Aortitis, a Serious Diagnosis with Rare Etiologies

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Disclosures

• Speaking and consulting for Janssen, Novo Nordisk, and Lexicon (not relevant).

• No financial conflict related to this talk.

• Some of the cases were modified for teaching purposes.

Objectives

• Discuss etiologies and differential diagnoses for aortitis.

• Share multiple cases with similar presentations but different etiologies.

• Review challenges making the right diagnosis.

• Discuss our published experience.

• Summary and recommendations.
Aortitis

• **Rare disease with different underlying pathogenesis:**
  - Inflammatory
  - Infectious
  - Malignant and paraneoplastic
  - Idiopathic

• **Diagnosis and differential diagnoses:** very challenging!
  - Similar clinical presentations, imaging and lab findings
  - Different diseases or one disease with a wide spectrum!? 

• **Management:**
  - Based on the underlying etiology
  - Expert multidisciplinary team including vascular medicine!

Sleik et al. Vasc Endovascular Surgery 20017 Oct;51(7):470-479
Inflammatory Aortitis

- Giant cell arteritis (GCA)
- Takayasu arteritis (TA)
- Immunoglobulin G4-related disease (IgG4-RD)
- Periaortitis (previously known as retroperitoneal fibrosis)

- Secondary to other disorders:
  - Other vasculitides (Cogan disease, Behcet’s, Kawasaki, small vessel vasculitis “ANCA associated”)
  - Connective tissue disorders: RA, SLE, seronegative spondyloarthropathies
  - Other autoimmune disorders: Relapsing polychondritis, sarcoidosis
  - Histocytosis: Erdheim-Chester disease
  - Radiation

- Idiopathic inflammatory aortitis

Infectious Aortitis

- Bacterial
  - Staphylococcus
  - Salmonella
  - Mycobacterium
  - Treponema
  - Neisseria
  - Rickettsia

- Viral:
  - Herpes
  - Varicella-zoster
  - Hepatitis B or C

- Fungi
  - Aspergillosis
  - Mucormycosis
Malignancy and Paraneoplastic Aortitis

• **Primary:**
  - Malignant fibrous histiocytoma
  - Epitheloid angiosarcoma
  - Unclassified sarcoma

• **Paraneoplastic**
  - Leukemia
  - Lymphoma

CASE: 1

• **History:** 52 y.o. F. with 5 years history of shoulders and hips pain

• **Presentation:** mid abdominal & lower back pain, fever, chills, and night sweats

• **VS:** normal

• **Exam:** Para-umbilical abdominal tenderness with tender shoulders and hips

• **Lab work:** WBC 11K, ESR: 48 and CRP: 22.7
  - Immunoglobin G subclasses, C3 and 4, ANCA vasculitis panel, and ANA were unremarkable
  - Blood cultures, HIV antibodies, syphilis, and viral hepatitis were also negative
CASE 1: Diagnosis and Management

- **Diagnosis:** Giant cell arteritis

- **Management:** prednisone 40 mg po daily for 4 weeks and then slow taper

- Significant symptom improvement in first 3 weeks

- ESR/CRP normalized in 4 weeks

- Imaging improved significantly 1-3-6 months
CASE 1: Follow-up Image After 6 Months

Giant Cell Arteritis (GCA)

- Granulomatous large vessel vasculitis mostly affecting females ≥ 50
- Large vessel or cranial GCA!

**Diagnosis: consider if ≥ 3 of the followings**
- Age ≥ 50 years
- New headaches
- Abrupt onset of visual disturbances (visual loss)
- Jaw claudication
- Unexplained fever, anemia, or other constitutional symptoms
- High ESR and/or CRP
- A current or prior diagnosis of PMR heightens the potential diagnosis

- Now US is now proposed as the first and main imaging modality (OMERACT US {OGUS} score).

Sleik et al. Vasc Endovascular Surgery 20017 Oct;51(7):470-479
## Large Artery vs. Cranial GCA

- **Younger age:**
  - Mean age 66-68 vs 72-76 years

- **Delay in Diagnosis:**
  - Mean 3.5 to 8 months vs 2 months

- **Lower frequency:**
  - Cranial manifestations: <40% vs >70%
  - Visual manifestations: 4% vs 11-19%

- **Positive temporal artery biopsy** in <50% !!

- **Less likely to meet ACR criteria**
  - 39% vs 95%

  

## Disease Activity

- **Disease activity:** symptoms, inflammatory markers and imaging (US!, CTA, MRA, PET scan)

- **Elevated ESR or CRP:** not always indicate a flare (may also be normal in active disease)!!

- **PET scan can be positive in multiple conditions** *(sensitivity 85% and specificity 41%):*
  - Postop (open and endo)
  - Atherosclerosis
  - Inflammation
  - Infection (much higher SUV!)
  - New ligands !
  - **FDG-PET scan, increase recent interest** *(sensitivity 73% and specificity 97%)*

  Arnaud L et al. Arthritis Rheum 2009 60 p1193
  K-H Lee et al Arthritis Rheum 2012 64 p866
  Alibaz-Oner et al. Mod Rheumatol 2015 25 p752
GCA: Management

• Prednisone 1mg/kg daily (maximum 60 mg) for 2-4 weeks.

• Reduce by 5 mg every 2 weeks to 20 mg/day.

• Then by 2.5 mg every 2 weeks to 10 mg/day if there are no flares of disease activity.

• Then: prednisone taper should be slowed, over the ensuing 6-12 months.
  - Tapering by 1 mg decrements each month once the daily dose is less than 10 mg can be considered. Recurrence!

Steroid Sparing Agents

• Tocilizumab: IL6 inhibitor, first FDA-approved drug for GCA!
  - Decreased rate of first relapse
  - Significant reduction in the cumulative dose of glucocorticoids
  - 162 mg SC qwk for a year
  - When to start and stop?! New data: taper; weekly for 12 and then bi-weekly for another 12 months.
  - How soon should prednisone be tapered? (3-4 months)!

• Methotrexate: at best, is only moderately effective!

• Abatacept: CD4+ inhibitor, borderline effective

• Ustekinumab: IL-12 (Th1) IL-23 (Th17) inhibitor, promising results

• DMARDs: Azathioprine, cyclophosphamide, leflunomide

• TNF inhibitors: not that effective!

Skeik et al. Vasc Endovascular Surgery 20017 Oct;51(7):470-479
Villiger PM et al. Lancet 2016; 387:1921
Skeik et al. Vasc Endovascular Surgery 20017 Oct;51(7):470-479
Giant Cell Aortitis; Clinical Presentation and Outcomes in 40 Patients Consecutively Operated on

- 40 pts with postop F/U 4.2 ± 2.3 years, mean age 66.7 ± 9.1 years
- PMR or temporal arteritis in 22.5%
- All pts had ascending aortic involvement (aneurysm, dissection)
- 4 pts needed emergency repair for dissection
- 85% had an arch replacement; 79.4% a hemiarch and 20.6% a full arch
- 13 patients had elevated ESR and or CRP

- Postoperative Complications:
  - AF 40%, stroke 2.5%, SZ 10% and wound infection 2.5%
  - Freedom from intervention 89.9% (4 cases had re-intervention)
  - 7/13 pts with elevated and none with normal ESR+/CRP had disease progression (increase aortic diameter)
  - The 5-year overall survival rate was 91%


Prognosis and Relapse

- Relapse reported in 34-74% (more common in large vessel form).
- Late relapses of GCA have been described.
- 50% of patients were still on treatment after five years.
- 54%-75% off steroids after 5 years.
- Survivorship is similar to general population.
- Aneurysms are associated with higher mortality.
- Large vessel related complications may occur years after diagnosis.

CASE: 2

• **History:** 48-y.o. F. with tobacco abuse

• **Presentation:** One-month of malaise and progressive bilateral hip and buttock pain with walking, and acute painful and purple left foot

• **Vital signs:** unremarkable except P: 119/min

• **Exam:** left purple and cold foot
  - Non-palpable bilateral popliteal, DP and PT pulses

CASE 2: Laboratory and Imaging Work Up

• CBC and CMP were normal

• ESR: 36 and CRP 2.76

• Lipid profile: Elevated TC, LDL and TG

• **Bilateral ABI:** 0.71 with diffuse monophasic waveform! Inflow disease!

• Immunoglobin G subclasses, C3 and 4, ANCA vasculitis panel, and ANA were unremarkable

• Blood cultures, HIV antibodies, syphilis, and viral hepatitis were also negative
CASE 2: CTA of Abdomen and Pelvis:
CASE 2: Diagnosis and Management

- Type IV Takayasu arteritis
  - Age, gender, severe stenoses and involvement

- Management:
  - Prednisone 60 mg daily
  - ASA and statins
  - Walking program
CASE 2: Complications

- **Presented 3 weeks later**: sudden onset of left leg pain

- **Exam**: cold left foot, non-palpable left DP and PT pulses, and livedo reticularis

- Acute CLI!? immediately started on IV UFH

CASE 2: CTA of Abdomen

Improved Aortic Wall Thickness
CASE 2: CTA of Abdomen

New Thrombosis of the Left CIA

CASE 2: CTA of Abdomen

Improved Aortic Wall Thickness but New Thrombosis of the Left CIA
CASE 2: Management of Complications

• Catheter-directed lytic therapy with aspiration thrombectomy resulting in significant reduction of the clot burden

• **Procedural complication:** right iliac artery dissection managed by bare-metal stenting *(inflamed fragile vessel!)*

• The patient continued to improve on prednisone taper, atorvastatin, aspirin and warfarin

• **2 Months:** Symptoms resolved, normalized ESR/CRP and ABIs

CASE 2: CTA of Abdomen, 2 Months Later

Further Improvement of Aortic Wall Thickening and Bilateral Iliac Artery Flow
CASE 2: CTA of Abdomen, 2 Months Later

Further Improvement of Aortic Wall Thickening and Bilateral Iliac Artery Flow
# Takayasu Arteritis

- Rare, chronic large vessel granulomatous vasculitis
- Primarily affects the aorta and large vessels
- Manifestations usually start < age 40
- Females are 8.5 times more commonly affected
- **Prevalence:** highest in Japan (150 cases/yr), Southeast Asia, India, and Mexico
- **Incidence in U.S. and Europe:** 1-3 new cases per million persons per year

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## Clinical Presentation

- **Early Symptoms:** fever, night sweats, malaise, weight loss, arthralgia, myalgia, and mild anemia.

- **Disease progression:** poor pulses, blood pressure discrepancies, vascular bruits, and hypertension due to renal artery stenosis.

- **Cardiovascular complications:** aneurysms, pulmonary hypertension, aortic rupture, and thrombosis.
Takayasu: Age > 40 years

- **Late-onset TAK** (> 40 years) in 13-43% cases

- **Japanese registry of 1372 patients with TAK**
  - 594 patients (43%) >40 years

- **Differences > 40 years vs < 40 years:**
  - Delay in diagnosis: 45 vs 28 months
  - Dyslipidemia: 50% vs 21%
  - Hypertension: 49-61% vs 14-39%
  - Less carotid involvement: 60% vs 90%
  - More coronary involvement; female only: 11% vs 4%

Watanabe et al. Circulation 2015
Schmidt et al. Mayo Clinic Proc. 2013

Diagnosis

- **Clinical presentation, labs and imaging** (US, CT, MR, or catheter-based angiography) showing stenoses of medium and large vessels

- **Laboratory findings:** Elevated ESR and CRP may reflect inflammation but can be unreliable in identifying disease activity!

- **PET scan** may also be helpful for initial diagnosis and monitoring of disease activity!

Angiographic classification
(Based on the 1994 Tokyo International Conference on Takayasu Arteritis)

According to the vessels involved, the most recently proposed angiographic classification divides TA into six types:

- Type I involves only the branches of the aortic arch.
- Type IIa involves ascending aorta, aortic arch and its branches.
- Type IIb affects ascending aorta, aortic arch and its branches, and thoracic descending aorta.
- Type III involves the descending thoracic aorta, the abdominal aorta and/or the renal arteries. The ascending aorta, the aortic arch and its branches are not affected.
- Type IV involves only the abdominal aorta and/or renal arteries.
- Type V has combined features of Type IIIb and IV.

Additionally, involvement of the coronary and pulmonary arteries should be indicated as C (n) or P (n), respectively. Type V has been documented as the most common type.

Global Prevalence of Takayasu Arteritis by Type Based on Our Literature Review

<table>
<thead>
<tr>
<th>Study</th>
<th>Location</th>
<th>Type I</th>
<th>Type IIa</th>
<th>Type IIIb</th>
<th>Type III</th>
<th>Type IV</th>
<th>Type V</th>
</tr>
</thead>
<tbody>
<tr>
<td>Singh et al. (2015)</td>
<td>India</td>
<td>32.2%</td>
<td>8.1%</td>
<td>1.6%</td>
<td>3.2%</td>
<td>17.7%</td>
<td>37.1%</td>
</tr>
<tr>
<td>Schmidt et al. (2013)</td>
<td>United States</td>
<td>20%</td>
<td>6%</td>
<td>7%</td>
<td>5%</td>
<td>5%</td>
<td>57%</td>
</tr>
<tr>
<td>Sahin et al. (2012)</td>
<td>Turkey</td>
<td>39.4%</td>
<td>6.4%</td>
<td>2.7%</td>
<td>3.9%</td>
<td>4.5%</td>
<td>43%</td>
</tr>
<tr>
<td>Cong et al. (2010)</td>
<td>China</td>
<td>40%</td>
<td>4.8%</td>
<td>1.6%</td>
<td>2.4%</td>
<td>20.8%</td>
<td>30.4%</td>
</tr>
<tr>
<td>Kechauo et al. (2009)</td>
<td>Tunisia</td>
<td>67.7%</td>
<td>0%</td>
<td>10.7%</td>
<td>0%</td>
<td>3.6%</td>
<td>25%</td>
</tr>
<tr>
<td>Petrovic-Rackov et al. (2009)</td>
<td>Serbia</td>
<td>50%</td>
<td>19%</td>
<td>0%</td>
<td>0%</td>
<td>0%</td>
<td>31%</td>
</tr>
<tr>
<td>Park et al. (2005)</td>
<td>South Korea</td>
<td>36.1%</td>
<td>2.8%</td>
<td>4.6%</td>
<td>7.4%</td>
<td>15.8%</td>
<td>33.3%</td>
</tr>
<tr>
<td>Sato et al. (1998)</td>
<td>Brazil</td>
<td>21%</td>
<td>4%</td>
<td>0%</td>
<td>4%</td>
<td>14%</td>
<td>57%</td>
</tr>
<tr>
<td>Suwanwela et al. (1996)</td>
<td>Thailand</td>
<td>0%</td>
<td>0%</td>
<td>11.1%</td>
<td>3.2%</td>
<td>19%</td>
<td>66.7%</td>
</tr>
</tbody>
</table>
Takayasu and Arterial Thrombosis

- **TA promotes a hypercoagulable state**: Significantly increase in PLT and coagulation activity

- Although TA-associated thrombosis has been reported in various inflamed vessels, we were unable to find any case of TA complicated by thrombosis of the iliac artery


Takayasu Management

- **Prednisone**: initial dose 40-60 mg daily

- Slow taper: no faster than 10 mg a week when improve

- **Steroids sparing or resistant patients**:
  - DMARDs (Methotrexate, azathuoprine or leflunomide): effective
  - Anti-TNF agents: etanercept, adalimumab or infliximab are effective
  - **IL-6 inhibitor**: Tocilizumab, based on study cohorts (CRT: low number!)

- **Revascularization**: when disease is inactive!

- **Patency in 9.4 years**:
  - Open: 79%
  - Endo 52%
  - Failure can be reduced by medical therapy

BJD 2014;101:43-50
Takayasu’s Arteritis; Operative Results and Influence of Disease Activity (J Vasc Surg, Jan 2006)

- Minority of patients require operation (42/251, 17%)
- Operation was safe, no significant increased mortality, MI, major stroke or renal failure (3 early graft thrombosis, 2 minor stroke)
- Patients with active disease (31%) are more likely to require revision or develop disease in another site

Prognosis

- **5 years survival**: 80 to 90%
- **15-year survival**: 66 and 96% for patients with and without a major complication

- The long-term prognosis among patients who required surgical revascularization (106 Japanese patients followed for a mean of 19.8 years):
  - Survival to hospital discharge was 89%
  - Overall survival at 20 years was 73.5%.
  - Anastomotic aneurysms developed in 13.8% at various times following surgery.

CASE: 3

- **Presentation:** 51 y.o. M. presented with 2 wks of fever, night sweats and chest pain.
- **Exam:** friction rub, erythematous rash all four extremities

- **Labs:** WBC: 17.1, Hgb: 10.4, ESR: 96, CRP 24.28, and normal troponin

- **CXR:** cardiomegaly
- **EKG:** AV flutter
- **TTE:** LV EF 30%, moderate pericardial effusion, thickened enlarged ascending aorta 6.5 cm

CASE 3: CTA of Chest

Ascending Aortic Wall Thickening with Aneurysm: 6.5 cm
CASE 3: CTA of Chest

Ascending Aortic Wall Thickening with Aneurysm: 6.5 cm

CASE 3: CTA of Abdomen

Significant wall thickening of The Abdominal Aorta
CASE 3: Further Work-up

- ANA positive
- C-ANCA (PR3-ANCA) strongly positive: > 1:160
- RF positive: 33.3 (Normal < 15.9)
- Rest of rheumatology work up was negative including IgG4 level

- Chest CT: diffuse mediastinal lymph node prominence
- PET scan: increased uptake around the ascending and abdominal aorta

- Multiple sets of blood cultures: negative

- Skin biopsy: perivascular neutrophilic infiltrate (nonspecific)

CASE 3: Differential Diagnoses and Treatment

- Differential Diagnoses:
  - Inflammatory Aortitis!
    - GCA?
    - ANCA associated vasculitis?
    - Rheumatoid arthritis related aortitis?

- Management:
  - Failed high dose steroids plus methotrexate and tocilizumab
  - Finally significant symptom, lab and imaging improvement on rituximab!
  - Prednisone dose is down to 20 mg and then had open repair

- Final diagnosis: C-ANCA related vasculitis
CASE 3: CTA after Treatment

Improved Abdominal Aorta Wall Thickening

CASE 3: Final Diagnosis: C-ANCA (PR3-ANCA) Vasculitis

- **ANCA associated vasculitis (AAV): necrotizing small vessel vasculitis**
  - C-ANCA (PR3-ANCA): Granulomatosis with polyangiitis
  - P-ANCA (MPO-ANCA): Microscopic polyangiitis, eosinophilic granulomatosis with polyangiitis, or renal-limited vasculitis

- **ACR Criteria for diagnosis of GPA:**
  - Nasal or oral inflammation
  - Abnormal chest radiograph showing nodules, fixed infiltrates, or cavities
  - Abnormal urinary sediment (microscopic hematuria)
  - Granulomatous inflammation on biopsy of an artery or perivascular area

- **Management:** Glucocorticoids combined with rituximab, or less favorably with cyclophosphamide.

CASE: 4

- **Presentation:** 70 y.o. with hx of HTN and DM presents with left side neck mass, right eye lid swelling, night sweat, significant weight loss, and abdominal pain

- **Physical Exam:** left submandibular tender mass, and periumbilical abdominal tenderness

- **Labs:** WBC: 12000, Hgb 11, ALT 350, AST 250, ALP: 300, Lipase 870, ESR: 70, CRP 1.5

- **CXR:** mediastinal lymphadenopathy

- **Abdominal CT:** pancreatitis with sausage-shaped pancreas!

- **Rest of autoimmune w/u and cultures:** unremarkable
CASE 4: Submandibular Gland Biopsy

IgG4 Related Disease

- Immune-mediated condition with multiple disorders sharing particular pathologic, serologic, and clinical features

- **Shared features:**
  - Tumor-like swelling of involved organs
  - Lymphoplasmacytic infiltrate with IgG4-positive plasma cells
  - Variable degree of fibrosis “storiform” pattern
  - Elevated serum IgG4 in 60 to 70 % of patients

- **Major Presentations:**
  - **Salivary gland disease**
  - **Orbital disease:** lacrimal glad inflammation, proptosis
  - **Retroperitoneal disease:** periaortitis, pancreatitis, cholangitis, hydronephrosis
  - **Others:** thyroid (Riedel’s thyroiditis), renal (nephritis), lungs and CNS
## IgG4 Related Disease Diagnosis and Treatment

### Diagnosis
- **Clinical**: classic organ involvement
- **Serology**: elevated serum IgG4, depressed C3/C4
- **Radiology**: periaortitis, sausage-shaped pancreas
- **Pathology**: classic pathology and positive stain for IgG4

### Treatment
- Prednisone: initial dose 40 mg po daily
- Rituximab: as a sparing agent or in resistant cases

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### CASE: 5

- **Presentation**: 55 y.o. M. with hx of HTN presents with abdominal and left flank pain that radiates to the groin area
- **Physical exam**:
  - Periumbilical and left flank moderate tenderness
- **Labs and Images**:
  - WBC 11.6, Hgb: 10.5
  - Creatinine 3.2
  - ESR: 80, CRP 4.6
  - **Abdominal ultrasound**: left side hydronephrosis
- Rest of autoimmune work up as well as blood cultures are negative
Periaortitis (Retroperitoneal Fibrosis)

- Rare condition with presence of inflammatory and fibrous retroperitoneal tissue that often encases the ureters or abdominal organs (hydronephrosis!)

- Idiopathic or secondary to other causes (IgG4 disease, autoimmune disorders, lymphoma, infection, radiation, meds <Ergots>, surgeries ..)

- **Diagnosis:** clinical, labs, biopsy (fibrous tissue with inflammation)

- **Treatment:** Prednisone, methotrexate, tocilizumab, rituximab
CASE: 6

- **Presentation:** 51 y.o F. presents with chronic legs and periumbilical abdominal pain

- **Labs:**
  - Normal CBC
  - ESR 32, CRP 1.5

- **Leg X ray:**
  - Bilateral and symmetric osteosclerosis of the diaphysis of the long bones

CASE 6: CTA of Abdomen

[Image of CTA of Abdomen]
Erdheim-Chester Disease

- Multisystem non-Langerhans cell histiocytosis
- Periadventitial infiltrate can mimic: IgG4-RD, TAK, GCA
- BRAF V600E in approximately half of cases

**Work Up:**
- Biopsy: non-Langerhans histiocytosis
- BRAF V600E
- Some cases with elevated ESR and CRP
- Imaging: Hairy kidney/Coated aorta

**Management:**
- BRAF V600E (+): Vemurafenib, Dabrafenib
- BRAF V600E (-): Steroids, IFN-α, anakinra, tocilizumab, infliximab

**Frequency of clinical or radiologic characteristics in Erdheim - Chester**

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Osteosclerosis</td>
<td>96%</td>
</tr>
<tr>
<td>Bone Pain</td>
<td>40-50%</td>
</tr>
<tr>
<td>CNS Involvement</td>
<td>15-51%</td>
</tr>
<tr>
<td>Diabetes Insipidus</td>
<td>25-27%</td>
</tr>
<tr>
<td>Exophthalmos</td>
<td>24-32%</td>
</tr>
<tr>
<td>Xanthelasmas</td>
<td>19-28%</td>
</tr>
<tr>
<td>Cardiac Involvement</td>
<td>42-52%</td>
</tr>
<tr>
<td>Pulmonary Involvement</td>
<td>22-43%</td>
</tr>
<tr>
<td>Periaortic Infiltration</td>
<td>60-66%</td>
</tr>
<tr>
<td>“Coated Aorta”</td>
<td>23-30%</td>
</tr>
<tr>
<td>“Hairy Kidney”</td>
<td>68%</td>
</tr>
</tbody>
</table>

Haroche et al. Curr Opin Rheumatol 2012

**CASE: 7**

**Presentation:** 59 y.o man with hx of obesity and HTN, presented with abdominal pain

**Exam:**
- Periumbilical moderate tenderness

**Labs:**
- ESR 34 and CRP 2.72
- CBC and CMP are unremarkable

**Extensive unremarkable rheumatological work up

**Negative multiple blood cultures**
CASE 7: CTA of Abdomen and Pelvis

Idiopathic Inflammatory Aortitis

• Treated successfully with taper down steroids and methotrexate

• **Diagnosis:** clinical presentation, labs (ESR and CRP elevation), and imaging findings when all other etiologies are ruled out

• **Management:** steroids, methotrexate, tocilizumab!
CASE 7: CTA of Abdomen 3 Months after Initial Treatment

CASE 8

- **Presentation:** 79 y.o. M with RA, DM, HTN, HL presented with chronic abdominal and lower back pain
- **Medications:** infliximab and methotrexate
- **Exam:** periumbilical abdominal and lower back tenderness

- **CBC:** WBC: 13.1, ESR 64, CRP 3.85
- **Blood cultures and lumbar MRI negative for infection**
- Urine culture was positive for MSSA

- **I.D. Recommendation:** ceftriaxone then cefalexin.

- **Management:** prednisone taper and continue on other immunosuppressive meds
CASE 8: Management and Follow up

- **1 month clinic F/U:** improved symptoms but ESR and CRP were still elevated
- **Blood cultures** were still negative!
- **CTA:** ulcerated ASO plaque!?  

- **Plan:** add statins, risk factor reduction, and bring back in 2-4 weeks for follow up

CASE 8: CTA 4 Days Later

Blood cultures came back positive for MSSA
Infectious Aortitis

- Rare and very hard to differentiate from inflammatory:
  - Leukocytosis (left shift)
  - Procalcitonin elevation
  - Hx of recent infection
  - Positive blood or tissue culture (50-75%)
  - Worsening symptoms on immunosuppression
  - Asymmetric aortic wall thickening or periaortic soft tissue
  - Very high SUV uptake with PET scan


Diagnosis, Management, and Outcome of Aortitis at A Single Center


Patient with aortitis, giant cell arteritis, Takayasu arteritis or retroperitoneal fibrosis treated at ANW between 1/1/2009 and 4/17/2015 (n=171)

Total Exclusions (n=156)
  - No aortic involvement (n=153)
  - Lack of clinical/imaging evidence (n=2)
  - Lost to follow-up (n=1)

Patients in Final Analysis (n=15)

Takayasu Arteritis (n=8)
Idiopathic inflammatory (n=5)
Giant cell arteritis (n=2)

Figure: Flow chart of the selection of aortitis study population with exclusion criteria

### METHODS

#### Table 1: Clinical criteria used to categorize patients with aortitis

<table>
<thead>
<tr>
<th>Takayasu Arteritis</th>
<th>Giant Cell Arteritis</th>
<th>IgG4-related system disease</th>
<th>Inflammatory Idiopathic Aortitis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Criteria (3 or more)</td>
<td>Criteria (3 or more)</td>
<td>Criteria (5 or more)</td>
<td>Based on exclusion (criteria for other diagnoses are not met)</td>
</tr>
<tr>
<td>Age at onset: &lt;40 years</td>
<td>Age at onset: ≥ 50 years</td>
<td>Diffuse or local swelling, masses, or thickness in single or multiple organs</td>
<td>Characteristic histopathological appearance (dense lymphoplasmacytic infiltrate, fibrosis, obliterative phlebitis)</td>
</tr>
<tr>
<td>Claudication of extremities</td>
<td>New headache</td>
<td>Increased numbers of IgG4+ plasma cells or IgG4/IgG+ plasma cell ratio of &gt;40%</td>
<td></td>
</tr>
<tr>
<td>Decreased brachial artery pulse</td>
<td>Temporal artery abnormality</td>
<td>Elevated erythrocyte sedimentation rate</td>
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<td>Blood pressure difference &gt;10 mm Hg</td>
<td>Abnormal bruit over subclavian arteries or aorta Arteriogram abnormality</td>
<td>Abnormal biopsy</td>
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#### METHODS: Study Definitions

- An ESR >21 mm/hr, a CRP > 0.50mg/dL, and an IgG4 > 135 mg/dL were considered abnormal

- Normal aortic thickness was considered to be <4 mm

- Follow-up was defined individually as the most recent hospital encounter or clinic visit during which imaging was performed

- Combined outcome was assessed using clinical presentation, laboratory data, and imaging findings. In cases of discordance between these three variables, imaging findings were considered to be the most accurate

## RESULTS: Study Population

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## Patient baseline clinical and laboratory characteristics

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<td>Length of hospital stay (days), median (25th, 75th percentile)</td>
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<td>5 (6, 9)</td>
<td>51 (28, 81)</td>
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### Aortitis Management Regimen

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<td>Other immunosuppressants/ modulators, (%)</td>
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### Indications, Outcomes, and Complications of Interventional Procedures

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<tr>
<th>Patient</th>
<th>Indication for Procedure</th>
<th>Type of Procedure</th>
<th>Procedure Outcome</th>
<th>Complication</th>
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<tbody>
<tr>
<td>2</td>
<td>Severe stenosis of left main coronary artery</td>
<td>Coronary artery bypass grafting</td>
<td>Successful</td>
<td>N/A</td>
</tr>
<tr>
<td>4</td>
<td>Severe stenosis and thrombosis of bilateral common iliac artery</td>
<td>Aspiration thrombectomy, bilateral drug-eluting balloon angioplasty, left stenting</td>
<td>Successful</td>
<td>Right iliac artery dissection</td>
</tr>
<tr>
<td>9</td>
<td>Severe stenosis of right renal artery, induced hypertension</td>
<td>Angioplasty and stenting of the right renal artery</td>
<td>Successful</td>
<td>N/A</td>
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<tr>
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<td>Symptomatic severe stenosis of bilateral subclavian artery</td>
<td>Balloon angioplasty and stenting of bilateral subclavian artery</td>
<td>Successful</td>
<td>Right subclavian artery re-stenosis</td>
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<tr>
<td>11</td>
<td>Aortic valve insufficiency</td>
<td>Ascending aortic aneurysm repair with Hemashield Dacron Graft</td>
<td>Successful</td>
<td>Ischemic brain lesion</td>
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<tr>
<td>12</td>
<td>Occlusion of right carotid artery</td>
<td>Left-to-right carotid-carotid bypass</td>
<td>Successful</td>
<td>Seizure</td>
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Case Study Conclusion

- More prevalent in females except in idiopathic inflammatory aortitis.

- 4/5 of pts with idiopathic type had hx of smoking.

- All idiopathic cases involved the abdominal aorta.

- Most patients with TA presented with arterial stenoses and/or occlusion.

- All patients received medical therapy and 33% underwent interventional procedures leading to 67% improvement of disease activity or related complications.

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Vascular Medicine Work-up Algorithm for Patients with Aortitis

**Patients with suspected aortitis based on history and clinical presentation**

- Imaging: US, CTA, MRA and/or PET
- Histopathology: microscopic tissue examination
- Laboratory examination: CBC, ESR, CRP, c- and p-ANCA, IgG4, C3, C4, ANA, RF, Cryo, and blood or tissue cx

**Diagnosis**

- Infectious Aortitis
- Inflammatory Aortitis

**Management:**

- Consult I.D. and vascular services
- Antibiotics +/- surgical debridement and/or excision

**Management:**

- Consult vascular services including vascular medicine
- Management of the underlying etiology (TA, GCA, IgG4 disease, ECD, periartitis, or other systemic autoimmune disorders)
- Prednisone + steroid sparing agents
- Surgical or catheter-based revascularization (if severe ischemia or large aneurysm when disease is under control)
Noticing the Difference ..

Which is Which?!?
**Differential Diagnoses: Very Challenging!**

- **GCA:** F, \( \geq 50 \) age, headache, elevated markers, PMR, temporal artery biopsy
- **TA:** F, < 40, BP difference, poor pulses, imaging findings
- **IgG4-RD:** classic organ involvement, elevated serum IgG4, imaging (periaortitis, sausage-shaped pancreas), pathology (storiform fibrosis, lymphoplasmacytic infiltrates with positive stain for IgG4)
- **Periaortitis:** ureter and periaortic involvement with fibrosis and inflammation
- **ECD:** multisystem non-Langerhans cell histiocytosis, periadventitial infiltrate, BRAF V600E, imaging (hairy kidney/coated aorta)
- **Idiopathic inflammatory aortitis:** exclusion of others

---

**Life is Full of Mysteries and Unknowns**

- There are known knowns
- These are things we know that we know
- There are known unknowns
- That is to say, there are things that we know we don't know, but there are also unknown unknowns
- There are things we don't know we don't know
Conclusion

- Aortitis (inflammatory, infectious, neoplastic, idiopathic) are very rare disorders.
- More research is needed to characterize and classify these disorders.
- Differential diagnoses can be very difficult.
- Requires extensive work up involving multidisciplinary approach including vascular medicine.
- Determining disease activity needs to be standardized.
- Management includes steroids and steroid sparing agents (new agents!).
- Revascularization when disease is inactive. Increasing interest in endovascular approach.

Thank you!
Questions?