Retroperitoneal Fibrosis

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Objectives

1. To Review the Epidemiology and Etiology of Retroperitoneal Fibrosis
2. To Discuss the Evaluation of Patients with Retroperitoneal Fibrosis
3. To Provide an Overview of Medical and Surgical Options in the Management of Retroperitoneal Fibrosis
Objectives

4. To avoid mentioning the 70 cm snowfall at Whistler over the weekend

Sources

Retroperitoneal fibrosis, A. Vaglio, C. Salvarani, and C. Buzio
Lancet 2006; Vol. 367
Definition

• An inflammatory, fibrotic process in the retroperitoneum encasing the aorta, IVC, and their branches, and the ureters, other retroperitoneal structures, and at times intraperitoneal structures including the GI tract

History

• First description 1905
  – Albarran (French Urologist)
    • Reported surgical treatment of RPF with ureteral obstruction

• Officially acknowledged 1948
  – Ormond (English Urologist)
    • Published 2 cases in English
Synonyms

- Ormond’s disease
- Periureteritis fibrosa
- Periureteritis plastica
- Chronic periureteritis
- Sclerosing retroperitoneal granuloma
- Fibrous retroperitonitis

Epidemiology

- Incidence: 1:200,000-500,000 per year
- M:F = 2-3:1
- No clear ethnic or genetic predisposition
- Mean age: 50 (40-60)
- Uncommon in children + elderly but has been described...
RPF in Children

Idiopathic retroperitoneal fibrosis in children.

Chan SL, Johnson HW, McLoughlin MG.

A case of idiopathic retroperitoneal fibrosis in an 11-year-old boy is presented. A review of 7 previous case reports revealed that there was no characteristic presentation in this disease process and that the diagnosis usually is suspected on an excretory urogram. The treatment is primarily surgical but corticosteroids may be helpful in selective cases.
Location

- Borders
  - Center
    • L4-5
  - Superior
    • Renal pedicle
    • Rarely extends to mediastinum
  - Inferior
    • Pelvic brim
    • Rarely extends to pelvis
  - Lateral
    • Outer edge of psoas
    • Envelopes ureters

- 15% extension outside retroperitoneum

Causes

- Idiopathic (70%)
- Other (30%)
  - Malignancy (8-10%)
  - Medications and chemicals
  - Retroperitoneal injury
  - Infections
Causes

- Idiopathic...
  - Immune-mediated chronic peri-aortitis
    - Leakage from atherosclerotic plaques
      - Ceroid (polymer of oxidized lipid + protein)
      - Oxidized LDL
    - ↑ Incidence of RPF in patients with atherosclerosis + AAA
    - Constitutional symptoms
    - Associated with other autoimmune diseases
      - SLE
      - RA
    - ↑ Acute phase reactants
    - ↑ Autoantibodies
    - Response to steroids + immunosuppression
    - Infiltrate includes plasma cells + lipid-laden macrophages

Idiopathic RPF
Causes

• Medications
  – Ergotamine alkaloids $\rightarrow$ serotonin
    • Methylsergide (Sansert)
    • LSD
  – Alpha-blockers
    • Methyldopa
    • Hydralazine
  – Beta-blockers
  – Dopamine agonists
    • Pergolide
    • Pramipexol
  – Phenacetin
  – Haloperidol
  – Amphetamines

Causes

• Chemicals
  – Asbestos
  – Talcum powder
  – Avitene
  – Methyl methacrylate
Causes

• Malignancies
  – Primary retroperitoneal
    • Lymphoma (most common)
    • Sarcoma
    • Multiple myeloma

Causes

• Malignancies
  – Metastatic retroperitoneal disease
    • Prostate
    • Colorectal
    • Breast
    • Pancreatic
    • Gastric
    → Desmoplastic reaction
Causes

• Malignancies
  – Carcinoid
    • Produces RPF without mets to RP
    • Seratonin-mediated
    • Profibrogenic growth factors
      – PDGF
      – IGF
      – EGF
      – TGF\(\alpha+\beta\)

Causes

• Retroperitoneal injury
  – Radiation
  – Trauma
    • Organizing retroperitoneal hematoma
  – Surgery
    • Retropertioneal
    • Intraperitoneal
  – Urinary extravasation
Causes

- Infectious
  - Repeated cholangial infections
  - TB
  - Chronic UTI
  - Actinomyces
  - Gonorrhea
  - Schistosomiasis
  - Syphilis
  - Ruptured liver echinococcal cyst

Associated Conditions

- Up to 15% of cases
- Underdiagnosed
- Presentation
  - Synchronous
  - Metachronous
Associated Conditions

- Autoimmune multifocal fibrosclerosis (rare)
  - RPF
  - Sclerosing mediastinitis
  - Sclerosing cholangitis
  - Orbital pseudotumour
  - Riedel’s thyroiditis

Related Conditions:

- Hashimoto’s thyroiditis
- Riedel’s thyroiditis
- Graves’ disease
- Small and medium-sized vessel vasculitis
- Wegener’s granulomatosis
- Polyarteritis nodosa
- Microscopic polyangiitis
- Hepatitis C virus-related cryoglobulinaemia
- Ankylosing spondylitis
- Systemic lupus erythematosus
- Rheumatoid arthritis
- Glomerulonephritis
- ANCA-positive rapidly progressive glomerulonephritis
- Membranous nephropathy
- Sclerosing cholangitis
- Primary biliary cirrhosis
- Sclerosing pancreatitis
- Uveitis

*ANCA, anti-neutrophil cytoplasmic antibodies.*
Presentation

• Symptoms
  – Non-specific
  – Insidious onset (4-6 months)
• PE
  – Typically unremarkable
• Labs
  – Non-specific

Symptoms

• Lower back or flank pain (90%)
  – Dull, NON-colicky, unchanged with posture
  – Radiation to lower abdomen or groin
  – Relieved by NSAIDS (not narcotics)
• Anorexia + nausea + weight loss + malaise
• Fever
• HTN 2’ RV encasement (up to 50%)
• Gross hematuria
• Testicular pain or swelling
• Claudication
• Lower extremity pain or edema
• Polyuria, oliguria, or anuria
• Constipation
Physical Exam

- Abdominal or lumbar tenderness
- Palpable abdominal mass
- Periumbilical bruit
- AAA
- Hydrocele or varicocele
- DVT
- Lower extremity edema

Laboratory Investigations

- Leukocytosis
- Eosinophilia
- Normochromic normocytic anemia
- ↑ Acute phase reactants (80-90%)
  - ↑ ESR
  - ↑ CRP
- + Autoimmune markers
  + ANA (60%)
  + RF
  + SMA
  + ENA
- Hypergammaglobulinemia
- Renal insufficiency (+ electrolyte abN)
- UA
  - Microscopic hematuria
  - Proteinuria
Structures Involved in RPF

<table>
<thead>
<tr>
<th>Common</th>
<th>Less Common</th>
</tr>
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<tbody>
<tr>
<td>Abdominal aorta</td>
<td>Kidney</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>Pancreas</td>
</tr>
<tr>
<td>Ureters</td>
<td>Duodenum</td>
</tr>
<tr>
<td>Iliac vessels</td>
<td>Spleen</td>
</tr>
<tr>
<td></td>
<td>Ovaries &amp; fallopian tubes</td>
</tr>
<tr>
<td></td>
<td>Ascending aorta</td>
</tr>
<tr>
<td></td>
<td>Biliary tract</td>
</tr>
<tr>
<td></td>
<td>Celiac axis</td>
</tr>
<tr>
<td></td>
<td>Portal vein</td>
</tr>
<tr>
<td></td>
<td>Spermatic vessels</td>
</tr>
<tr>
<td></td>
<td>Inferior mesenteric artery</td>
</tr>
<tr>
<td></td>
<td>Bladder</td>
</tr>
<tr>
<td></td>
<td>Seminal vesicles</td>
</tr>
<tr>
<td></td>
<td>Spermatic cord</td>
</tr>
<tr>
<td></td>
<td>Scrotum</td>
</tr>
<tr>
<td></td>
<td>Diaphragm</td>
</tr>
<tr>
<td></td>
<td>Pleura</td>
</tr>
<tr>
<td></td>
<td>Right main bronchus</td>
</tr>
<tr>
<td></td>
<td>Pulmonary parenchyma</td>
</tr>
<tr>
<td></td>
<td>Pericardium</td>
</tr>
<tr>
<td></td>
<td>Myocardium</td>
</tr>
<tr>
<td></td>
<td>Coronary artery</td>
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</tbody>
</table>

Imaging

- Modalities
  - US
  - CT
  - MRI
  - IVP
  - Retrograde or antegrade pyelography
  - Renography
  - FDG-PET

- NO modality can reliably distinguish idiopathic from malignant RPF
US

- First-line imaging modality
- Shows hydroureteronephrosis
  - Present in >97.5%
- Occasionally demonstrates fibrotic plaque
  - Poorly defined
  - Echo-poor

Sonography in the diagnosis of retroperitoneal fibrosis.

Sander RS, Duffy J, McLaughlin M, Walsh PC.

The ultrasonic appearance of retroperitoneal fibrosis is characteristic of a smooth-bordered and relatively echo-free mass anterior to the sacral promontory. Sonography can be used to confirm the diagnosis, follow response to therapy and detect hydronephrotic changes in the kidneys.
CT

- Gold-standard
- Contrast contraindicated in renal impairment

CT

- Findings
  - Hydronephrosis (usually bilateral)
  - Smooth tapering of ureter at level of RPF
    - Ureteric filling defect 2˚ invasion rarely seen
  - Retroperitoneal soft tissue mass
    - Isodense with muscle on unenhanced studies
    - Variable contrast enhancement
    - Envelopes great vessels + ureters
  - Evaluation of lymphadenopathy + underlying malignancy
CT

• Idiopathic RPF
  – Medial deviation of proximal-mid ureter
    • Begins at L3-4
    • Non-specific
      → seen in 18% of normal subjects

• Malignant RPF
  – ANTERIOR displacement of aorta
  – LATERAL deviation of ureters
MRI

- Superior soft tissue delineation
  - Fat-saturation
  - Distinguish RPF from encased structures
- Avoids radiation
- NOT contra-indicated in renal impairment

MRI

- Characteristics findings
  - T1
    - Hypointense (muscle)
  - T2
    - ↑ T2 = acute phase
    - ↓ T2 = mature plaque
    - Inhomogenous = malignancy
  - Gadolinium
    - Variable enhancement (↑ in acute phase)
Hydroureter

Aorta

MRI

– Seldom performed now
– Contraindicated in renal impairment
– Classic triad
  1. Proximal hydronephrosis (usually bilateral)
  2. Medial deviation of proximal-mid ureter
  3. Extrinsic compression of proximal-mid ureter

IVP
Retrograde (or Antegrade) Pyelography

- NOT contraindicated in renal impairment
- Similar findings to IVP
- Performed when stent or NT placed in uremic or septic patient
- Delineates level + extent of ureteral involvement
Retrograde Pyelogram

Renography

- Differential renal function
- Useful in surgical planning
PET

- Fluorodeoxyglucose (FDG) PET
- Well established in oncology
- Low specificity
- Useful in assessing metabolic activity + treatment response
- Detects occult neoplastic or infectious processes
- Detects other disease sites (multifocal fibrosclerosis)

Differential

- Retroperitoneal fibromatosis
- Inflammatory myofibroblastic tumour
- Inflammatory malignant fibrous histiocytoma
- Inflammatory fibrosarcoma
**Differential**

- **Retroperitoneal fibromatosis**
  - Uniform fibroblast proliferation
  - Originates from muscle + fascia
  - Infiltrative
  - Recurs
  - Does not metastasize
  - Associated with Gardner’s syndrome (familial adenomatous polyposis)

- **Inflammatory myofibroblastic tumour**
  - Mainly children
  - Huge mass with infiltrative borders
  - Myofibroblast proliferation
  - Recurs
  - Rarely metastasizes
Differential

• Inflammatory malignant fibrous histiocytoma AND fibrosarcoma
  – ↑ Cellularity + vascularity
  – Nuclear atypism
  – Mitoses
  – Atypical cells may be rare

Initial Management

• Principles
  – Prompt diagnosis
  – Relief of symptoms
  – Preservation of renal function
    • >50% present with ureteric obstruction + uremia
    • Some progress to ESRD
Initial Management

- Uremia
  - Decompression of hydronephrosis
    - Percutaneous NT
    - Ureteric stenting
      - Typically NOT difficult
  - Watch for POD
Management

• Discontinue precipitant medications
  – Case reports of spontaneous resolution

• Search for occult malignancy
  – Appropriate imaging
  – Percutaneous biopsy

Percutaneous Biopsy

• Indications
  – Prior to primary medical management (disputed)
  – Prior to primary surgical management if
    • Atypical features or location on MRI or CT
    • Lymphadenopathy on MRI or CT
    • History of prior malignancy
Percutaneous Biopsy

- **Technique**
  - CT Guidance
  - Tru-Cut needle (preferable to FNA)

- **Problems**
  - Sampling error → ↑ FALSE NEGATIVES

Pathology

- **Gross**
  - Smooth
  - Flat
  - Grayish-white
Pathology

• Micro
  – Non-specific inflammatory process
    • Early stage
      – Collagen bundles
      – Inflammatory infiltrate
        » Lymphocytes, plasma cells, fibroblasts, lipid-laden
          macrophages, and occasionally eosinophils
      – Capillary proliferation with perivascular infiltrate
      – Edema
    • Late stage
      – Acellular + avascular
      – Sheets of hypocellular collagen
      – Scattered calcification
    • Inflamatory process most notable at margins of mass

Pathology

• Micro
  – Diagnostic clues in 2° RPF
    • Tumour islands → Malignancy
    • Monoclonal infiltrate → Lymphoma
    • Granulomas → TB
    • Hemosiderin deposits → Trauma
    • Sclerotic with minimal inflammatory infiltrate → XRT
Pathology

- Inflammatory infiltrate
  - Diffuse pattern
  - Nodular pattern

Pathology

- Perivascular inflammatory aggregate
  - Monocytes
  - Eosinophils (rare)

- Arrowhead = vessels
- Arrow = monocyte
Pathology

- Diffuse mononuclear infiltrate
- Thick collagen bundles

Treatment Goals

1. Stop progression
2. Relieve obstruction of ureters + other structures
3. Suppress acute-phase reaction + systemic manifestations
4. Prevent recurrence + relapse
Medical Management

• Now considered 1st line
  – Morbidity of ureterolysis
  – Efficacy shown in small case series only
  – Accurate image-guided biopsy techniques to exclude malignancy
  – More sensitive imaging for diagnosis + follow-up

• No RCTs comparing medical vs. surgical management

Medical Management

• Options
  – Corticosteroids
  – Immunosuppressants
  – Others
Corticosteroids

• Mechanism
  – ↓ Cytokine production
  – ↓ Inflammation
  – ↓ Collagen synthesis + maturation

• Indications
  – Historical
    • Poor surgical candidate
    • Neo-adjuvant in context of extensive vascular involvement
    • Failed surgery or recurrence
  – Contemporary
    • Initial management
    • Adjuvant therapy in combination with surgery

Corticosteroids

• 80% clinical response rate
  – Based on 140 cases in ++series of idiopathic RPF
  – Clinical response
    • ↓ Pain + constitutional symptoms (within days)
    • ↓ Size
    • ↓ Ureteric or IVC compression
    • ↓ ESR
    • Diuresis

• 10% recurrence rate
• No consensus on drug, dose, or duration
Corticosteroids

- Kadar (JU 2002)
  - Case series (11 patients)
  - Steroid monotherapy
    - Prednisolone 60 mg po on alternate days x 2 months then tapered to 5 mg over next 2 months for 24 months total
  - Follow-up: 63.1 months
  - Clinical response: 9 (82%)
    - Regression of fibrotic plaque
    - Resolution of hydronephrosis
    - Typically seen within 6-20 months
  - Clinical failure: 2 (18%)
    - Reinitiation of steroids in 1 patient
    - Ureterolysis in 1 patient

Corticosteroids

- Predictors of response
  - Systemic manifestations
  - Leukocytosis
  - ↑ ESR
  - +ANA
  - Active inflammation on biopsy
Corticosteroids

- Disadvantages
  - Adverse effects
    - Weight gain
    - Diabetes mellitus
    - Osteoporosis
    - Avascular necrosis
    - Moon facies
    - etc.
    - Contraindicated in RPF 2° infection

- Minimizing adverse effects
  - H2 Blockers
  - Calcium supplementation
  - Lower dose in combination with immunosuppressants

Immunosuppressants

- Case reports only
- Agents studied
  - Azathioprine
    - 2.5 mg/kg po OD x 3-6 mo. then 1.5 mg/kg x 6 mo.
  - Cyclophosphamide
    - 2 mg/kg OD x 3 mo. tapered to 6 mo. total
    - 1000 mg/m2 q1mo. x 6 mo.
    - More side effects
  - Cyclosporine
  - Penicillamine
  - Mycophenolate mofetil
<table>
<thead>
<tr>
<th><strong>Immunosuppressants</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>• Typically used in combination with corticoids</td>
</tr>
<tr>
<td>• Avoid in elderly or uremic patients</td>
</tr>
<tr>
<td>• Multiple toxicities</td>
</tr>
</tbody>
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<table>
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<tr>
<th><strong>Other</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>• Case reports only</td>
</tr>
<tr>
<td>– Tamoxifen (20 mg po OD x 12 mo.)</td>
</tr>
<tr>
<td>• Non-steroidal anti-estrogen</td>
</tr>
<tr>
<td>• Alters TGF-B</td>
</tr>
<tr>
<td>• Effective in desmoid tumours</td>
</tr>
<tr>
<td>• Used as monotherapy or in combination with corticoids</td>
</tr>
<tr>
<td>• Good alternative if corticoids fail</td>
</tr>
<tr>
<td>– Medroxyprogesterone acetate</td>
</tr>
<tr>
<td>– Progesterone</td>
</tr>
</tbody>
</table>
Surgical Management

- **Indications**
  - Failure of medical management

- **Benefits**
  - Deep biopsies possible → exclude malignancy

- **Disadvantages**
  - Does NOT prevent progression or recurrence
  - Recurrence in up to 50%
  - Minimal effect on systemic manifestations

Surgical Options

- Ureterolysis
  - Open
  - Laparoscopic
- Ileal (or appendiceal) interposition
- Boari flap
- Autotransplantation
- Chronic ureteral stenting
- Nephrostomy drainage
- Nephrectomy
Open Ureterolysis

- Gold standard

- Approach
  - Pre-operative stenting
    - Facilitate identification + dissection of ureters
    - Remove 6-8 weeks post-op
  - Midline transperitoneal
  - Medial mobilization of colon
  - Deep biopsies

Open Ureterolysis

- Dissection
  - Bilateral dissection (even with unilateral hydronephrosis)
  - Begin at distal non-dilated ureter
    - Avoid injury to thin dilated proximal segment
  - Right-angle clamp between ureter + fibrotic mass
  - Excise ureter from fibrous bed
  - Avoid ureteral injury
    - Urine leak → recurrence
    - Late ischemic stricturing
  - UU seldom necessary
Open Ureterolysis

• Options for preventing recurrence

1. Lateral displacement
   – Retract laterally to uninvolved retroperitoneal fat
   – Secure peritoneum medial to psoas

2. Intraperitoneal displacement
   – Tresidder 1972
   – Close peritoneum behind ureters
   – Avoid obstruction at hiatus
Open Ureterolysis

- Options for preventing recurrence

3. Intraperitoneal displacement with omental wrap
   - Carini 1982
   - Definitive approach for extensive RPF

Omental Wrap

- Technique
  - Mobilize omentum from TC attachments
  - Divide along midline to gastric attachment
  - Divide short gastric vessels at gastric wall
  - Blood supply from gastroepiploic arteries
  - Tack in place with absorbable suture

- Alternative
  - PTFE vascular graft

- Benefits
  - Protect from extrinsic compression
  - Provide blood supply to prevent ischemia
Open Ureterolysis

- Barbalias and Liatsikos (Int Urol Nephrol 1999)
  - Retrospective review
  - 21 patients with idiopathic RPF
  - Mean age 55 (44-71)
  - Two techniques
    1. Intraperitoneal displacement (9)
    2. Lateral displacement (12)
  - NO difference in radiologic or clinical outcome

Laparoscopic Ureterolysis

- First reported by Kavoussi and Clayman (JU 1992)
Laparoscopic Ureterolysis

• Fugita et al (J Endourol 2002)
  – 13 patients (7 bilateral + 6 unilateral)
  – Preoperative stenting
  – Transperitoneal 4-port approach
  – Intraperitoneal displacement of ureters
  – Conversion rate: 15% (iliac vein injury + dense fibrosis)
  – OR time: 381 min (bilateral); 192 min (unilateral)
  – Hospital stay: 4 days
  – Complication rate: 30%
    – Prolonged ileus
    – Urinary retention
    – Port-site erythema
    – Epididymitis
  – Follow-up: Lack of UT obstruction in 92% at 30 months

Laparoscopic Ureterolysis

• Technically difficult
• Prolonged operative time

• AUA Update Series 2005 Lesson 3
  → “This approach has been PROVEN to be less morbid”
    – Elashry et al (JU 1996)
      » ↓ Analgesic requirements
      » ↓ Hospital stay
      » ↓ Recovery time
Laparoscopic Ureterolysis

- Elashry et al (JU 1996)
  - Case control

### TABLE 4. Patient characteristics

<table>
<thead>
<tr>
<th></th>
<th>Laparoscopic Ureterolysis</th>
<th>Open Ureterolysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. female/male pts.</td>
<td>6/0</td>
<td>2/0</td>
</tr>
<tr>
<td>Period</td>
<td>8/00–9/95</td>
<td>9/95–10/95</td>
</tr>
<tr>
<td>Mean age (range)</td>
<td>36.5 (15–51)</td>
<td>55.7 (31–76)</td>
</tr>
<tr>
<td>Mean cm. stricture length (range)</td>
<td>4.2 (2–5)</td>
<td>3.3 (2–5)</td>
</tr>
<tr>
<td>Mean mins. operative time (range)</td>
<td>225 (120–330)</td>
<td>225 (120–380)</td>
</tr>
<tr>
<td>Mean cc estimated blood loss (range)</td>
<td>140 (100–250)</td>
<td>373 (100–700)</td>
</tr>
<tr>
<td>Mean days to oral intake (range)</td>
<td>1</td>
<td>4.7 (3–6)</td>
</tr>
<tr>
<td>Mean days hospital stay (range)</td>
<td>2.8 (2–6)*</td>
<td>10.5 (6–20)</td>
</tr>
<tr>
<td>Mean mg. parenteral morphine sulfate (range)</td>
<td>7.8 (6–20)*</td>
<td>125 (50–267)</td>
</tr>
<tr>
<td>Mean hrs. to return to usual activities (range)</td>
<td>2.1 (2–3)*</td>
<td>7 (6–8)</td>
</tr>
<tr>
<td>% Complications (No. pts/total):</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Major</td>
<td>None</td>
<td>14 (7/7)</td>
</tr>
<tr>
<td>Minor</td>
<td>None</td>
<td>57 (4/7)</td>
</tr>
</tbody>
</table>

*p < 0.05, Wilcoxon rank sum and Mann-Whitney tests were used to analyse data.

AAA

- AAA → ↑ Incidence of RPF
AAA

• Aneurysmatectomy
  – Ureterolysis remains controversial
  – Without concomitant ureterolysis
    • Some resolve
    • Some persist
    • Some progress

AAA

• Speziale et al (JU 2001)
  – Case series
  – 14 patients with
    – Inflammatory AAA
    – RPF
    – Severe renal impairment
  – Open aneurysmatectomy alone
  – Follow-up 48 months
  – Response rate 75%
    – Disappearance of RPF
    – Resolution of hydronephrosis
  – Mortality rate 7% (1 patient)
  – Stent requirement 7% (1 patient)
AAA

• Endovascular stent graft repair
    → Case report
    → Resolution of hydronephrosis in RPF

Surgical Complications

• Ureteral stricture
  – Ureterotomy
  – Urine leak
  – Ureteral devascularization
• RPF recurrence
• Late intestinal obstruction
  – ↑ with omental wrap
• POD
Follow-up

- Long-term follow-up warranted
  - Recurrence reported up to 16 years

- Monitoring
  - Symptoms
  - Renal function
  - ESR + CRP
  - US
    - Assess ureteral obstruction
  - CT/MRI
    - Assess size of mass

Management Algorithm

![Management Algorithm Diagram]

*Defined as basis of clinical findings or persistent symptoms, normalization of concentrations of acute-phase reactant, CT or MRI evidence of regression of intrapelvic mass, or resolution of obstructive complications.*
Summary

- Obscure and multifaceted disease
- Difficult to diagnose
- Difficult to establish cause
- Medical management now first line
  - No RCTs

Thank you.