Chronic periaortitis imaging: What radiologists should know.

Poster No.: C-0812
Congress: ECR 2014
Type: Educational Exhibit
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Keywords: Arteries / Aorta, Cardiovascular system, Vascular, CT, CT-Angiography, Computer Applications-Detection, diagnosis, Computer Applications-3D, Diagnostic procedure, Aneurysms, Inflammation, Acute

DOI: 10.1594/ecr2014/C-0812
Learning objectives

The purpose of our educational exhibit is to:

1. Describe the classification, the main pathophysiology mechanisms and the clinical features of chronic periaortitis (CP)
2. Illustrate a review of the main imaging findings of the disease with different techniques including multidetector computed tomography (MDCT), magnetic resonance imaging (MRI) and 18F-fluorodeoxyglucose-positron emission tomography (18F-FDG PET/CT) imaging.
Background

CP is a rare fibro-inflammatory disease, which comprehends a variety of conditions characterized by common clinical manifestations and similar histopathological characteristics. Imaging studies are essential for the diagnosis and management of the disease, allowing prompt recognition of possible complications.
Findings and procedure details

CP includes a variety of conditions with common clinical manifestation and similar histopathological findings, including adventitial and periadventitial inflammation, medial thinning, and advanced atherosclerosis [1-7]:

1. idiopathic retroperitoneal fibrosis
2. inflammatory abdominal aortic aneurysm
3. perianeurysmal retroperitoneal fibrosis
4. thoracic periaortitis

1. **Idiopathic retroperitoneal fibrosis**: the aorta is not dilated and the surrounding fibro-inflammatory tissue might encase the adjacent structures or not [1-7], Fig. 1 on page 8.

2. **Inflammatory abdominal aortic aneurysms**: the tissue develops around a dilated aorta, but does not cause obstructions of proximal organs [1-8], Fig. 2 on page 8.

3. **Perianeurysmal retroperitoneal fibrosis** involves an inflammatory aneurysm: the tissue surrounding the aneurysm entraps the adjacent organs, for example, at the level of the infra-renal abdominal aorta, fibrosis might cause encasement and extrinsic compression or obstruction of the ureters and of the venous and lymphatic drainages [1-9], Fig. 3 on page 9.

4. **Thoracic periaortitis** [10]: the adventitia is thick with predominantly inflammatory infiltrate and fibrotic change of varied intensity; the inflammatory process extends to the medial layer with severe medial elastic fiber damage. The process may be accompanied by aortic dilatation and atherosclerotic changes Fig. 4 on page 10.

Pathogenesis

The main forms of CP are:

- Idiopathic in 75% of cases, pure or associated with autoimmune systemic diseases (eg, systemic lupus erythematosus, vasculitis, auto-immune pancreatitis) [1-7].

- Secondary in 25% of cases to malignancies, infections, drugs, radiation or surgery.

The pathogenesis of CP is unclear and probably multifactorial.

Three main pathogenetic hypotheses have been proposed:
1) an inflammatory disease secondary to advanced atherosclerosis [5]: CP could be a consequence of a local inflammatory reaction to antigens in the atherosclerotic plaques (oxidized low-density lipoproteins (LDL) and ceroid) that induces a self-perpetuating local inflammatory reaction that eventually leads to an aortic wall inflammation mainly concentrated in the media and adventitia;

2) a manifestation of a systemic disease with autoimmune origin [5]: adventitial inflammation, with involvement of the vasa vasorum, is considered the primum movens with subsequent extension to the retroperitoneum and fibrosing reaction, centripetal extension, thinning of the tunica media, aneurysmal dilatation and development of atherosclerosis.

3) a manifestation of IgG4-related systemic disease [8-14]: the adventitia is the most compromised layer; the inflammatory process can also disrupt the lamellar elastic fibers in the media, which may lead to aneurismal formation.

Regarding the last hypothesis, recent works in the last few years have revealed that for each of the members of the family of CP, a fraction of the cases are due to a recently recognized systemic inflammatory condition known as IgG4-related systemic disease [8-14]. This condition is characterized by elevated circulating levels of IgG4 in the serum (in 60-70% of cases), by the infiltration of organs throughout the body by a lymphoplasmacytic infiltrate rich in IgG4-expressing plasma cells and T-lymphocytes, obliterative phlebitis and by variable degree of fibrosis with a characteristic "storiform" pattern [8-14].

According to a recent review, a significant fraction of thoracic lymphoplasmacytic aortitis cases, about 40% of inflammatory abdominal aortic aneurysms/abdominal periaortitis cases, and a portion of retroperitoneal fibrosis cases are all caused by IgG4-related systemic disease [10].

**Clinical manifestations**

The clinical manifestations of idiopathic and secondary retroperitoneal fibrosis often overlap. The clinical signs and symptoms are nonspecific, of varying duration and thus insidious. Patients usually report two types of manifestations: localized, likely due to the retroperitoneal mass and its mechanic or compressive effects, or systemic, expression of the inflammatory nature of the disease [1-6].

Localized symptoms include side, back and abdominal pain (dull, constant and not exacerbated by movement or palpation, or else colic-like if the ureter is involved), lower extremity oedema (related to extrinsic compression of retroperitoneal lymphatics and veins) and deep vein thrombosis [1-6].
Often the localized symptoms are preceded by or coexist with systemic or constitutional symptoms, which include fatigue, low-grade fever, nausea, anorexia, weight loss, and myalgias [1-7].

Patients with idiopathic retroperitoneal fibrosis often have constitutional symptoms, elevated concentrations of acute phase reagents, positive autoantibodies and associated autoimmune diseases, suggesting that idiopathic retroperitoneal fibrosis is a manifestation of a systemic autoimmune disease, rather than an exaggerated local reaction to atherosclerosis, similarly to systemic large-vessel vasculitis [6].

**Role of Imaging**

Imaging studies are essential in the diagnosis and management of retroperitoneal fibrosis, and can sometimes help to differentiate between idiopathic and secondary disease.

However in some cases histopathologic examination of the retroperitoneal tissue is needed for definitive diagnosis.

The first instance radiologic methods are CT and MRI, that allow to study the thoraco-abdominal aorta, its vessels and adjacent organs involvement, showing the peri-aortic and retroperitoneal fibrosis and the possible impaired patency of the ureters [14-16]. These two diagnostic techniques are frequently integrated with PET (or PET-CT) with 18F-fluorodeoxyglucose (18F-FDG) to study the metabolic activity of the disease [17-19].

**MDCT Imaging**

MDCT play a key role in assessing the extent of disease and in studying the effects on other organs [14-15]. On unenhanced CT, periaortic/retroperitoneal fibrosis usually appears as a homogeneous plaque, isodense with muscle, surrounding the aorta [14-15], Fig. 5 on page 11. At the level of the lower abdominal aorta (Fig. 6 on page 12, Fig. 7 on page 13) and of the thoracic aorta (Fig. 8 on page 14, Fig. 9 on page 15), fibrotic tissue may extend, respectively, downstream to the iliac arteries and upstream to the epiaortic vessels.

In the retroperitoneum, fibrotic process may spread laterally to involve the inferior vena cava, the ureters and the lymphatic drainages, causing varying degrees of obstruction (Fig. 10 on page 16, Fig. 11 on page 17).

The fat plane between the mass and the adjacent muscle may be obliterated. Unlike malignancy, benign mass doesn't have the tendency to displace the aorta anteriorly and does not produce local bone destruction [14-15] (Fig. 6 on page 12).
The administration of contrast medium improves the visibility in the early inflammatory stages of retroperitoneal/periaortic fibrosis, but not in later stages. However, it is not reliable in the evaluation of the metabolic activity [14-15]. Contrast-enhancement is variable and depends on the maturity of the fibrotic process, with early or active stage of the disease showing a greater enhancing pattern due to greater vascularity [14-15] (Fig. 10 on page 16).

**MRI Imaging**

MRI allows the avoidance of nephrotoxic contrast medium and provides a better definition against the surrounding tissues, mainly when fat-saturation images are used. MRI has an excellent soft-tissue contrast and great value in distinguishing benign from malignant fibrosis [16]. Retroperitoneal/periaortic fibrosis is hypointense in T1-weighted images; in T2-weighted images its intensity is high in the early or active stages of disease because of tissue oedema and hypercellularity, and low in the late avascular, acellular fibrotic stages [15-16] (Fig. 12 on page 18, Fig. 13 on page 19, Fig. 14 on page 20). The presence of an inhomogeneous signal on T2-weighted images may suggest malignant retroperitoneal fibrosis, although the diagnosis cannot be certain on the basis of MRI appearances alone. Furthermore, diffusion-weighted imaging (DWI) and dynamic contrast-enhanced imaging may help in differentiating benign from malignant soft-tissue masses (e.g. lymphoma) [16] (Fig. 13 on page 19, Fig. 14 on page 20).

**18F-FDG PET/CT Imaging**

This technique, although it has low specificity, shows high sensitivity for wall inflammation and periaortic tissue (Fig. 15 on page 21). At the same time, being a full body examination, it's able to locate any remote location of disease (fibrosis, multifocal), occult malignancy, infectious or inflammatory processes. This method is also useful in the evaluation of the patient after medical treatment and in case of recurrence.
Fig. 1

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Fig. 3

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3) Thoracic Aortitis

Fig. 4

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Basal axial CT shows atherosclerotic alterations of non enlarged abdominal aorta (A-C), and the periaortic tissue with soft tissue density. It can be easily differentiated from intramural or periaortic ematoma, that is tipically hyperdense compared with the aortic lumen. The periaortic tissue is not clearly noticeable between iliac arteries because of his isodensity, compared to the lumen. (D-E)

Fig. 5

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Late phase contrast-enhanced axial CT, shows intense enhancement of periaortic tissue (A-F). It presents quite smooth margins and doesn’t displace the abdominal aorta anteriorly away from the spine (C): this finding can help differentiate between benign retroperitoneal fibrosis and malignant one (associated with neoplasms or retroperitoneal metastasis).

Fig. 6

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Fig. 7

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Axial MDCT images after contrast media administration from cranial to caudal (A-F) show extensive periaortic fibro-inflammatory crescent tissue at the level of the ascending aorta (C-F, blue arrows), of the origin of the left subclavian artery (A), and of the descending aorta that is aneurysmatic (B-F, yellow arrows). Note the important wall thickening especially at the descending tract, with mural calcification of the ascending aorta (D-F).

Fig. 8

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Fig. 9

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73 y/o male with dorso-lumbar pain, high inflammatory markers and intermittent FUO

Contrast enhanced axial CT in arterial (A-C) and late phase (D-F), shows bilateral perirenal and periaortic fibro-inflammatory tissue that extends from the superior mesenteric artery to the subrenal aortic tract, characterized by late enhancement (D-F). This tissue encases the ureters (F).

Fig. 10

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73 y/o male with dorso-lumbar pain, high inflammatory markers and intermittent FUO.

Axial (A), 3D VR reconstruction (B) and MIP (C) CT images highlight the fibro-inflammatory tissue that encases the ureters. It causes proximal ureteral obstruction and mild dilatation of left pelvis and calyces, in the late phase.

Fig. 11

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Fig. 12

73 y/o male with dorso-lumbar pain, high inflammatory markers and intermittent FUO

Axial T2w MRI images (A-B) show peri-renal and peri-aortic tissue and mild bilateral hydronephrosis, without signs of acute or recent bleeding (C). Note aortic wall thickening (C).
73 y/o male with dorso-lumbar pain, high inflammatory markers and intermittent FUO

Axial DWI (A-B) e T2 fat-sat STIR (C) MRI sequences demonstrate the presence of bilateral perirenal and periaortic oedematous tissue, a sign of active disease.

Fig. 13

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73 y/o male with dorso-lumbar aching, high inflammatory markers and intermittent FUO

Axial (A) and coronal late enhancement MRI sequences reconstructions (B-C) that confirm the perirenal and peri-aortic fibro-inflammatory tissue contrast enhancement.

**Fig. 14**

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Chronic Periaortitis: role of MDCT and $^{18}$F-FDG PET-TC

MDCT axial images after contrast media administration (A-C) demonstrating multifocal fibro-inflammatory tissue surrounding the aorta at the level of the origin of the left subclavian artery (A) and at the level of the medial wall of the ascending aorta (B-C). Note the intense enhancement and uptake of the $^{18}$F-FDG (D-F), correlated with metabolic activity. Instead, the periaortic tissue at the level of the descending aorta is characterized by poor contrast-enhancement and uptake of the radionuclide (white arrows in B-C; blue arrow in F).

**Fig. 15**

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Conclusion

The knowledge of the main radiological manifestations of the fibro-inflammatory periaortitis permits prompt diagnosis and early medical treatment to reduce disease progression. Non-invasive imaging modalities such as MDCT and MRI are reliable methods of diagnosis and follow-up.
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References
