Idiopathic Retroperitoneal Fibrosis

Key points:

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Idiopathic Retroperitoneal Fibrosis (RPF) is usually benign, but malignancy must be ruled out by adequate biopsy specimen.</td>
</tr>
<tr>
<td>2.</td>
<td>RPF presents insidiously and non-specifically, therefore is often diagnosed by CT scan as an incidental finding.</td>
</tr>
<tr>
<td>3.</td>
<td>RPF is treatable by a combination of medical and surgical approaches.</td>
</tr>
</tbody>
</table>


Manifestations: Usually presents gradually as:

- Pain over the flank, low back, abdomen, or poorly localized midsection pain in >90% pts
  - Early sign, unchanged with position, progressive
  - Helped by NSAIDS due to inflammatory nature of the fibrosis
- Constitutional symptoms: malaise, anorexia, fever, weight loss, N/V
- Vascular compromise:
  - Venous: IVC involvement → lower extremity edema, phlebitis, DVT
  - Arterial: Claudication, mesenteric ischemia
- Symptoms of renal insufficiency or failure from obstructive uropathy at the ureteral level

Demographics: Age 40-60, Males twice as common than females

- Diagnosis: Primarily made via imaging studies such as US, Urography, CT (exam of choice), MRI and confirmed with biopsy. Biopsy helps confirm presence of fibrosis and rule out malignancy as the inciting cause. Open biopsy is usually preferred, though No RCCT studying the difference between CT guided needle biopsy and open biopsy
  - CT classically appears as a confluent symmetric mass encasing the aorta, vena cava, and ureters.
  - Malignancy (mets and primary) typically distinguishable from the vessels on CT.

Differential diagnosis:
Secondary causes of RPF and other causes of obstructive uropathy:
- Malignancy, connective tissue disease, beta blocker use, methysergide use.

Associated Findings:
- 67% idiopathic, no associated conditions or exposures
- 12.4% methysergide exposure
- 8% malignancy
- 2.5% aortic arteritis
- <2% sclerosing cholangitis, Crohn’s disease, thyroiditis, abdominal aortic aneurysm

Mechanism: inflammatory fibroblast proliferation with extracellular matrix deposition—usually benign.

Treatment:
Medical:
- Glucocorticoids and azathioprine.
  - Pts with s/s of inflammation (high ESR, white count, (+) ANA) benefit most
  - Often used in conjunction with surgery
- Mycophenolate mofetil: case report of MM used in conjuction with steroids resulted in cure.

Surgical: Relief of mechanical obstruction by lysis and removal of fibrosis. Allows for biopsy and adequate exclusion of malignancy. Usually medical therapy is combined with surgical approaches +/- IR stenting of structures being compressed by fibrosis or percutaneous nephrostomy tube placement.