

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 |

Pathology

Smooth, flat tan-coloured mass of tissue overlying ureters

| | | |
|-----------|----------------|---|
| Histology | Early phase | collagen and inflammatory cells |
| | Late phase | collagen |
| | Malignancy RPF | collagen and occasional islands of tumour cells |

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 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