Retroperitoneal fibrosis

Definition
- Inflammatory mass that surrounds and obstructs retroperitoneal structures. It is a rare condition. When the cause is idiopathic, it is referred to as Ormond’s disease.

Incidence
- More common in males.
- Estimated incidence of 1/200,000.
- Average age at diagnosis is 50 years.
- An underlying cancer is found in only 8-10% of cases.
- 70% of cases are idiopathic.

Etiology
- Idiopathic: the most common cause.
- Drugs: Methisergide, Hydralazine, L-dopa methyl, LSD, Reserpine, Haloperidol, β-blockers, Ergotamine, Phenacetin, Amphetamine, ergot alkaloids.
- Chemicals: talc, methyl-methacrylate, microfibrillar collagen hemostat.
- Retroperitoneal tumors: lymphoma (the most common), multiple myeloma, carcinoid, pancreatic cancer, sarcoma.
- Inflammatory processes: sclerosing lymphangitis, pelvic inflammatory disease, ankylosing spondylitis, Wegener’s granulomatosis.
- Bleeding disorders: Henoch-Schönlein purpura, pelvic or abdominal surgery, ruptured viscera.
- Periarteritis: aortic or iliac artery aneurism, inflammatory response to advanced atherosclerosis, collagen disease with vascular involvement, etc.
- Infections: TB, gonorrhea, syphilis, actinomycosis.
- Membranous glomerulonephritis.
- Radiation injury.
- Other causes: endometriosis, sarcoidosis, biliary tract disease.

Pathology
- A whitish, fibrous plaque extending from the renal hilum to the pelvic rim (L4-L5). The plaque surrounds and compresses the aorta, inferior vena cava, and the main branches of both along with the ureter and structures of the gastrointestinal tract.
- There are two phases:
  - Inflammatory: infiltration of inflammatory cells (plasma cells, lymphocytes, macrophages, and eosinophils) resulting from an autoimmune response, probably against a protein (ceroid) loss in the atheromatous plaques of the aorta.
  - Fibrotic maturation: development of fibrotic tissue with limited cellularity.

Symptoms
- Constitutional syndrome: malaise, fatigue, weight loss, anorexia.
- Non-specific symptoms including:
  - Abdominal and back pain.
  - High blood pressure (50% of cases).
  - Lower limb edema.
  - Fever or feverishness.
**Diagnosis**

- **Laboratory results:** elevated ESR (80-90% of cases), normochromic-normocytic anemia, elevated blood *creatinine*, and hypergammaglobulinemia.

- **Imaging tests:**
  - *IVP:* medial deviation with extrinsic compression of the ureter and hydronephrosis (not a specific sign, as it is observed in 18% of healthy subjects).
  - *Ultrasound:* hydronephrosis and paravertebral hypoechoic mass.
  - *Retrograde pyelography:* medial deviation of the ureter and segmentary stenosis with no filling defect.
  - *CT:* well-defined retroperitoneal mass, isodense with respect to muscular tissue. Contrast enhancement is greatest in the early stages.
  - *MRI:* in T1 sequences fibrous plaque shows a decreased or medium signal intensity while in T2 sequences it is variable. The use of *gadolinium* facilitates both the evaluation of disease activity and the monitoring of the response to treatment. A low signal indicates mature plaque. In general, MRI allows for better differentiation of soft tissue, especially at the interface with larger vessels.

**Treatment**

- **Ureteral catheterization** or nephrostomy in cases of ureteral obstruction.
- **Percutaneous biopsy** to rule out malignancy. Some authors regard this as unnecessary in the presence of typical signs of retroperitoneal fibrosis (absence of lymphadenopathy).
- **Drug treatment:** the initial treatment of choice, although no prospective randomized studies have evaluated its effectiveness compared to surgery. The patient should be informed of the risk of side effects of medical treatment, the risk of progression, and the absence of a diagnosis of malignancy if a biopsy has not been performed.
  - *Therapy of choice:* oral Prednisolone (PREDNISOLONE BEACON®) 60 mg/day for two months, with gradual reduction of the dose for the next two months to a basal dose of 5 mg/day, to be maintained for 2 years.
  - *Other therapies:*  
    - *Tamoxifen* (TAMOXIFEN TABLET®) 10-20 mg/day for 1-3 years in combination with the aforementioned corticoid therapy. Several studies have demonstrated its therapeutic benefits stemming from its effect on TGF-β, potentially limiting fibrosis.
    - Immunotherapies such as *Azathioprine* or *Cyclophosphamide*. Less commonly used.
- **Surgical treatment:** bilateral ureterolysis. The ureter should be moved to an intraperitoneal position by closing the posterior peritoneum or by wrapping it with epiplon:
  - *Double J catheter:* recommended, to be removed after 6-8 weeks, and associated with the administration of *oral corticosteroids* to accelerate the resolution process and reduce the likelihood of recurrence.
  - *Success rate:* 66-100%.
  - *Follow-up:* long term due to the possibility of recurrence.