
Incidence 1:200,000
Peak age 40-60
Males > females 2:1

Aetiology

**Idiopathic (70%)**

? immune response to ceroid, a polymer of oxidised lipids and protein from atherosclerotic plaques

**Drugs**

- Methysergide* migraine treatment
- LSD*
- Bromocriptine*
- Methyldopa* *all ergot alkaloids
- Beta blockers
- Phenacetin
- Amphetamine

**Malignancy**

- Lymphoma
- Sarcoma
- Carcinoma of breast, colon, stomach and prostate

**Infection**

- TB
- Schistosomiasis
- Chronic UTI
- Syphilis
- Gonorrhoea
- *Actinomyces*

**Radiation**

**Inflammatory**

- IBD
- Sarcoidosis
- Endometriosis
- Collagen diseases
- Inflammatory AAA
- Multifocal fibrosclerosis rare condition characterised by RPF mediastinal and mesenteric fibrosis sclerosing cholangitis, Reidel’s thyroiditis and orbital pseudotumour

**Presentation**

Relatively non-specific often with delayed diagnosis

- Vague back pain
- Lethargy
- Anorexia
- Weight loss

Symptoms attributable to disease process

**Ureters** oliguria, weight gain, SOB
**IVC** DVT
**Renal vein** hypertension, haematuria
Retroperitoneal fibrosis

Pathology
Smooth, flat tan-coloured mass of tissue overlying ureters

Histology
<table>
<thead>
<tr>
<th>Early phase</th>
<th>Late phase</th>
<th>Malignancy RPF</th>
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<tbody>
<tr>
<td>collagen and inflammatory cells</td>
<td>collagen</td>
<td>collagen and occasional islands of tumour cells</td>
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Diagnosis
Raised ESR, CRP, anaemia and leucocytosis common but non-specific
No current diagnostic serological test
IVU shows varying degree of obstruction with medial deviation of ureters (NB. medial deviation seen in ~20% of normal individuals)
CT or MRI with or without contrast are the investigations of choice
Except in a few obvious cases of lymphoma, cross-sectional imaging not sufficient to exclude malignancy: Campbell's states that:
‘Representative biopsies of the mass need to be obtained ..to rule out malignancy and allow one to proceed with treatment for RPF’ However some believe that biopsy not required if:
- Classic features of RPF on CT/MRI
- No lymphadenopathy
- No history of prior malignancy
- ? a role for PET scanning in this situation to exclude requirement for biopsy - both lymphoma and sarcoma positive on FDG-PET

Management
Initial management comprises decompression of urinary tracts or Mx of DVT
Primary high dose steroid therapy
Effective in ~80%
?better in those with high ESR, leucocytosis or inflammatory cells on biopsy
Duration of treatment unknown; > 6 months recommended, but because of relapse rate (?value) some advocate longer term Rx
Rx schedule (Kardar 2002) in Campbell’s:

(ESR). The steroids were started with an initial oral intake of 60 mg. prednisolone on alternate days for 2 months. This dose was tapered during the next 2 months (40 mg. for 2 weeks, 20 mg. for 2 weeks and 10 mg. for 2 weeks) to a maintenance dose of 5 mg. daily. Total duration of steroid use was 2 years.

Steroid-sparing immunosuppressants including azathioprine, cyclosporine, mycophenylate mofetil and tamoxifen all reported to have efficacy. Possible role for non-steroid immunosuppressants in preventing relapse after short-course of high-dose steroids (Swartz RD 2009)
Surgical management for non-responders:
- Open ureterolysis
- Midline incision
- Medial mobilisation of left and right colon
- Initial biopsy +/- frozen section
- Right-angle from normal ureter to abnormal
Placement of ureters in peritoneum or omental wrap
If inadvertent uretotomy leave stents longer post-op

Laparoscopic ureterolysis
First report Clayman and Kavoussi 1992
Data on 13 patients reported in 2002
All pre-stented and placed in peritoneum by tacking white line back to original site *underneath* ureter
Conversion in 15% for bleeding/failure to progress; no intrabdominal complications; 92% unobstructed at 30 months