Retroperitoneal Fibrosis (Periaortitis)

Retroperitoneal fibrosis is a rare disorder characterised by the presence of a fibro-inflammatory tissue, which usually surrounds the abdominal aorta and the iliac arteries and extends into the retroperitoneum to envelop neighbouring structures - eg, the ureters.[1]

- Retroperitoneal fibrosis is thought to be an autoimmune response to an insoluble lipid that has leaked through a thinned arterial wall from atheromatous plaques.
- Fibrous tissue covers the retroperitoneal structures such as the aorta, vena cava, ureters and psoas muscle. It may extend from the renal pedicle to below the pelvic brim.
- The centre of the plaque is usually located at the level of the aortic bifurcation. The fibrous tissue may bifurcate and follow the common iliac arteries.

Epidemiology

Retroperitoneal fibrosis is a rare disease. Idiopathic retroperitoneal fibrosis accounts for over 70% of cases. The incidence of the idiopathic form has been reported to be 0.1 per 100,000 person-years with a prevalence of 1.4 per 100,000.[2]

Causes

In approximately 70% of patients, no underlying cause is found (idiopathic retroperitoneal fibrosis). The recently advocated concept and diagnostic criteria of immunoglobulin G4 (IgG4)-related disease has led to widespread recognition of retroperitoneal fibrosis associated with IgG4-related disease.[3, 4]

Identified secondary causes include:

- Drugs: eg, methysergide, beta-blockers, methyldopa, amfetamines, phenacetin, pergolide and cocaine.
- Abdominal aortic aneurysm.
- Trauma to the renal tract.
- Infection.
- Retroperitoneal malignancy.
- Post-irradiation therapy or chemotherapy.

Retroperitoneal fibrosis may also be associated with primary biliary cirrhosis, fibrosing mediastinitis, panhypopituitarism, glomerulonephritis, rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosa, ankylosing spondylitis, hemilaminectomy, hypothyroidism, carcinoid tumour and Hashimoto’s thyroiditis.

Presentation

Symptoms may occur such as low back pain, nonspecific systemic complaints, and lower limb oedema. The diagnosis is usually late when a patient is evaluated for renal insufficiency and obstructive uropathy.[2]

- Most patients present with nonspecific symptoms, including dull abdominal pain, of less than 12 months in duration.
- Patients may present with the complications of retroperitoneal fibrosis (see 'Complications', below).
- Early clinical features depend on any underlying cause.
- Advanced disease causes obstructive uropathy. The patient may present with acute kidney injury or chronic kidney disease resulting from ureteric involvement.
- Reduced blood flow to the lower limbs may cause features of peripheral vascular disease.
- The most common presentation is pain, which may occur in the loin, back, scrotum or the lower abdomen.
- Fever, weight loss, nausea and vomiting, malaise and peripheral oedema may occur.
- Urinary features include polyuria, polydipsia, anorexia, nocturia, oliguria, urinary frequency and haematuria.
- Children may present with hip or gluteal pain.

Differential diagnosis

- Other conditions that can cause ureteric obstruction and either acute kidney injury or chronic kidney disease - eg, retroperitoneal abscess, periaortic haematomata, pelvic surgery, radiation therapy and amyloidosis.
- Similar radiological appearances may be caused by abdominal aortic aneurysm, lymphomas, sarcomas, pancreatic carcinomas and metastatic malignancies.

Investigations

There needs to be a high index of suspicion of a diagnosis of retroperitoneal fibrosis when patients present with an elevated ESR and CRP and renal insufficiency from obstructive uropathy.[2]
CT and MRI help to exclude secondary causes, but biopsy remains the gold standard for diagnosis.\[^{5}\]

- Blood and urine tests: findings may include renal function tests (renal dysfunction), FBC (anaemia, raised white cell count), raised ESR, and urinalysis and urine culture (pyuria).
- Plain X-ray: non-specific but may show evidence of complications - eg, bowel obstruction, pulmonary oedema (acute kidney injury).
- Ultrasound: may help in identifying the retroperitoneal mass; can demonstrate the degree of obstruction to the ureters and kidneys.
- Barium follow-through and enema: bowel obstruction.
- Intravenous urography (IVU): shows dilated ureters with medial deviation of ureters. IVU may lead to contrast nephropathy; therefore, good hydration is essential and IVU should be used with caution in the elderly and those with renal impairment (always check renal function beforehand).
- Retrograde pyelography: for patients with severely impaired renal function.
- Aortography, venography, and lymphangiography help in assessing the level and extent of occlusion.
- CT and MRI scanning: delineation of the extent of the retroperitoneal fibrosis.
- Isotope renography is useful in the serial assessment of renal function.
- Biopsy under CT guidance: differentiate benign masses from malignant retroperitoneal masses; biopsy in retroperitoneal fibrosis shows periaortic inflammation with lymphocyte and plasma cell infiltrate.
- The diagnosis may not be established until surgical exploration.

Management

- In drug-related retroperitoneal fibrosis, stopping the offending drug may result in resolution of urinary tract obstruction and symptoms.
- Medical treatment of retroperitoneal fibrosis depends on the underlying cause. In patients with idiopathic retroperitoneal fibrosis, glucocorticoids are traditionally considered the mainstay of treatment.\[^{2}\]
- Immunosuppressive drugs (eg, azathioprine, cyclophosphamide and tamoxifen) have been used.
- Drainage of the upper urinary tract can be performed as a temporary measure. Percutaneous nephrostomy helps restore renal function, fluid, electrolyte and acid-base balance prior to surgery.
- Surgery may be required to resolve urinary tract obstruction or obstruction of other structures.
- Laparoscopic ureterolysis is very effective for patients with retroperitoneal fibrosis of all causes with morbidity and efficacy comparable to open surgery.\[^{6}\]

Complications\[^{1}\]

- Hypertension is common.
- Fibrosis may cause compression of the major arteries, veins and lymphatics, resulting in thrombophlebitis, arterial insufficiency and lower limb oedema.
- Obstruction of the duodenum and colon may cause bowel obstruction.
- Obstruction of the common bile duct may cause jaundice.
- Spinal involvement may cause neurological abnormalities in the lower limbs.

Prognosis

- Prognosis depends on the degree of renal impairment at presentation and the degree of obstruction of the urinary tract, bowel and blood vessels.
- Idiopathic (non-malignant) retroperitoneal fibrosis has a generally good prognosis unless not appropriately diagnosed or treated, when the disease can cause severe complications - eg, end-stage kidney disease.\[^{1}\]
- Malignant retroperitoneal fibrosis has a poor prognosis. Most patients only live for 3-6 months after receiving a diagnosis of malignant retroperitoneal fibrosis.
- Lifelong follow-up is required for possible progressive or recurrent disease.\[^{7}\]

Further reading & references

- Retroperitoneal Fibrosis; MedlinePlus


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