Aortic inflammation (aortitis) is a rare pathology, with infectious (fungal pseudoaneurysm, syphilis) and noninfectious etiology (arteritis, idiopathic aortitis, ankylosing spondylitis, among others); it has a difficult clinical diagnosis and a variable prognosis. The use of various imaging methods such as multidetector computed tomography (MDCT), magnetic resonance imaging (MRI), positron emission tomography–computed tomography (PET-CT) and ultrasound (US) facilitate the identification, monitoring and treatment of this entity.

The aim of this paper is to perform a literature review and update on aortitis and its various etiologies, exemplifying with cases seen at our institution.

Keywords
Aortitis, Multidetector computed tomography, Takayasu arteritis, Giant Cell Arteritis, Cardiovascular syphilis

Introduction
The term “aortitis” encompasses inflammatory changes of the aortic wall, irrespective of the infectious or noninfectious etiology.1,2 To complement clinical symptoms (headache, back pain, polymyalgia rheumatica and fever), imaging features and the pattern of aortic involvement help to distinguish the underlying cause and monitor disease activity.

The most widely used imaging modalities are angiography, multidetector computed tomography (MDCT), positron emission tomography-computed tomography (PET-CT), magnetic resonance imaging (MRI) and, to a lesser extent, ultrasound (US).1,3 The aim of this paper is to perform a literature review and update on this disease, providing as examples cases seen at our institution.

Discussion
“Aortitis” is a pathologic term that refers to the presence of inflammatory changes of the aortic wall.4,5 Even if the classification of this condition is controversial, it is useful to classify it as “infectious” or “noninfectious”. The latter is the most common category and includes small-, medium-, or large-vessel vasculitis (Takayasu arteritis, giant cell arteritis, chronic ankylosing spondylitis, Cogan syndrome and relapsing polychondritis).1,4 Large-vessel inflammation, including aortitis, predominates in adults and, if there is clinical suspicion, an emergent evaluation of the aorta with an appropriate imaging technique is required for diagnosis and effective management.6 Multiple imaging modalities can be used for the evaluation of this condition. MDCT is currently the modality of choice because of its high sensitivity and specificity (95% and 100%, respectively) for detecting disease and guiding interventional procedures.1 By means of intravenous contrast, multiplanar resolution, adequate image acquisition times and three-dimensional reconstruction software, we can evaluate changes in the wall and lumen of the vessel, assess the extent of disease and monitor disease activity, as well as guide percutaneous interventional procedures.2 Differently from MDCT, MRI provides better tissue differentiation (being able to demonstrate vessel wall edema on T2-weighted sequences) and it does not require the use of ionizing radiation, being an alternative modality when MDCT and/or the iodinated contrast material are contraindicated. PET-CT shows disease activity by increased 18F-FDG radiotracer uptake in the areas involved, and US is helpful in evaluating the wall of the vessels and measuring blood flow, but it is unable to detect active disease.5
Noninfectious inflammatory diseases

**Takayasu arteritis**

Takayasu arteritis is a chronic inflammatory disease of unknown etiology that affects medium and large vessels, such as the aorta and its branches, and the supra-aortic trunks. It affects predominantly middle-aged women. The abdominal aorta is affected most often, followed by the descending thoracic aorta and, less frequently, the aortic arch. Pathologic examination demonstrated granulomas and inflammation of the arterial wall, with infiltration and proliferation of mononuclear cells in the adventitia, followed by fibrosis and calcification in chronic stages. Findings on MDCT include:

- Concentric thickening of the vessel wall, seen as a "double ring" in the early stage of disease (enhancement of the swollen intima and enhancement of the media and adventitia)
- Thrombosis
- Stenosis
- Occlusion
- In addition, vessel ectasia, aneurysms and ulcers may be seen (Fig. 1).1,7

The classification based on angiographic findings divides the disease into five types with different degrees of involvement of the aorta and its branches, making a special distinction when there is pulmonary and/or coronary artery involvement, and being helpful for surgery planning (Table 1).8

**Table 1:** Classification of Takayasu arteritis (Takayasu Conference 1994).

<table>
<thead>
<tr>
<th>Type</th>
<th>Vessels involved</th>
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<tbody>
<tr>
<td>Type I</td>
<td>Branches of the aortic arch</td>
</tr>
<tr>
<td>Type II (a)</td>
<td>Ascending aorta, aortic arch and its branches</td>
</tr>
<tr>
<td>Type II (b)</td>
<td>Ascending aorta, aortic arch and its branches, thoracic descending aorta</td>
</tr>
<tr>
<td>Type III</td>
<td>Thoracic descending aorta, abdominal aorta and/or renal arteries</td>
</tr>
<tr>
<td>Type IV</td>
<td>Abdominal aorta and/or renal arteries</td>
</tr>
<tr>
<td>Type V</td>
<td>Combined features of types II (b) and IV</td>
</tr>
</tbody>
</table>

Involvement of the coronary arteries is indicated as C+. Involvement of the pulmonary arteries is indicated as P+.
Takayasu arteritis. Coronal (A) and MIP vascular reconstruction (B) MR angiography. PET-CT (C-D). Occlusion of the left common carotid artery at its origin (arrowhead) and absence of signal in the proximal third of the left subclavian artery (arrow) with a filiform lumen in the rest of its course. Irregular uptake in the walls of the ascending aorta, aortic arch and origin of supra-aortic vessels (hollow arrow). The highest metabolic activity is observed at the origin of the carotid and left subclavian arteries (line).

PET-CT allows for more precise anatomic localization of disease activity with increased uptake of 18F-FDG as marker of inflammation (Fig. 2).7,9

Giant-cell arteritis

Giant-cell arteritis or temporal arteritis is a large- and medium-vessels granulomatous vasculitis that affects people of over 50 years of age.10 It affects mainly the external carotid branches, especially the superficial temporal artery, vertebral arteries, coronary arteries and the aorta in 15% of cases.11 Giant-cell arteritis is found in association with polymyalgia rheumatica (60% of cases), with symptoms such as stiffness and pain in the shoulder and pelvic girdles, and signs such as elevated levels of acute phase reactants in laboratory tests. MDCT and MRI show:

• Ectasia
• Ascending aortic aneurysm extending into the aortic arch
• Acute dissection
• Aortic insufficiency
• Abdominal aortic aneurysms
• Circumferential mural thickening12

MDCT allows visualization of mural changes, such as calcifications or thrombosis, while MRI detects vessel wall edema when the disease is active.13

The main differential diagnosis from the clinical viewpoint is intramural hematoma (IMH) of atherosclerotic etiology, with a partial crescent-shaped mural thickening (Fig. 3).1,14

Ankylosing spondylitis

Ankylosing spondylitis affects the aortic root and the aortic valve in 80% of cases. MDCT shows aortic wall thickening, present in about 60% of affected patients, and aortic valve thickening with nodularity and associated insufficiency.1 The cellular inflammation of the aortic root results in a marked fibroblastic reparative response, adventitial thickening, focal destruction of the elastic tissue and intimal proliferation. The surrounding inflammation causes obliterative endarteritis and then root dilatation. These chronic or acute changes of the aortic root and valve lead to aortic insufficiency.15

Idiopathic aortitis

Idiopathic aortitis is asymptomatic and diagnosed by pathologic examination. It affects mainly women and it presents as aortic dilatation without any symptoms of systemic disease. Irregular aortic wall thickening involving ascending aorta and aortic arch may be seen.2
Fig. 3 Giant-cell arteritis. MDCT angiography. Native axial images with no intravenous contrast (b), with intravenous contrast (A-C) and coronal 3D maximum intensity projection reconstruction (D) showing “circumferential” periaortic mural thickening (star in A-D) and spontaneous periaortic crescent-shaped hyperintensity in the ascending and descending aorta (arrow in B-C-D) consistent with intramural hematoma. MR angiography (E) shows visible and diffuse para-aortic hyperintensity on T2-weighted sequence corresponding to the vascular inflammatory process (star). Correlation with surgical specimen (F) evidencing drainage of hematoma (arrow) and aortic replacement with Dacron graft (line).
Idiopathic inflammatory aortic aneurysm

Idiopathic inflammatory aortic aneurysm differs from atherosclerotic aneurysm because of the presence of a thickened aortic wall with dense fibrosis that may entrap adjacent structures, such as the duodenum, the ureter and the inferior vena cava. It affects the infrarenal portion of the abdominal aorta in patients younger than those with atherosclerotic aneurysm. A distinctive feature of idiopathic inflammatory aneurysm is that the thickening of the aortic wall predominantly affects the anterior wall of the vessel (Fig. 4). Main differential diagnoses include retroperitoneal fibrosis, in which, in addition to fibrotic inflammatory reaction, there is aortic dilatation, and mycotic aneurysm because of the similarities in clinical, imaging and laboratory findings (elevated C-reactive protein levels). As regards imaging features, the mycotic aneurysm is mostly saccular, irregular, and surrounding air bubbles may be present, unlike the inflammatory aneurysm, which is typically fusiform (Fig. 5). A distinctive feature of idiopathic inflammatory aneurysm is that the thickening of the aortic wall predominantly affects the anterior wall of the vessel (Fig. 4).1

Main differential diagnoses include retroperitoneal fibrosis, in which, in addition to fibrotic inflammatory reaction, there is aortic dilatation, and mycotic aneurysm because of the similarities in clinical, imaging and laboratory findings (elevated C-reactive protein levels). As regards imaging features, the mycotic aneurysm is mostly saccular, irregular, and surrounding air bubbles may be present, unlike the inflammatory aneurysm, which is typically fusiform (Fig. 5).1

Infectious inflammatory diseases

Infectious processes usually occur in vessels with preexisting disease, such as atherosclerosis, aneurysms, diabetes, etc. The most common pathogens involved are Staphylococcus aureus and Salmonella. Other microorganisms include Treponema pallidum, M Tuberculosis, Listeria, Bacteroides fragilis, Clostridium septicum and Campylobacter jejuni.2

Mycotic pseudoaneurysm

The term “mycotic” refers to aneurysms caused by an infectious process. Mycotic aneurysm is highly infrequent (prevalence of
Mycotic aneurysm is a consequence of an infection that weakens the vessel wall, creating a false lumen or pseudoaneurysm. The most common location is the infrarenal aorta, followed by the descending aorta, and the most common organism implicated is Salmonella. On MDCT scan, mycotic pseudoaneurysm appears as a saccular form aneurysm in 90% of cases, with perianeurysmal gas. Vertebral body destruction, psoas abscess and kidney infarct may also be found (Fig. 6 and 7).

Syphilitic aortitis
Syphilis is a sexually transmitted systemic disease caused by Treponema Pallidum. The tertiary stage of disease manifests 5 to 30 years after the primary infection and it is characterized by neurological and cardiovascular involvement (aortitis, aneurysm, aortic valvulitis with regurgitation and coronary arterial stenosis). Invasion of the aortic adventitia causes obliteratoris of vasa vasorum, resulting in impaired blood supply, weakening of aortic wall and development of aneurysm. Syphilitic aortitis involves the ascending aorta in 60% of cases and the aortic arch in 30%.

Conclusion
Aortic inflammatory disease has various etiologies. The use of multiple imaging modalities enables us to classify them, which provides guidance for an appropriate diagnosis.

Conflicts of interest
The authors declare no conflicts of interest.
Fig. 7 Mycotic pseudoaneurysm and abscess. Coronal (A), axial (B-D) and sagittal (C) MDCT angiography. Paravalvular abscess surrounding the aortic root, with dissecting pseudoaneurysm originating from a solution of continuity in the left ventricle outflow tract (arrow in A-B). Increased density of the adjacent mediastinal fatty tissue (arrowhead in D), and filling defect in the pulmonary valve consistent with vegetations (star in C).

References

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