Ormond's disease or secondary retroperitoneal fibrosis: An overview of retroperitoneal fibrosis

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Learning objectives

This poster illustrates the spectrum of retroperitoneal fibrosis (RF) and describes how to assess its activity. It should give an overview of the epidemiology, aetiology, pathology and imaging findings of retroperitoneal fibrosis and help to differentiate between malignant and benign RF.

Background

Retroperitoneal fibrosis is a rare disease, which is characterized by a chronic non-specific inflammation of the retroperitoneum. The plaque typically begins at the level of the fourth or fifth lumbar vertebra and than spreads cranial towards the renal hila [1]. This retroperitoneal tissue can entrap the ureters or other abdominal organs [2]. In most cases RF is detected during the evaluation of newly diagnosed renal failure or hypertension [3]. The idiopathic form of the disease, called Ormond's disease, accounts for more than two thirds of cases, with the rest being secondary to other factors - e.g. neoplasm, infections, trauma, radiotherapy, surgery, and the use of certain drugs [4].

The epidemiological characteristics of RF are not well established and only data on the idiopathic form are available [4]. Its true incidence is unknown and varies from 1:200.000 to 1:500.000 per year [5, 6]. The peak incidence is seen in patients 40 to 60 years of age and mostly in men with a male to female ratio approximating 3:1 [3, 7]. However there are also reports of affected persons under 18 years of age [8-10].

Both the idiopathic and the secondary RF have an insidious clinical onset, characterized by flank, abdominal or back pain which is often described as dull, constant and not exacerbated by movement or palpation [11]. But if the ureter is involved, the pain can be more colic-like. Constitutional symptoms are malaise, fatigue, headache, nausea, vomiting, anorexia, weight loss and constipation [12]. Scrotal swelling, hydrocele and/or varicocele are frequent and probably result from involvement of the gonadal vessels [4]. Less frequently, urinary symptoms like urinary infections, pollakisuira, polyuria, dysuria, oliguria or anuria are present [3]. Ureteral involvement is reported in 80-100% of cases [13] and the urinary tract obstruction may cause hypertension and secondary acute or chronic renal failure [11], which is the most common and severe complication [4]. Rare symptoms include fever, testicular pain, abdominal angina, intermittent claudication, oedema, gross hematuria and vein thrombosis of the lower limbs [3, 5, 12, 14]. Physical examination is usually inconclusive and may show abdominal or lumbar tenderness. Some patients present a palpable and tender abdominal mass [4]. Because of the non-specific nature of the presented clinical manifestations and the paucity of physical findings, there is often a considerable delay between onset of the symptoms and diagnosis, which leads to the late complications of advanced retroperitoneal fibrosis.
In Ormond’s disease as well as in secondary RF the results of routine laboratory tests are consistent with inflammatory disease: the levels of acute phase reactants such as ESR and CRP are high in 80-100% of patients [4, 15, 16]. Leucocytosis, eosinophilia, proteinuria and hematuria are less common laboratory findings [2]. Azotemia usually depends on the extent of ureteral obstruction [4]. If RF involves the kidney or the biliary tract, high levels of creatinine or bilirubin may result. If the disease leads to intestinal ischemia, lactate increases [11].

Secondary RF is caused by a broad spectrum of factors. It can be associated with the use of drugs like methysergid, bromocriptin, ergotamine, methyldopa hydralazine, analgetics or β-blocker [5, 17-19]. Secondary to malignant diseases, RF may be caused by exuberant desmoplastic response to retroperitoneal tumours like Hodgkin or Non-Hodgkin lymphomas or to retroperitoneal metastases of carcinomas of prostate, breast, stomach or colon [4, 5]. Infections like tuberculosis, histoplasmosis and actinomycosis can also cause RF usually by local spread of the contiguous infection focus [20]. Because of the sclerosing effect of radiation, radiotherapy can induce RF, usually limited to the radiation field [21]. Uncommon causes of secondary retroperitoneal fibrosis include trauma and major abdominal surgery [4].

In up to 15% of patients, RF is associated with other fibrotic processes like Riedel's thyroiditis, fibrous pseudotumour of the orbit, sclerosing cholangitis and mediastinal fibrosis [22, 23]. Their histopathologic characteristic overlap, but the pathogenic mechanisms linking these conditions are still unknown [4].

**Imaging findings OR Procedure details**

Ultrasound is in most cases the first-line examination. On ultrasound, RF appears as a smooth-bordered, hypoechoic or isoechoic mass. By involving the ureter, the mass can cause in one third of patients unilateral or in two thirds of patients bilateral hydronephrosis [24, 25].

The scout films of excretory urograms typically show normal psoas outlines bilaterally, because the mass usually does not extend the lateral borders of the muscle [1]. After application of contrast media, intravenous urography (IVU) usually reveals the triad of medial deviation and extrinsic compression of the ureters in combination with hydronephrosis [1, 4]. However, these aspects can also be caused by primary ureteral tumors, periureteral adenopathy, postoperative strictures of the ureter or inflammatory processes [1, 4] (Fig. 1 on page 28). Furthermore, medial ureteral placement can be seen in up to 20% of healthy individuals [4]. MRU using HASTE (half Fourier-acquired single shot turbo spin echo) sequences can be used to scan the renal tract of patients with RF and offers a safe alternative to IVU without the need for a contrast medium application. According to IVU it shows bilateral hydronephrosis combined with hydroureter and medial deviation of the ureters [25].
CT and MRI are the modalities of choice for evaluating the extent of the disease [24]. Unenhanced CT shows homogenous plaques, which are isodense to muscle [23] and encase the aorta and vena cava inferior without displacement (Fig. 2 on page 30). Usually the mass incorporates the ureters, displacing them medially (Fig. 3 on page 25). The perivascular tissue typically ranges from the origin of the renal arteries up to the bifurcation of the common iliac arteries [3, 12] (Fig. 4 on page 30). Rarely, the fibrotic process is seen as only minimal soft tissue stranding around the great vessels, therefore, thin sections make it easier to visualize [1]. In up to 15% of cases calcifications inside the RF can be identified [11]. After application of contrast-media fibrous tissue enhances to a variable degree, depending on the stage of the disease. In the early, inflammatory stages the plaque is more vascular and enhances contrast media well. But long standing plaques are relatively avascular and only show poor enhancement (Fig. 5 on page 31) [24, 25].

MRI provides a better definition of the fibrous tissue against the surrounding tissue, especially when fat-saturation images are used [4]. The extent of the fibrosis may be demonstrated more sensitively and MRI may show fibrosis in regions which appeared normal on CT [26]. RF is hypointense on T1-weighted images (Fig. 6 on page 32) and of variable intensity on T2-weighted images. At early and active stages of the disease, the tissue may have a high signal according to its oedema and hypercellularity (Fig. 7 on page 26). Mature fibrosis, which is composed predominantly of collagen and few cells, shows low signal intensity on T2-weighted images (Fig. 8 on page 33) [4, 24]. According to CT, retroperitoneal plaques in early and inflammatory stages (particular in untreated disease) show a strong enhancement after administration of gadolinium containing contrast media (Fig. 7 on page 26, 9 on page 24), whereas there is a minimal enhancement in long-standing plaques (Fig. 8 on page 33) [25, 27]. In a study by Burn et al. a significant difference in the dynamic enhancement ratio (defined as the ratio of maximum enhancement to non-enhancement during dynamic sequences) between the patients with acute and those with chronic or treated RF was able to be demonstrated [27]. After treatment, the acute patients showed a reduction in the dynamic enhancement ratio, as well. This may have a role in assessing disease activity, monitoring of the disease progression and resolution following treatment [27]. However, to avoid nephrogenic systemic fibrosis in patients with renal impairment gadolinium-based contrast agents should only be used after careful consideration of the risk factors [28].

FDG-PET is not useful for the diagnosis of RF because of its low specificity, but it can be considered a reliable means of assessing the metabolic activity of the retroperitoneal mass [4]. Whole body imaging by PET can reveal other diseased sites, such as those seen in multifocal fibrosclerosis, and detect occult neoplastic or infectious processes to which RF can be secondary or associated [4]. It may also have a role in differentiating idiopathic disease from that revealed to lymphoma, because fibrous tissue usually shows a low FDG uptake in contrast to lymphoma, which shows a high FDG uptake [25]. But RF can also cause accumulation of FDG, giving a false impression of malignancy [25].
Differential Diagnosis

Ormond's disease and secondary RF may present equal imaging findings (Fig. 10 on page 35, 11 on page 37, 12 on page 38). However, imaging findings such as an anterior displacement of the aorta-iliac vessels or a lateral displacement of the ureters by the mass should heighten suspicion of a secondary form [4]. Also the presence of an inhomogeneous signal in T2-weighted images may suggest RF secondary to a malignant disease [29].

The retroperitoneum can be affected by a number of diseases which may have an appearance similar to RF such as carcinoid, multiple myeloma (Fig. 13 on page 39, 13b on page 40) or pancreas-carcinoma (Fig. 14 on page 41) [11]. RF may also be mimicked by inflammatory tumours such as inflammatory myofibroblastic tumour (inflammatory pseudotumour), which mainly affects children, by inflammatory malignant fibrous histiocytoma and inflammatory fibrosarcoma [4]. Other entities in the differential diagnosis are amyloidosis, retroperitoneal hematomas, retroperitoneal fibromatosis [1] or infectious spondylodiscitis (Tab. 1 on page 43)[30]. When RF is found in other than its typical location, the differential diagnosis is expanded significantly [1].

Echo-planar diffusion weighted MRI with calculated apparent diffusion coefficient (ADC) values may be useful in the differential diagnosis of retroperitoneal solid masses. In a study by Nakayama et al. the ADC values of malignant and benign lesion were significantly different with lower values for lymphoma and carcinoma [31]. However, because of the small number of patients, especially those with RF, further studies are required to ensure the usefulness of diffusion weighted imaging (DWI).

The definitive diagnosis usually has to be established by means of histological evaluation [1, 24], especially when the mass shows atypical localisations or when clinical or laboratory findings suggest the presence of underlying malignant disease or infections [4]. The histological examination is still the most reliable diagnostic tool because it may rule out other malignant, benign, or infectious lesions mimicking idiopathic RF [12]. In malignant RF, however, the metastatic cells are dispersed so diffusely in the fibrotic plaque that multiple deep biopsies throughout the lesion are necessary to establish the diagnosis [1]. While percutaneous biopsy is often diagnostic (Fig. 11b on page 37, 14b on page 42), surgical specimens may be required [24]. Although several authors have stressed the importance of laparoscopic or open biopsies to rule out malignancy, both methods are potentially fallible in that malignant areas may be missed [3]. Therefore, a careful search for occult malignancy using the available imaging modalities is essential and percutaneous biopsies should be performed in each case of diagnostic doubt [3].
Images linked within the text of this section:
**Fig.:** After application of contrast media, intravenous urography (IVU) reveals the extrinsic compression of the right ureter in combination with hydronephrosis. This hydronephrosis is caused by lymphoma.

**Fig.** Ormond’s disease: CT shows a mass, which encases the aorta and ranges to the bifurcation of the common iliac vessels. After application of contrast-media the mass only shows poor enhancement (red arrows). Stent in the right ureter (red arrowhead).
Fig.: MRI of idiopathic RF (Ormond’s disease): Coronar T1 weighted sequence. Hypointense lesion, which begins at the level of renal arteries and extends to the iliac bifurcation. The mass surrounds the aorta and reaches the inferior vena cava.
Fig.: MRI of Ormond’s disease after treatment. Axial sequence. Both, in T1w and T2w hypointense lesion (red arrow), which surrounds the aorta and reaches the inferior vena cava without displacement. After administration of a gadolinium-based contrast medium the retroperitoneal fibrosis shows a poor enhancement.
Fig.: MRI of Ormond’s disease. The plaque (red arrows) encases the aorta and vena cava inferior without displacement. On T1-weighted images the mass is hypointense and shows a strong enhancement after administration of gadolinium containing contrast media.

Fig.: MRI of Ormond’s disease. Coronar T1-weighted image. Medial deviation of the ureters (white arrowheads) caused by hypointense retroperitoneal fibrosis. Cirrhosis of the left kidney, ureter stent on the right sight (white arrow).
**Fig.**: MRI of RF in an early and inflammatory stage. On T2-weighted images the tissue has a high signal (red arrows). On T1-weighted images the mass is hypointense and shows a strong enhancement after administration of gadolinium containing contrast media (red arrows).
**Fig.:** Patient with inflammatory aortic aneurysm: CT shows a mass, which encases the aorta and ranges to the bifurcation of the common iliac vessels. The mass has no significant enhancement after application of contrast-media. Note the stent in the left ureter.

**Fig.:** CT shows a mass, encasing the aorta, with no significant enhancement after application of contrast-media. Note the stent in the right ureter. b) Percutaneous biopsy was done. Histological result: Secondary RF caused by breast cancer.
**Fig.**: CT shows a mass, encasing the aorta, with no significant enhancement after application of contrast-media. Note the stent in the right ureter. Percutaneous biopsy was done. Histological result: Secondary RF caused by breast cancer.
**Fig.:** Unenhanced CT shows a lesion which is isodense to muscle and encases the aorta. Hydronephrosis left with thinned renal parenchyma. Histological result after percutaneous biopsy: secondary retroperitoneal fibrosis caused by prostate-carcinoma.
**Fig.**: CT shows a mass, encasing the aorta with no significant enhancement of contrast-media. CT-guided biopsy (Fig. 14b) revealed the diagnosis of secondary retroperitoneal fibrosis caused by pancreas-carcinoma.
Fig.: CT shows a mass, encasing the aorta with no significant enhancement of contrast-media. CT-guided biopsy revealed the diagnosis of secondary retroperitoneal fibrosis caused by pancreas-carcinoma.
## Retroperitoneal masses

| Benign retroperitoneal fibrosis | Idiopathic  
| Secondary to  
| Drugs  
| Aortic aneurysm  
| Retroperitoneal infection  
| Hemorrhage  
| Retroperitoneal radiation therapy  
| Surgery / trauma  
| Inflammation |
|---|---|
| Malignant retroperitoneal fibrosis | Secondary to infiltration of retroperitoneum by malignant cells producing desmoplastic and sclerotic reaction, primary malignancies of:  
| breast  
| stomach  
| colon  
| prostate  
| lung  
| kidney  
| lymphoma |
| Differential diagnosis | Non Hodgkin lymphoma, carcinoid, multiple myeloma, pancreas-carcinoma, sarcoma  
| retroperitoneal fibromatosis  
| inflammatory myofibroblastic tumour (inflammatory pseudotumour)  
| inflammatory malignant fibrous histiocytoma  
| amyloidosis  
| infectious spondylodiscitis |

**Fig.**: Differential diagnosis of retroperitoneal masses
Fig.: CT shows a hypodense mass, which encases the aorta and ranges to the bifurcation of the common iliac vessels. The mass has no significant enhancement after application of contrast-media. Hydronephrosis right. Is it Ormond's disease?
Fig.: Thoracic CT of the same patient as in Fig. 13: The mediastinal and axillary lymph nodes are enlarged. The patient has a secondary RF, caused by multiple myeloma.
**Fig.**: Ormond’s disease: Unenhanced CT shows a homogenous plaque (red arrows), which is isodense to muscle and encases the aorta without displacement. Note the sclerosis of the aorta.
**Fig.:** Ormond’s disease: Unenhanced CT shows a homogenous plaque (red arrows), which is isodense to muscle and ranges to the bifurcation of the common iliac arteries. Note the sclerosis of the iliac arteries.

**Additional images for this section:**
Fig. 1: MRI of Ormond’s disease. The plaque (red arrows) encases the aorta and vena cava inferior without displacement. On T1-weighted images the mass is hypointense and shows a strong enhancement after administration of gadolinium containing contrast media.
Fig. 2: MRI of Ormond’s disease. Coronar T1-weighted image. Medial deviation of the ureters (white arrowheads) caused by hypointense retroperitoneal fibrosis. Cirrhosis of the left kidney, ureter stent on the right sight (white arrow).
Fig. 3: MRI of RF in an early and inflammatory stage. On T2-weighted images the tissue has a high signal (red arrows). On T1-weighted images the mass is hypointense and shows a strong enhancement after administration of gadolinium containing contrast media (red arrows).
**Fig. 4:** After application of contrast media, intravenous urography (IVU) reveals the extrinsic compression of the right ureter in combination with hydronephrosis. This hydronephrosis is caused by lymphoma.

![Image of intravenous urography revealing extrinsic compression and hydronephrosis.]

**Fig. 5:** Ormond’s disease: Unenhanced CT shows a homogenous plaque (red arrows), which is isodense to muscle and encases the aorta without displacement. Note the sclerosis of the aorta.

![Image of CT scan showing Ormond's disease.]

**Fig. 5:** Ormond’s disease: Unenhanced CT shows a homogenous plaque (red arrows), which is isodense to muscle and encases the aorta without displacement. Note the sclerosis of the aorta.
Fig. 6: Ormond’s disease: Unenhanced CT shows a homogenous plaque (red arrows), which is isodense to muscle and ranges to the bifurcation of the common iliac arteries. Note the sclerosis of the iliac arteries.
**Fig. 7:** Ormond’s disease: CT shows a mass, which encases the aorta and ranges to the bifurcation of the common iliac vessels. After application of contrast-media the mass only shows poor enhancement (red arrows). Stent in the right ureter (red arrowhead).
Fig. 8: MRI of idiopathic RF (Ormond’s disease): Coronar T1 weighted sequence. Hypointense lesion, which begins at the level of renal arteries and extends to the iliac bifurcation. The mass surrounds the aorta and reaches the inferior vena cava.
Fig. 9: MRI of Ormond’s disease after treatment. Axial sequence. Both, in T1w and T2w hypointense lesion (red arrow), which surrounds the aorta and reaches the inferior vena cava without displacement. After administration of a gadolinium-based contrast medium the retroperitoneal fibrosis shows a poor enhancement.
**Fig. 10:** Patient with inflammatory aortic aneurysm: CT shows a mass, which encases the aorta and ranges to the bifurcation of the common iliac vessels. The mass has no significant enhancement after application of contrast-media. Note the stent in the left ureter.

![CT images showing a mass encasing the aorta and a stent in the left ureter.](image)

**Fig. 11:** CT shows a mass, encasing the aorta, with no significant enhancement after application of contrast-media. Note the stent in the right ureter. b) Percutaneous biopsy was done. Histological result: Secondary RF caused by breast cancer.
Fig. 12: CT shows a mass, encasing the aorta, with no significant enhancement after application of contrast-media. Note the stent in the right ureter. Percutaneous biopsy was done. Histological result: Secondary RF caused by breast cancer.
Fig. 13: Unenhanced CT shows a lesion which is isodense to muscle and encases the aorta. Hydronephrosis left with thinned renal parenchyma. Histological result after percutaneous biopsy: secondary retroperitoneal fibrosis caused by prostate-carcinoma.
**Fig. 14:** CT shows a hypodense mass, which encases the aorta and ranges to the bifurcation of the common iliac vessels. The mass has no significant enhancement after application of contrast-media. Hydronephrosis right. Is it Ormond’s disease?
**Fig. 15:** Thoracic CT of the same patient as in Fig. 13: The mediastinal and axillary lymph nodes are enlarged. The patient has a secondary RF, caused by multiple myeloma.
**Fig. 16:** CT shows a mass, encasing the aorta with no significant enhancement of contrast-media. CT-guided biopsy (Fig. 14b) revealed the diagnosis of secondary retroperitoneal fibrosis caused by pancreas-carcinoma.
Fig. 17: CT shows a mass, encasing the aorta with no significant enhancement of contrast-media. CT-guided biopsy revealed the diagnosis of secondary retroperitoneal fibrosis caused by pancreas-carcinoma.
## Retroperitoneal masses

| Benign retroperitoneal fibrosis | Secondary to:  
|                               | • Idiopathic  
|                               | • Secondary to  
|                               |   • Drugs  
|                               |   • Aortic aneurysm  
|                               |   • Retroperitoneal infection  
|                               |   • Hemorrhage  
|                               |   • Retroperitoneal radiation therapy  
|                               |   • Surgery / trauma  
|                               |   • inflammation  

| Malignant retroperitoneal fibrosis | Secondary to infiltration of retroperitoneum by malignant cells producing desmoplastic and sclerotic reaction, primary malignancies of:  
|                                   | • breast  
|                                   | • stomach  
|                                   | • colon  
|                                   | • prostate  
|                                   | • lung  
|                                   | • kidney  
|                                   | • lymphoma  

| Differential diagnosis | Non Hodgkin lymphoma, carcinoid, multiple myeloma, pancreas-carcinoma, sarcoma  
|                        | • retroperitoneal fibromatosis  
|                        | • inflammatory myofibroblastic tumour (inflammatory pseudotumour)  
|                        | • inflammatory malignant fibrous histiocytoma  
|                        | • amyloidosis  
|                        | • infectious spondylodiscitis  

**Fig. 18:** Differential diagnosis of retroperitoneal masses
Conclusion

RF is a rare, but complex condition in which the role of the radiologist is essential, both in suggesting the diagnosis as well as in aiding management. Therefore contrast-enhanced CT and especially MRI play an important role. T2-weighted images and dynamic gadolinium enhancement can help in assessing activity, monitoring response to therapy and detecting relapse whereas ADC values may provide useful information in differentiating benign and malignant retroperitoneal masses.

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References


