Learning objectives

Retroperitoneal fibrosis is a rare disease. It has been associated with benign and malignant processes. Diagnosis can be suspected due to the characteristics of the pain or in the assessment of an obstruction of the urinary tract or a venous or arterial failure. Imaging studies are the first diagnostic approach.

Our objectives are to know the characteristics of retroperitoneal fibrosis with different radiological techniques and to define its management.

Background

Introduction

Retroperitoneal fibrosis is a disease characterized by the presence of retroperitoneal tissue, consisting of chronic inflammation and marked fibrosis, which often catches the ureters or other abdominal organs.

The first description of this pathology appeared in 1905, when the French urologist Albarran reported the surgical treatment of a retroperitoneal process causing ureteral obstruction. However, retroperitoneal fibrosis was not established as an entity until Ormond published two cases in 1948.

The most common etiology is idiopathic, and it accounts for two thirds of all cases, but it can also be secondary to drugs, infections, radiotherapy, surgery and, in 8% of the cases, tumors (Table 1).

Epidemiology

The incidence of retroperitoneal fibrosis is 0.1 cases per 100,000 people per year, and its prevalence is 1.4 per 100,000 people.

Men are two to three times more likely to develop retroperitoneal fibrosis than women. There is no ethnic predilection.
The age of occurrence is between 50 and 60 years, but it is not uncommon to find it in children and elderly patients\(^2\).

**Pathogenesis**

The pathogenesis of idiopathic peritoneal fibrosis is unknown.

There are two theories\(^1\):

- Exaggerated local inflammatory reaction to low-density lipoproteins (LDL) and to ceroid (lipoprotein polymer that appears as a result of the oxidation of LDL inside macrophages). When the middle layer of the aorta slims down, the macrophages expose the ceroid to B- or T-lymphocytes, which triggers an inflammatory response that repeats itself, leading to an inflammation of the aortic wall.

- Autoimmune systemic disease. This theory is based on the fact that patients with retroperitoneal fibrosis present constitutional symptoms, increased levels of acute-phase reactants and positive antibodies, and they are associated to autoimmune diseases that involve other organs. Also, they respond to treatment with immunosuppressant agents.

**Pathology**

Macroscopically, the fibrosis is a retroperitoneal mass that surrounds the abdominal aorta and the iliac vessels, and it can also involve the ureters\(^4\). Sometimes it occurs in atypical sites, with periduodenal, peripancreatic, pelvic or periureteral locations, or near the renal hilum, where it looks like a poorly-defined mass\(^1\).

Microscopically, the sclerotic tissue is infiltrated by mononuclear cells but the proportion of these two components varies depending on the stage of the disease. In early stages, the tissue is edematous and highly vascularized, and due to the active chronic inflammation there are many mononuclear cells, including fibroblasts and collagen. Later stages reveal sclerosis and calcifications\(^4\).

The aortic wall also suffers changes that include atherosclerotic degeneration of the intima, slimming of the media and inflammation of the adventitia\(^1\).

**Symptoms**
Clinical manifestations of idiopathic or secondary retroperitoneal fibrosis are unspecific, and they depend on the stage of the disease. The symptoms can be divided into localized, due to the retroperitoneal mass and its compression effect; and systemic\textsuperscript{1}.

With regard to the localized symptoms, the most common one is a constant dull ache that does not increase with movement, located on the lower part of the back, sides or abdomen. There are other symptoms related to the mass compression, such as edema of the lower limbs, deep vein thrombosis, varicocele, hydrocele and constipation. In advances stages with bilateral ureteral obstruction, oliguria and uremia may occur\textsuperscript{1}.

Systemic symptoms are fatigue, fever, nausea, anorexia, weight loss and myalgia\textsuperscript{1}.

**Treatment**

The objectives of the treatment of retroperitoneal fibrosis are to stop the progression of the inflammatory reaction, to inhibit or eliminate the obstruction of the ureters or other retroperitoneal structures, to eliminate systemic manifestations and to prevent recurrences\textsuperscript{1} (Table 2 on page 6).

Corticoids are the basis of the treatment, because they improve the symptoms, they reduce the size of the mass and they resolve the obstruction. However, there are patients refractory to corticoids who receive immunosuppressants or tamoxifen\textsuperscript{1}.

In order to temporarily solve the ureteral obstruction, a percutaneous nephrostomy is performed and stents are placed on the ureters. If the excretory function does not improve, surgery with ureterolysis with intraperitoneal transposition is required, and the ureters are surrounded by omentum\textsuperscript{1}.

**Images for this section:**
### Table 1: Causes of secondary retroperitoneal fibrosis

<table>
<thead>
<tr>
<th>Secondary causes</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Drugs</td>
<td>Methylsergide, Pergolide, Bromocriptine, Ergotamine, Methyldopa, Hydralazine, analgesics, β blockers</td>
</tr>
<tr>
<td>Malignant diseases</td>
<td>Carcinoid, Lymphomas, Sarcomas, Carcinomas of the colon, prostate, breast, stomach</td>
</tr>
<tr>
<td>Infections</td>
<td>Tuberculosis, Histoplasmosis, Actinomycosis</td>
</tr>
<tr>
<td>Radiotherapy</td>
<td>Testicular seminoma, Colon carcinoma, Pancreatic carcinoma</td>
</tr>
<tr>
<td>Surgery</td>
<td>Lymphadenectomy, Colectomy, Hysterectomy, Aortic aneurysmectomy</td>
</tr>
<tr>
<td>Others</td>
<td>Histiocytoses, Erdheim-Chester disease, Amyloidosis, Trauma, Barium enema</td>
</tr>
</tbody>
</table>
Fig. 1: 55-year-old man with abdominal pain. Axial and sagittal reconstruction CT scans show soft tissue mass surrounding the aorta accompanied by increased density of the adjacent fat and extraluminal air bubbles. It is a secondary retroperitoneal fibrosis caused by an abscess.
Table 2: Management of retroperitoneal fibrosis
Imaging findings OR Procedure details

There are no standard criteria for the diagnosis of retroperitoneal fibrosis. The radiological finding is a mass of soft tissue that surrounds the abdominal aorta and the iliac vessels, and that may also reach neighboring structures such as the ureters and the inferior vena cava. This is associated with increased levels of acute-phase reactants in the blood test.

Conventional X-ray

Most of the times, the X-ray does not reveal significant findings, although in advanced stages of the disease, a central mass with the density of a soft tissue can be seen, or also the complications associated to this disease\(^2\).

Excretory urography

A classic triad has been defined, consisting of: delayed renal excretion of contrast with hydronephrosis (Fig. 2 on page 10), medial deviation of the middle third of both ureters and reduction of the ureteral caliber at the level of the L4-L5 vertebral bodies\(^2\).

Ultrasonography

It presents a hypoechoic or anechoic well-defined retroperitoneal mass with irregular edges, as well as hydronephrosis secondary to ureteral obstruction.

CT

The CT makes it possible to establish the location and size of the fibrosis, as well as the involved organs.

Retroperitoneal fibrosis usually appears as a well-defined paraspinal mass (Fig. 3 on page 11) with irregular edges, isodense to muscle. The mass starts around the aorta and the iliac arteries (Fig. 4 on page 11) and then it spreads towards the retroperitoneum and around the ureters.

The intensity of the enhancement of the mass after the administration of contrast depends on the activity of the process. Acute stages reveal an intense enhancement with an increase of 20-60 UH after the administration of contrast, whereas in chronic manifestations of the disease the mass is barely enhanced or nothing\(^2\).
MR

In magnetic resonance imaging the retroperitoneal mass presents low signal intensity in T1-weighted sequences (Fig. 5 on page 12), whereas in T2-weighted sequences, the signal may vary depending on the degree of acute inflammation and the associated edema. The signal is hyperintense in the initial stages and hypointense in the chronic stages (Fig. 4 on page 11). The intensity of the signal in T2-weighted sequences is used to monitor response to treatment, because the edema is reduces, and so is the signal in those sequences. The enhancement after the administration of contrast also depends on the degree of the edema² (Fig. 6 on page 13).

PET

FDG-PET establishes the degree of inflammatory activity. Therefore, it is used to check the effectiveness of the treatment, as well as to assess the existence of persistent residual tissue².

Biopsy

There are no guidelines for the determination of the need of a biopsy to confirm diagnosis and rule out malignancy. Biopsy is not usually performed on patients with typical imaging studies for retroperitoneal fibrosis until surgery is made.

However, some situations in which biopsy is necessary have been defined:

- Atypical location of the mass.
- Clinical, laboratory or radiological findings that suggest malignancy or infection.
- Patients who do not respond to the initial treatment.

The biopsy can be open or it can be a CT-guided puncture, although closed biopsy can provide a false negative result, depending on the site of the puncture¹.

Radiological characteristics that differentiate between benign and malignant lesions

A set of criteria has been defined in order to try to differentiate between benign and malignant retroperitoneal fibrosis, although the specificity of these characteristics is low, which means that there can be no certainty on the nature of the process until a biopsy is performed¹.
• Malignant retroperitoneal fibrosis usually is larger and more extensive. It causes a mass effect and displaces the aorta and the inferior vena cava towards the anterior region and the ureters towards the lateral regions.

• With regard to the distribution of the lesion, lymphomas have a tendency to be located over the renal hilum, while idiopathic retroperitoneal fibrosis is usually caudal to the hilum.

• With regard to its morphological characteristics, retroperitoneal fibrosis is a mass with poorly defined edges and peripheral infiltrate, whereas neoplasms present lobulated and nodular edges.

Images for this section:

Fig. 2: 55-year-old man with idiopathic retroperitoneal fibrosis. The intravenous urography shows delay in the excretion in the right kidney and hydroureteronephrosis in the upper third of the ureter. Axial T2W MR shows a mass surrounding the aorta without increase of intensity, so there is absence of edema. Coronal T1 fat saturation with contrast MR
shows a mass surrounding the aorta from the renal hilium to the iliac bifurcation without enhancement.

**Fig. 3:** Axial CT with coronal and sagittal reconstructions in a 50-year-old man with retroperitoneal fibrosis. The images show a well-marginated soft tissue mass surrounding the aorta.
**Fig. 4:** 8-year-old man with idiopathic retroperitoneal fibrosis. In the first row are the coronal and sagittal CT reconstructions, which showed the soft tissue mass surrounding the iliac bifurcation. In the second row are the MR, the first image is a coronal T1W fat saturation with contrast medium where there isn't enhancement of the mass, the second image is a sagittal T1W where the mass has an intensity signal similar to the muscle, and the third image is an axial T2W where there isn't increase of intensity of the mass.
Fig. 5: Retroperitoneal fibrosis in a 50-year-old man. T1W phase and out of phase MR show a mass surrounding the aorta, with an intensity signal similar to the muscle. In the T1W fat saturation with contrast image the mass has enhancement.
**Fig. 6:** Comparative CT and MR images after contrast medium administration in a 44-year-old man with retroperitoneal fibrosis. There is a soft tissue mass surrounding the aorta and iliac arteries, with enhancement in MR images.
Conclusion

Retroperitoneal fibrosis is a rare entity in which there is an underlying cause in at least 30% of the cases. It develops insidiously, and diagnosis is therefore difficult. Radiological findings are necessary for the initial diagnosis as well as for the suspicion of malignancy, although biopsy is the only way for diagnostic certainty.

References


Personal Information