Aortitis – What does Radiologist need to know to help the Clinician?

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Outline

Background

Imaging techniques

Diseases

- Takayasu arteritis
- Giant cell arteritis
- Behçet disease
- Ankylosing spondylitis
- Inflammatory diseases
- Infectious aortitis

Summary
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Infectious aortitis

Summary
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Imaging is only **ONE** of many techniques

- Gene analysis, family studies
- Clinical assessment
- Biopsy, Histology, Immunochemistry

Imaging

- CT, US, TEE, MRI, PET, PET-CT
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CT technique

 Rather “standard’ protocols used for the aorta

I. Unenhanced Chest – “low” dose (40 mA, 100 kVp)

II. Enhanced Chest

i.v. contrast/saline: 100/30 mL, 4 mL/sec

Trigger at LA (100 HU)

If positive: abdomen and pelvis (50 mL)

100 kVp, dose modulation, (NI ~16)

ECG-gating to be considered

III. Axial, MPR, 3D VR – 3D imaging Lab
CT technique
Modality of choice for followup, particularly in young patients
Assessment of the vessel can be done with:

- Non-contrast MRA techniques
- Oblique sagittal contrast-enhanced 3D MR angiography
Nuclear Imaging technique

• FDG does not accumulate in normal vascular structures
  Any uptake of 18-FDG in the aortic wall is abnormal because of inflammatory or infectious processes
  No specific protocol is needed for assessing vasculitis with PET-CT

• Morphologic assessment: limited - low special resolution
• Benefit when obtained in conjunction with either CT or MRI
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Summary
Takayasu arteritis

- „Pulseless disease”
- Idiopathic vasculitis of the elastic arteries
- Initially described in Japan - found worldwide
- All ethnic groups - mild Asian overrepresentation
- Third decade (i.e., the 20s) of life
- Women 10x > men

- Japanese type:
  thoracic aorta and branches
- Indian type:
  abdominal aorta and renal arteries

Dr. Takayasu
Clinical presentation

Acute:
• “B” symptoms, such as weight loss, fatigue, night sweats, anorexia, and malaise

Chronic:
• Symptoms referable to the organs involved
• Claudication, cerebrovascular insufficiency, carotid artery pain, renal artery involvement
• Aortic aneurysm or stenosis (32%)
1990 American College of Rheumatology criteria

- 1) Age of onset younger than 40 years,
- 2) Intermittent claudication,
- 3) Diminished brachial artery pulse,
- 4) Subclavian artery or aortic bruit,
- 5) Blood pressure ≠ > of greater 10 mm Hg (arms)
- 6) Angiographic (CT, MR) evidence of stenosis

- 3/6 criteria present – Sens 90%, Spec 98%
Takayasu arteritis  Case 1

25-year-old previously healthy woman
3 weeks of malaise, low grade fever, leukocytosis and mild to moderate chest pain
Takayasu arteritis  Case 1

- Aneurysmal dilatation of both ascending and descending thoracic aorta
- Substantial aortic wall thickening of abdominal aorta

Oblique sagittal contrast-enhanced 3D MR angiography
Takayasu arteritis  Case 2

57-year-old ♂, 2 months of fever and weakness

Marked thickening of aortic wall
Complications

- Stenosis (up to 52%)
- Aneurysm (up to 32%)
- Any segment or branch may be involved!
Pulmonary arterial involvement is rare, but possible

Diffuse thickening and enhancement of both aortic and pulmonary artery wall
Takayasu Aortitis Case 5

Complications:

- Diffuse thickening of both aortic and PA wall
- Pulmonary infarct due to PA narrowing
Takayasu Aortitis Case 6

Complications:
Aortic root aneurysm and RCA aneurysm

Case courtesy Dr. Karen Song and Dr. Tony Lin, John Hopkins Medicine
Takayasu Aortitis - Treatment

• **Acute phase** - anti-inflammatory medications

  Inflammation reduction with corticosteroids
  50% of patients relapse during tapering
  require additional immunosuppression

• **Chronic phase** - interventional procedures (dilatation, stent placement) for strictures and stenosis due to fibrosis

  Revascularization: ↓ secondary organ insufficiency
  No randomized trials of percutaneous or surgical intervention
• Exclusion of other pathologies (AAS, raptured aneurysm)

• Presence of complications: aneurysm, stenosis, end-organ hypoperfusion

• Involvement of aortic branches and pulmonary arteries

• Assessment of treatment response/ recurrence
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Summary
Giant cell arteritis

- GCA, “temporal arteritis”
- Elastic vessel vasculitis
- Aorta, secondary and tertiary branches
- Affects patients > 50 years with an incidence peaking > 70
- 3:2 ratio women to men
- Incidence ↑ Scandinavia, ↓ southern Europe
- Genetic predisposition in certain populations?
Clinical presentation

- Constitutional symptoms, such as weight loss, night sweats, malaise, and fever
- Cranial symptoms common (tenderness, headache)
- Jaw claudication
- Visual changes, neurologic symptoms (stroke)
- Aortic stenosis less common than in Takayasu
- Aortic aneurysm more common than in Takayasu
1990 American College of Rheumatology criteria

- 1) Age older than 50 years,
- 2) Recent-onset localized headache,
- 3) Temporal artery pulse attenuation or tenderness,
- 4) Erythrocyte sedimentation rate $> 50$ mm/h,
- 5) Arterial biopsy: necrotizing vasculitis

No imaging
Giant cell arteritis

Imaging

- CTA: Diffuse dilatation and thickening of aorta
- MR: Enhancement of aortic wall
- PET-CT: Diffuse 18-FDG uptake
Complications and therapy

- AoD in the setting of an aneurysm reduces survival to an average of 1.1 years
- Aortic aneurysm rupture or dissection caused two thirds of deaths in a series of patients with GCA
GCA – intramural hematoma
Complications and therapy

- Corticosteroids (1-2 years): standard therapy
- Rapid improvement but ↑ relapse rate
- Revascularization: same as in Takayasu arteritis
GCA – What do we bring to the table

- Exclusion of other pathologies (AAS)
- Diagnosis of complications: IMH, aortic dissection, raptured aneurysm
- Involvement of aortic branches
- Assessment of treatment response/ recurrence
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Summary
Behçet disease

- Eponymous syndrome based on 3 symptoms
- Uveitis - aphthous stomatitis - genital ulcers
- Most common in Turkey
- Incidence of 80 to 370 / 100 000
- Commonly associated with HLA B51 allele
Diagnosis

International Group for Behçet’s disease

- Oral ulceration and 2 of these 3
  - 1) Recurrent genital ulceration,
  - 2) Uveitis or retinal vasculitis,
  - 3) Skin lesions (E. nodosum, pseudofolliculitis)

- Vascular involvement in 1/3 of patients
Vascular involvement

- May also involve veins
- Most commonly superficial thrombophlebitis
- Thrombosis in vena cava, varices, cerebral sinuses
- Small vessel disease $\rightarrow$ nonvascular complaints
- Aneurysm, $\rightarrow$ pseudoaneurysm with rupture
- In multiple sites and in different sites
- Aortic involvement is rather rare
- Any artery or vein, large or small, systemic or pulmonary, may be involved by the vasculitic process
Behçet's Disease - arterial involvement
Behçet's Disease - venous involvement
Behçet disease – PA involvement
Complications and therapy

- Aneurysm rupture: unpredictable and fatal
- Systemic corticosteroids
- Surgical repair, ↑ anastomotic pseudoaneurysms
- Endovascular repair (stent grafts)
- No evidence from randomized trials
Behçet disease – What do we bring to the table

- The main cause of sudden death in Behçet's disease is rupture of a large aortic or arterial aneurysm

- Exclusion of those complication – main goal of imaging

- Presence of complications: arterial/ venous stenosis, end-organ hypoperfusion, collaterals location

- Assessment of treatment response/ recurrence
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Summary
Ankylosing spondylitis

• Group of diseases labeled “spondyloarthropathies”
• Strong association of major histocompatibility complex \( HLA \) \( B-27 \) and the absence of rheumatoid factor
Ankylosing spondylitis

Common features:

- Sacroilitis
- Inflammatory arthritis or enthesitis
- Associations with inflammatory bowel disease or psoriasis
- Aortitis and heart block
Clinical presentation

- Ankylosing spondylitis: most common variant
- Often begins with back pain and stiffness
- Second or third decade of life
- Affects men 2 to 3 times as often as women
- Worsens with inactivity
- Often takes years for the diagnosis to be made

- Constitutional symptoms (malaise or fever)
- Acute anterior uveitis (up to 40% of patients)
Diagnosis requires 4 of the 5 criteria:

- Onset of pain at younger than 40 years,
- Back pain for longer than 3 months,
- Morning stiffness,
- Subtle symptom onset, and
- Improvement with exercise
Ankylosing spondylitis
Complications and therapy

- Aortic root and aortic valve involvement are reported in up to 80% of patients
- Aortic valve may have a nodular appearance
- Aortic valvular regurgitation (50% of patients)
- Treatment of aortic root expansion and aortic valvular abnormalities is the same as for other conditions
Ankylosis Spondylitis – What do we bring to the table

• Pre-surgical planning in cases of aortic root expansion or aortic valvular malfunction

• Assessment of coronary arteries involvement on CCTA in asymptomatic patients
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- Takayasu arteritis
- Giant cell arteritis
- Behçet disease
- Ankylosing spondylitis
- Inflammatory diseases
  - Relapsing polychondritis
  - Reumatoid arthritis
  - Idiopathic isolated aortitis
- Infectious Aortitis

Summary
Idiopathic Isolated Aortitis

- Rare form of aortitis
- Lack of any known systemic disease
- Male:female ratio is 2:3.
- Symptoms: related to aortic inflammation such as back pain, abdominal pain and/or elevated inflammatory markers, and both the thoracic and abdominal aorta can be involved
- Incidental diagnosis - possible
Idiopathic Isolated Aortitis

• Complications: aneurysms in the thoracic aorta or abdominal aorta, or a combination of both
• Dense perianeurysmal fibrosis and a thickened aortic wall
• Acute renal failure caused by ureterine obstruction due to retroperitoneal fibrosis
• New aneurysms can develop in 25% of patients who did not receive anti-inflammatory therapy
Idiopathic Isolated Aortitis

75-year-old male with abdominal pain
Idiopathic Isolated Aortitis – What do we bring to the table

- Diagnosis of exclusion – eliminating other causes of Aortitis
- Assessment of retroperitoneal fibrosis
- Assessment of the entire aorta given the systemic nature of the disease and involvement of both thoracic and abdominal aorta aorta
Inflammatory diseases

- “Mycotic endarteritis” (Osler)
- “Infected aneurysm” or “infectious aortitis”
- (because majority of etiologic agents nonfungal)

Infection
- bacterial
- fungal
- viral
- spirochetal
- tuberculous

Sir William Osler, Bt
Causative agents

- *Staphylococcus aureus* and *Salmonella*
- *Pneumococcus* and *Escherichia coli*
- *Treponema pallidum* and other *Treponema species* (10 to 25 years after the initial spirochetal infection)
- *Candida* and *Aspergillus*
- *Tuberculosis*
- (typically in the setting of impaired immunity)
Spread of disease

- **Contiguous spread** from adjacent thoracic structures (mediastinitis, abscess, infected lymph nodes, infectious pericarditis, empyema, paravertebral abscess)
- **Septic emboli** from underlying bacterial endocarditis
- **Hematogenous dissemination** of bacteria in the setting of sepsis or intravenous drug abuse
- **Risk Factors**: atherosclerosis, vascular grafts or catheters, joint prostheses, immunosupression (chemotherapy, alcoholism, steroids, DM)
Presentation

• Saccular aneurysms are most common, but infected aneurysms can be fusiform and often even pseudoaneurysms.

• Ascending thoracic aorta, aortic arch, and descending thoracic aorta, prosthetic aortic grafts and aortic homografts.

• Infected aneurysms are opposite the great vessels in the aortic arch or opposite the visceral arteries.
54F with Type II diabetes, HTN, GERD, and history of alcohol abuse, with 4 days of low grade fever, epigastric pain and odynophagia

CT Abd/Pelvis Day #1

DDX: Esophagitis or aortitis
Next 2 days: patient maintained low grade fever, but WBC and epigastric pain increased

• ESR (102) & CRP (118) were elevated

• CTA Chest was performed to assess for potential aortic pathology

Abdominal CT  Day #3
Antibiotics initiated but abdominal pain increased
Abdominal CT was repeated

Abdominal CT Day #6
Imaging Findings

Rapid progression suggests infectious etiology

Day 1

Day 7

Blood cultures grew H. Influenza
Concern for impending aortic rapture: patient underwent **endovascular aortic stent** placement, despite suspicion for infectious etiology.
Infectious Aortitis – What do we bring to the table

- Diagnosis
- Source of infection
- Assessment of rapid disease progression
- Assessment of complications: abscess, fistula, pseudoaneurysm, saccular aneurysm
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• Large vessel vasculitis: group of rare diseases
• Imaging seldom used for primary diagnosis
• No specific protocols or technique
• More “general” protocols for aortic examinations
• Imaging is only ONE of the diagnostic methods

• Diseases have specific characteristics
• Tangible for detection at a high level of suspicion and within the appropriate clinical context
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