AORTITIS: Causes and imaging characteristics of inflammation of the aorta

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Learning objectives

- Describe the causes and imaging spectrum of inflammatory conditions of the aorta.
- Show many real cases of aortitis where the combination of clinical manifestations and imaging findings was the diagnostic key.

Background

The term aortitis, a subtype of vasculitis, refers to an inflammatory condition that involves the aortic and its branches wall, regardless of its cause.

The etiology of this condition can be infectious or non-infectious. However, we will only review the second type, as they are the most common.

The non-infectious causes of aortitis can be divided in rheumatological diseases and non-rheumatological disorders.

Clinical manifestations and laboratory findings are non-specific. Patients usually show constitutional symptoms such as fever, pain, weight loss, vascular insufficiency and elevated levels of acute phase reactants.

Consequently, aortitis can be overlooked if it is not suspected as part of the initial differential diagnoses and imaging can be critical in the detection and therefore the initiation of appropriate management.

Findings and procedure details

IMAGING TECHNIQUES

There is no single specific examination protocol or algorithm for the assessment of potential vasculitis.
• **CT:**

It's usually the first performed technique because of its high disponibility. Biphasic CT is typically performed, with a non-enhanced CT of the chest (that can be omitted in patients younger than 40 y.o.) and a contrast-enhanced scan of the aorta. Nevertheless, in our experience, the wall thickening and enhancement is frequently better seen in a portal phase. ECG-gated CTA is useful for better imaging of the ascending aorta.

The CTA common findings are wall thickening more than 3 mm, known as the "doble ring" sign (with a poorly enhanced intima and a hyperdense media and adventitia, as in Fig. 1 on page 12), periadventitial soft tissue, wall enhancement, stenosis, dilatation or wall calcification.

• **MRI:**

Because of not using ionizing radiation this is the modality of choice for follow-up (particularly in young patients). In case of impaired renal function, non-contrast MR angiography techniques can be used.

The aortic wall may show oedema (high signal in T2, SPIR), gadolinium enhancement of the wall, stenosis and dilataations (Fig. 2 on page 12). It can be useful in the assesment of treatment response.

• **18-FDG-PET-CT:**

Increased metabolism in the wall of the aorta due to its inflammation may be seen. The uptake of FDG in the aortic wall must be higher than the uptake in the liver (Fig. 3 on page 13).

It can be useful in follow-up and treatment response.

**RHEUMATOLOGICAL DISORDERS (VASCULITIS)**

Vasculitis have been classified with regard to its predilection for vessels of a specific size by the Chapel Hill Consensus Conference (CHCC) and the American College of Radiology (ACR).

The Chapel Hill Consensus Conference of 1994 had classified vasculitis in three major categories: great vessel, medium vessel and small vessel arterities. However, vasculitis of all three major categories can affect arteries of any size.
A more recent CHCC has taken place in 2012, adding four new categories: variable vessel vasculitis, single-organ vasculitis, vasculitis associated with systemic disease and vasculitis associated with probable etiology (Table 1 on page 14).

The aorta and its branches are more frequently affected by large-vessel vasculitis (Tayakasau arteritis and giant cell arteritis).

A) Vasculitis with high prevalence of aortic involvement (>10%):

- **Takayasu arteritis (TA)/Pulseless disease/Martorell Syndrome:**

  It is a necrotizing and obliterative segmental large-vessel and T-cell mediated panarteritis.

  Its cause is unknown; it is thought to be secondary to an autoimmune process. It is mostly seen in young women (third decade), frequently from Asia.

  There are two stages: an early stage (systemic phase) when clinical manifestations are vague and therefore diagnosis is difficult and a late stage (pulseless phase) which is characterized by symptoms secondary to arterial stenosis, occlusion or dilatation.

  Diagnosis is made when three out of the six 1990 ACR criteria are present (Table 2 on page 15).

  Aortic involvement has been described in 30-90% of cases. It mostly affects the abdominal aorta (type IV of TA) (Table 3 on page 16).

  The imaging most common findings are vessel wall concentric thickening more than 3 mm in the early stage, thrombosis, stenosis, occlusion, vessel ectasia, aneurysms and ulcers. Linear aortic wall calcification can be seen after 5 or more years of inflammation and it tends to spare the ascending aorta.

  The treatment is made with high-dose glucocorticoids. Up to 50% of patients relapse, requiring additional immunosuppression. Revascularisation in cases of stenosis is performed when there is secondary vascular organ insufficiency.

**CASE 1:**

18-year-old woman with recurrent episodes of febricula, dyspnea, chest pain and elevated acute phase reactants.
A CECT was performed, showing wall thickening and stenosis of both carotid arteries from their origin to bifurcation (Fig. 4 on page 16). Complete stenosis of both lobar inferior pulmonary arteries and parcial stenosis of right and left superior segmentary arteries (Fig. 5 on page 17 and Fig. 6 on page 18) were detected. Note the increased diameter of the pulmonary trunk, meaning pulmonary hypertension (Fig. 7 on page 19). Treatment with corticosteroids and metotrexate was stablished with no clinical improvement, so stents in the affected pulmonary arteries had to be placed (Fig. 8 on page 20).

CASE 2:

A porcelain thoracic ascendent aorta consequence of long-standing aortitis was found as an incidental finding (Fig. 9 on page 21) in a CT of a woman of 67 years old.

- Giant cell arteritis (GCA):

A granulomatous vasculitis, the most prevalent form of aortitis in Europe, especially among white women of around 80 years old. The age of the patient is the major discriminator between TA and GCA. During the acute phase of the disease, constitutional symptoms are the most common and cranial symptoms, jaw claudication and visual or neurological changes are also frequent. In up to 60% of patients it is closely related to polymyalgia rheumatica, with proximal muscle stiffness and pain in the morning.

According to ACR diagnostic criteria (Table 4 on page 22), no imaging finding is required for diagnosis, but it might be helpful. A negative temporal artery biopsy doesn't exclude the diagnosis of GCA.

Giant cell arteritis affects the aorta in 15-80% of cases. It commonly affects its branches, particularly the superior temporal artery, branch of the external carotid artery ("temporal arteritis"). The other most frequently affected arteries are the subclavian, axillary, femoral, popliteal, tibial and peroneal.

A long segment involvement with significant wall thickening and smooth tapering proximal and distal to the lesion are the most common CT and MR findings. CTA may
show changes similar to TA, like stenosis, occlusion, dilatation, aneurysm formation, calcification and mural thrombi.

Aortic involvement manifests itself more frequently as annuloaortic ectasia or as an ascending aortic aneurysm but it can also appear as acute dissection, aortic valve insufficiency, or abdominal aortic aneurysm. These findings indicate a very high risk of rupture and death, with an average survival of 1 year in patients with thoracic aortic dissection. Ascending aortic aneurysms are usually a late complication.

FDG-PET is only sensitive for extracranial vasculitis whereas US are useful in assessing inflammation of cranial vessels, showing diffuse increased intima-media complex (IMC) thickness, reflecting oedema or increased vascularity.

The standard treatment is high-dose steroids for 1-2 years, with a rapid improvement but a high relapse rate. Unlike TA, additional immunosuppressive therapy does not affect the course of the disease. Revascularisation can also be done. Early treatment is important to avoid complications such as vision loss.

**CASE 3:**

82-year-old woman with anorexia, weight loss, shoulder girdle pain, hyperesthesia of temporal regions, anemia and acute phase reactants elevation.

A CECT was performed, showing diffuse wall thickening of abdominal aorta, both iliac arteries and superior mesenteric artery (Fig. 10 on page 23). The patient had clinical and analytical improvement with corticosteroids.

After 10 days of treatment, a MR was performed, showing diminution of vascular wall thickening but severe stenosis of SMA and left renal artery (Fig. 11 on page 23). The CT-PET was normal. Temporal artery biopsy was positive for GCA.

**CASE 4:**

59-year-old woman with fever of unknown origin during 5 weeks.

CECT and MR showed diffuse thoracic and abdominal aortic wall inflammation and aortic root ectasia (Fig. 12 on page 24 and Fig. 13 on page 24). In PET-CT high uptake of FDG in the wall of aorta, supraaortic trunks and both iliac arteries (Fig. 14 on page 25) had been seen. Corticotherapy was stablished, with clinical improvement.

**CASE 5:**
74-year-old man with temporal artery biopsy positive for GCA and antecedent of right ischemic optic neuritis with vision loss despite corticoid therapy.

He presented an acute reactants elevation and a thoracic CT was performed. A diffuse wall thickening of the aorta was detected, as well as a dilatated ascending aorta and an elongated brachiocephalic trunk (Fig. 15 on page 27). In MR hyperintensity in T2 STIR and gadolinium enhancement of the aortic arch and supraaortic trunks were seen (Fig. 16 on page 27). CT-PET was normal.

- **Ankylosing spondylitis:**

This HLA-B27 spondyloarthropathy was the first rheumatic disease found to be associated with aortitis.

It is usually seen among men during the second or third decade of life. The first clinical manifestation is back pain and stiffness which worsens with inactivity. Constitutional symptoms and acute anterior uveitis are commonly associated.

Aortic involving (60%) more commonly affects the aortic root and the aortic valve causing aortic valvular insufficiency, which is associated with significant morbidity and death.

- **Cogan Syndrome:**

It is an autoimmune disease that causes ocular, inner ear and vascular inflammation. It affects young white adults.

Cardiovascular manifestations include aortitis complicated by aortic insufficiency and necrotizing vasculitis, which may induce coronary, iliac or renal artery stenosis.

- **Relapsing polychondritis:**

This entity, characterized by recurrent episodes of connective tissue inflammation and degeneration, has an autoimmune etiology.

Vascular involvement includes aortic dilatation and regurgitation, mitral regurgitation and aortitis. Aortic wall calcification and ossification as well as aneurysm formation have also been described.

**B) Uncommon but well-documented aortic involvement:**
• **Behçet disease:**

This disease is characterized by the clinical triad of oral ulcers, genital ulcers and uveitis. It is more common in the Mediterranean region (notably in Turkey), among young males and a strong association with HLA-B51 serotype has been detected.

Vascular involvement happens in 25-30% of cases and it can involve various sized arteries and veins of the systemic and pulmonary circulation. The most common vascular manifestations are venous, such as superficial thrombophlebitis or deep vein thrombosis and arterial involvement is rare (less than 5% of patients). The aorta is the most often affected artery, with aortitis, aneurysms or, less commonly, thrombosis. The main case of mortality in Behçet disease is the rupture of a large aortic or arterial aneurysm.

The treatment is based on corticosteroids and cytotoxic agents, as well as on anticoagulant therapy, that should be carefully given in patients with pulmonary arterial aneurysms because of the risk of haemoptysis. Surgical treatment or endovascular procedures with a stent graft may be considered.

• **Rheumatoid arthritis:**

Aortitis is rare and may be associated with rheumatoid vasculitis in other vessels.

• **Systemic Lupus Erythematosus:**

It affects the cardiovascular system mostly as serositis of pericardium. Aortitis is uncommon and it is associated with aortic dissection, thrombus and aneurysm formation. The latter tends to occur at a younger age.

**NON-RHEUMATOLOGICAL DISORDERS:**

• **Erdheim Chester syndrome:**

This syndrome is a rare form of non-Langherhans cell histiocytosis of unknown origin. It is more common among males over 40 years old.

Bone involvement is the most frequent abnormality present radiologically and it is asymptomatic in at least 60% of cases. It consists of symmetric osteosclerosis of the long tubular bones, predominating in the metaphysis and diaphysis of the lower limbs.
The extraosseous manifestations may constitute a potentially life-threatening systemic disorder, affecting the retroperitoneum, the central nervous system, the lungs (interstitial pulmonary diseases) and the cardiovascular system.

Vascular involvement is relatively uncommon, although its frequency is thought to be underestimated because it can be asymptomatic.

CT typically shows a periaortic tissue, extending from the ascending aorta to the iliac junction and creating the appearance of a "coated aorta". It affects the adventitial and periadventitial periaortic space, sparing the wall itself. The extension to aortic branches can occur as well, affecting supraaortic trunks, coronary arteries and abdominal branches.

CASE 6:

56-year-old woman with one year progressive dyspnea, dry cough and constitutional symptoms.

A CECT showed bilateral interstitial diffuse pulmonary pattern and a soft tissue mass surrounding the aortic wall from the origin of supraaortic trunks to the iliac juncton (Fig. 17 on page 28).

Erdheim Chester disease was then suspected, so an x-ray and MR of both tibia were performed. They showed a normal fatty bone marrow replacement by a heterogeneous tissue exhibiting low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig. 18 on page 28).

Bone biopsy confirmed histocitary cells infiltration and treatment with immunosuppresors was stablished, with clinical improvement.

- **Paraneoplastic aortitis:**

Autoimmune manifestations in myelodysplastic syndromes have been described. Its presence was usually associated with very rapid clinical deterioration.

These manifestations include vasculitis, which more commonly affects small vessels; large-vessel vasculitis are rare. In most cases described in the literature, the diagnosis of vasculitis was established simultaneously with that of myelodisplastic syndrome.
In all cases described, patients had prominent systemic inflammatory symptoms (fatigue, malaise, fevers and marked elevations of acute-phase reactants).

Pathogenesis is unknown and in the absence of histopathologic evidence of vasculitis, it is not possible to differentiate true vasculitis from leukemic infiltration of the vessel wall.

In the reported cases, CT showed diffuse thickening of the wall of the aorta including or not its branches.

Patients showed a good response to corticosteroids, which must be monitored because of the risk of infection.

**CASE 7:**

47 year-old male with spontaneus right ankle hematoma, fever, diarrhea and weight loss since last 20 days. Anemia, trombopenia and 2% of blasts in peripheric blood count were detected.

Looking for a neoplastic cause, a CECT was performed, showing wall thickening of the aortic arch and the origin of supraaortic trunks (Fig. 19 on page 29) as well as mural thickening of a long segment of distal ileon (Fig. 20 on page 29). Aspiration of bone marrow was compatible with MDS/MPN type myelomonocytic leukemia type 2 from the WHO classification.

Treatment with corticosteroids and 5-azacytidine was established, with consequent inprovement (Fig. 21 on page 30).

- **Chronic periaortitis/retroperitoneal fibrosis/Ormond disease/retroperitoneal granuloma:**

This disorder consists in a fibrous tissue proliferation in the retroperitoneum.

It is idiopathic in 70% of cases and the other 30% it is associated to inflammatory diseases (primary biliary chirrosis, rheumatoid arthritis, systemic lupus erythematosus...) or subjacent neoplasms.

The chronic aortitis tends to affect men between 20 and 40 years old.
In imagin, it is characterized by a soft tissue mass in the retroperitonem that surrounds the abdominal aorta and can extend itself to adjacent viscera, including the inferior vena cava and the ureters (causing obstructive uropathy).

The retroperitoneal mass is typically isodense to the muscle and it does not displace the aorta and inferior vena cava anteriorly from the spine. Its enhancement in CECT and MR depends on the activity of the fibrosis. In T1 weighted images it is hypointense and its hyperintensity in T2 is also variable.

Its treatment is based in corticosteroids, adding immunosuppresors if necessary and surgery can be performed in case of no response to medical treatment.

**CASE 8:**

67-year-old woman with follicular lymphoma. A CT for work up was performed, showing a retroperitoneal soft-tissue mass surrounding thoracic and infrarrenal abdominal aorta (Fig. 22 on page 30).

- **Esclerosing systemic IgG4-related disease:**

  This disorder is characterized by multiple organ fibrosis, mainly glandular tissues, with elevated serum IgG and IgG4 and autoantibodies.

  The principal symptom is usually autoimmune pancreatitis.

  Aortic involvement has been reported as wall thickening with or without luminal changes (more commonly dilatation than stenosis) and homogeneous enhancement of the wall at the late phase. Periaortitis and retroperitoneal fibrosis have also been described. Steroid therapy has been effective in their treatment.

- **Iatrogenic aortitis:**

  Radiation-induced vasculitis can appear after more than 10 years of high doses of therapeutic radiation.

  Thrombosis, pseudoaneurysm, rupture, stenosis and accelerated wall calcification can be caused. It is normally confined to the irradiated field (i.e. ascending aorta inflammation as a late complication of mediastinal radiation therapy for Hodgkin disease).
Drug-induced aortitis has been described after treatment with different therapeutic agents, such as gemcitabine, propylthiouracil, minocycline, retinoids and leukotriene receptor antagonists.

Images for this section:

**Fig. 1**: "Doble ring" sign. CECT showing an poorly enhanced inner-ring (edematous intima, red arrow) and a hyperenhanced outer-ring (inflamed media and adventitia, green arrow)

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Fig. 2: MR demonstrating mural edema in aortic arch in STIR and gadolinium uptake in THRIVE and in MR perfusion.

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Fig. 3: Coronal 18FDG-PET-CT showing high uptake in descendent thoracic aortic wall. Note that the uptake in the liver is lower.

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<table>
<thead>
<tr>
<th>Type of Vasculitis</th>
<th>Conditions</th>
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<tbody>
<tr>
<td>Large vessel vasculitis</td>
<td>Takayasu arteritis, Giant cell arteritis</td>
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<tr>
<td>Medium vessel vasculitis</td>
<td>Polyarteritis nodosa, Kawasaki disease</td>
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<tr>
<td>Small vessel vasculitis</td>
<td>ANCA-associated Microscopic polyangiitis, Granulomatosis with polyangiitis</td>
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<tr>
<td></td>
<td>(Wegener’s granulomatosis), Eosinophilic granulomatosis with polyangiitis</td>
</tr>
<tr>
<td></td>
<td>(Churg-Strauss syndrome)</td>
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<tr>
<td></td>
<td>Immune-complex Anti-glomerular-basement-membrane disease</td>
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<td></td>
<td>Cryoglobulinemic vasculitis</td>
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<td></td>
<td>IgA vasculitis (Henoch-Schönlein purpura)</td>
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<td></td>
<td>Hypocomplementemic urticarial vasculitis (anti-C1q vasculitis)</td>
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<tr>
<td>Variable vessel vasculitis</td>
<td>Cogan syndrome, Behçet disease</td>
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<tr>
<td>Single-organ vasculitis</td>
<td>Cutaneous leukocytoclastic angiitis, Cutaneous arteritis, Primary central</td>
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<td>nervous system vasculitis, Isolated aortitis</td>
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<tr>
<td>Vasculitis associated with systemic</td>
<td>Lupus vasculitis, Rheumatoid vasculitis, Sarcoïd vasculitis, Others</td>
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<tr>
<td>disease</td>
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<td>Vasculitis associated with probable</td>
<td>Hepatitis C virus-associated cryoglobulinemic vasculitis</td>
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<td>etiology</td>
<td>Hepatitis B virus-associated vasculitis, Syphilis-associated aortitis</td>
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<td></td>
<td>Drug-associated immune-complex vasculitis, Drug-associated ANCA-associated</td>
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<tr>
<td></td>
<td>Cancer-associated vasculitis, Others</td>
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**Table 1:** 2012 CHCC for nomenclature of vasculitis

### 1990 ACR criteria for Takayasu arteritis

| Age of onset younger than 40 years                  
| Intermittent claudication                          
| Diminished brachial artery pulse                   
| Subclavian artery or aortic bruit                  
| Blood pressure difference greater than 10 mm Hg between arms |
| Angiographic/CT/MR abnormalities                    

**Table 2:** If three out of six criteria are present, the sensitivity and specificity for diagnosis are 90% and 98% respectively.

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<table>
<thead>
<tr>
<th>Takayasu arteritis types</th>
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<tbody>
<tr>
<td>I</td>
</tr>
<tr>
<td>Aortic arch branches</td>
</tr>
<tr>
<td>II</td>
</tr>
<tr>
<td>a Ascendent aorta and/or aortic arch +/- branches of the aortic arc</td>
</tr>
<tr>
<td>b Descendent thoracic aorta +/- ascending or aortic arch +/- branches</td>
</tr>
<tr>
<td>III</td>
</tr>
<tr>
<td>Thoracic and abdominal aorta distal to aortic arch +/- major branches</td>
</tr>
<tr>
<td>IV</td>
</tr>
<tr>
<td>Abdominal aorta and/or renal arteries</td>
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<tr>
<td>V</td>
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<tr>
<td>General involvement of all aortic segments</td>
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**Table 3**

Fig. 4: Case 1. Takayasu arteritis. CECT. Long stenosis of both carotid arteries as a consequence of diffuse wall thickening from their origin to the bifurcation.

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Fig. 5: Case 1. Takayasu arteritis. CECT. Stenosis of right superior lobe segmentary artery, occlusion of right inferior lobe artery and stenosis of left superior lobe segmentary artery.

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**Fig. 6:** Case 1. Takayasu arteritis. CTA. Lack of inferior lobes arterial enhancement due to complete stenosis of both inferior lobar arteries.

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**Fig. 7:** Case 1. Takayasu arteritis. CECT. Pulmonary trunk and principal pulmonary arteries enlarged due to pulmonary hypertension.

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Fig. 8: Case 1. Takayasu arteritis. CTA. After stent emplacement.

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**Fig. 9:** Case 2. Porcelain aorta. ECG-gated CTA. Thickening of ascendent aortic wall with periaortic calcification.

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<table>
<thead>
<tr>
<th>1990 ACR criteria for giant cell arteritis</th>
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<tbody>
<tr>
<td>Age of onset older than 50 years</td>
</tr>
<tr>
<td>Recent-onset localized headache</td>
</tr>
<tr>
<td>Temporal artery pulse attenuation or tenderness</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate &gt;50 mm/h</td>
</tr>
<tr>
<td>Arterial biopsy: necrotizing vasculitis</td>
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<tr>
<td>No imaging findings are required</td>
</tr>
</tbody>
</table>

**Table 4:** The presence of three out of five criteria has a sensitivity of 94% and a specificity of 91% for the diagnosis of GCA.

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**Fig. 10:** Case 3. GCA. CTA showing wall thickening of aorta and common iliac arteries. Superior mesenteric artery is stenotic due to the thick wall.

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Fig. 11: Case 3. GCA. MR MIP performed after 10 days showing SMA and left renal artery stenosis.

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Fig. 12: Case 4. GCA. CECT demonstrating diffuse thickening of the thoracic and abdominal aortic wall.

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Fig. 13: Case 4. GCA. MR showing mural edema in aortic arch, abdominal aorta and enhancement of thoracic aortic wall.

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**Fig. 14:** Case 4. GCA. FDG-PET-CT showing high uptake in aortic and supraaortic trunks wall.

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**Fig. 15:** Case 5. GCA. Axial and coronal CECT showing thoracic aortic wall and supraaortic trunks wall thickening, aortic root ectasia and elongated brachiocephalic trunk.

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**Fig. 16:** Case 5. GCA. MR showing mural oedema and gadolinium enhancement of the aortic and supraaortic trunks.

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**Fig. 17:** Case 6. Erdheim Chester disease. Coronal CECT showing diffuse lung septal thickening and aortic wall thickening.

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**Fig. 18:** Case 6. Erdheim Chester disease. X-ray of both knees showing lytic lesions with surrounding sclerosis in the proximal portion of both tibiae. MR showing normal fatty bone marrow replacement by a heterogeneous tissue exhibiting low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

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**Fig. 19:** Case 7. CMML type 2 associated aortitis. CTA showing aortic arch and supraaortic trunks wall thickening.

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Fig. 20: Case 7. CMML type 2 associated aortitis. CECT showing mural thickening of a long segment of pelvic ileon.

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Fig. 21: Case 7. CMML type 2 associated aortitis. CTA after treatment showing resolution of the arterial wall thickening.

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Fig. 22: Case 8. Secondary retroperitoneal fibrosis. CECT showing a soft tissue mass surrounding thoracic and abdominal aorta. Notice that the mass doesn’t displace the aorta anteriorly from the spine.

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Conclusion

Aortitis is an overlooked condition and imaging plays an important role in its diagnosis and follow-up, so radiologists must be familiar with its clinical manifestations and imaging findings.

Personal information

References