2022 MID-ATLANTIC CONFERENCE

10th ANNUAL CURRENT CONCEPTS IN

VASCULAR THERAPIES

2022

Hilton Virginia Beach Oceanfront
Virginia Beach, Virginia

APRIL 28-30

Sentara Vascular Specialists

@adityasharmamd

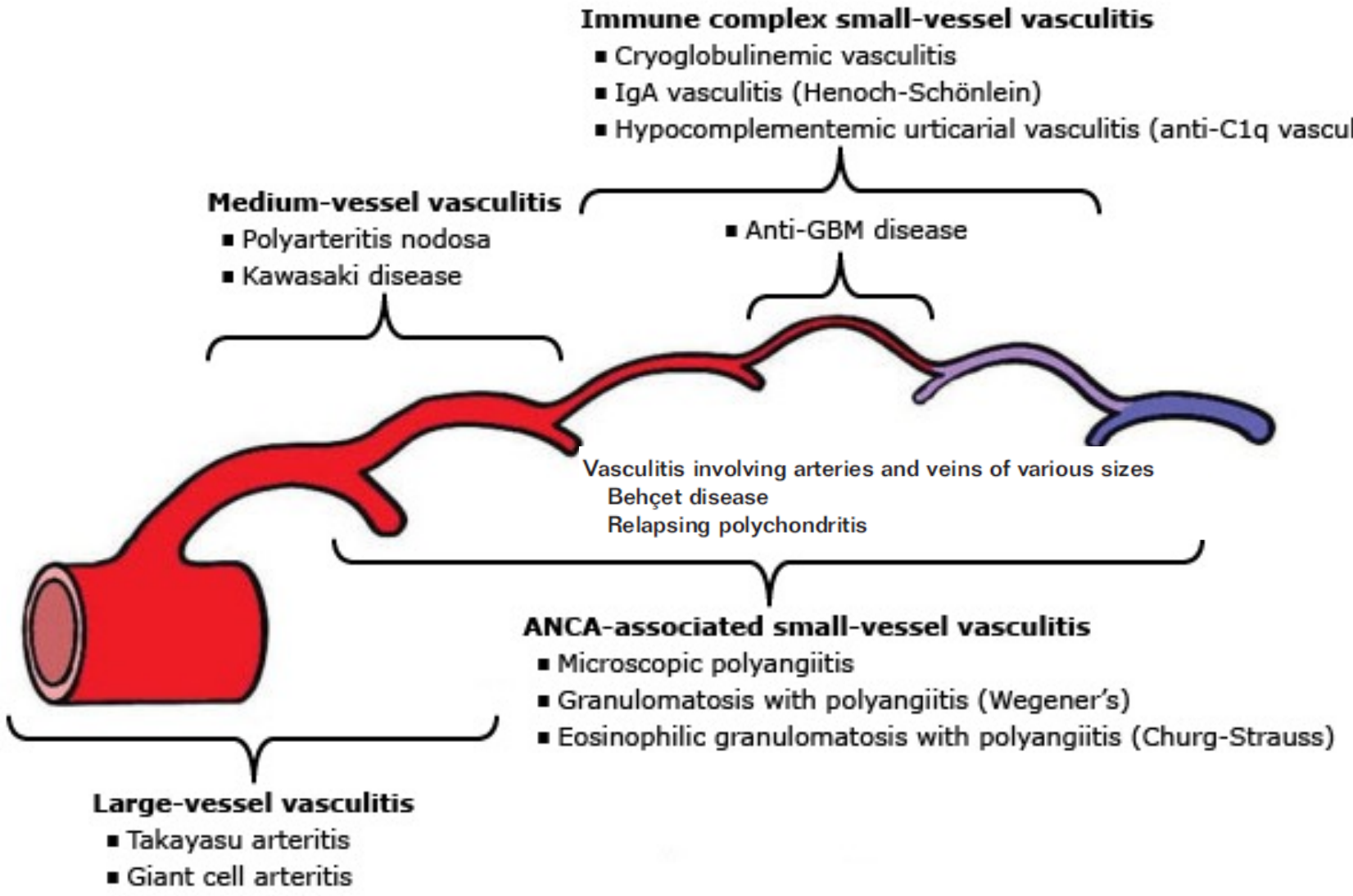
Vasculitis

Aditya Sharma, MD, FSVM
Associate Professor of Medicine
Medical Director, Vascular Medicine and PE Response Team
University of Virginia

Introduction

- Heterogeneous and complex group of diseases
- Cardinal feature: inflammation (systemic/ localized)
  - This often leads to narrowing, occlusion, or rupture of the involved vessel leading to end-organ or tissue damage
- Diverse presentations including varied organ distribution and size of vessel involvement
  - challenging to diagnose and treat

Vasculitis: Primary



Arthritis Rheum 2013; 65:1

Vasculitis: Secondary

Infectious

Infectious etiology
- Virus
  - Hepatitis B and C
  - Human immunodeficiency virus
  - Parvovirus B19
  - Cytomegalovirus
  - Herpes simplex virus
  - Varicella zoster
- Bacteria
  - Salmonella
  - Streptococcus
  - Staphylococcus
  - Clostridium septicum
  - Chlamydia pneumoniae
  - Mycobacterium tuberculosis
  - Treponema pallidum
  - Borrelia burgdorferi
  - Mycoplasma
  - Cryptococcus
  - Neisseria
  - Coccidioides

Connective tissue disease

- Relapsing Polychondritis
- Cogan Syndrome
- Rheumatoid arthritis
- Sjögren syndrome
- Systemic lupus erythematosus
- Scleroderma

Drugs

- D-Penicillamine
- Penicillin
- Propylthiouracil
- Hydralazine
- Minocycline
- Cocaine
- Leukotriene inhibitors

- Sulfasalazine
- Ciprofloxacin
- Pantoprazole
- Phenytoin
- Allopurinol
- Sulfonamides
- Thiazides

Malignancies

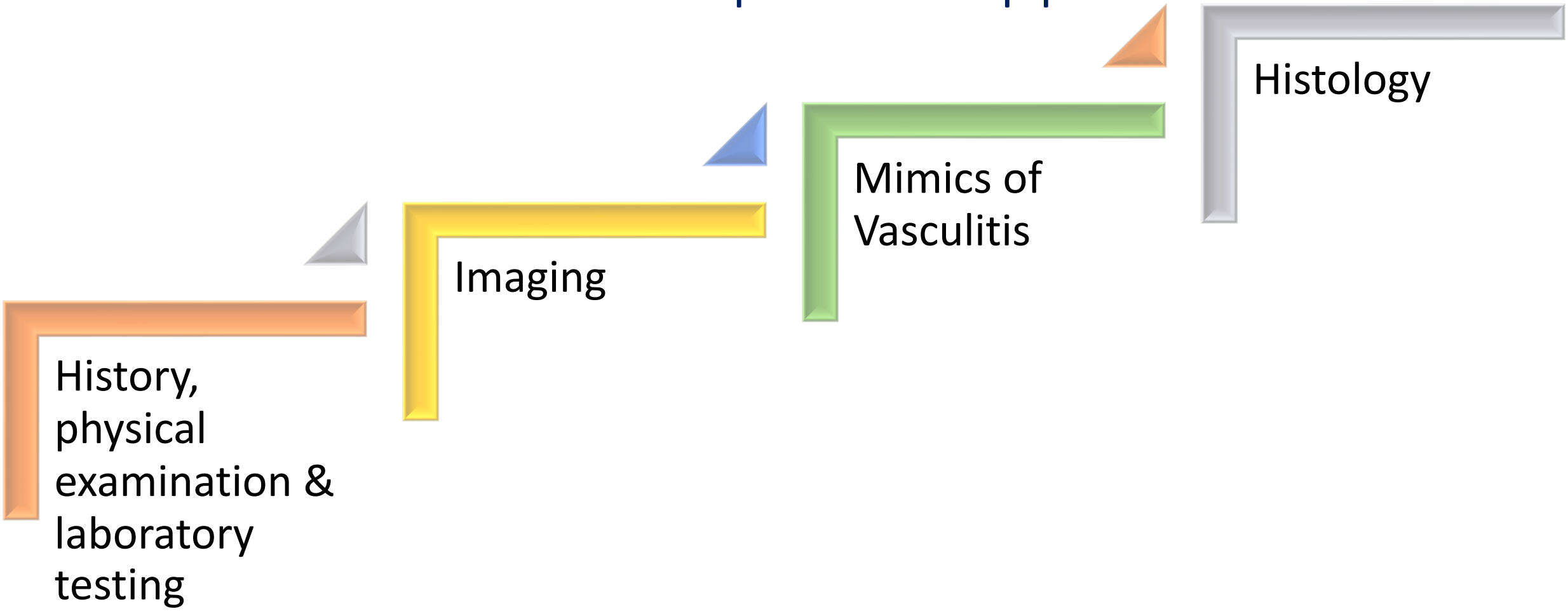
- Hematologic malignancies (myeloproliferative and lymphoproliferative disorders)
- Solid organ tumors including lung, colon, and GI carcinomas

Tech Vasc Interv Radiol. 2014 Dec;17(4):226-33

Vasculitis: Approach

- Is this a condition that could mimic the presentation of vasculitis?
- Is there a secondary underlying cause?
- What is the extent of vasculitis?
- How do I confirm the diagnosis of vasculitis?
- What specific type of vasculitis is this?

Vasculitis: Step-Wise Approach



Tech Vasc Interv Radiol. 2014 Dec;17(4):226-33

Step 1: Clinical and Laboratory Assessment

- Preceding illnesses
- New medications
- Constitutional symptoms
  - fever, night sweats, malaise, weight loss, arthralgia, myalgia
- Symptoms and signs localizing to specific organs

Laboratory Tests

Complete blood count
Inflammatory markers (ESR and CRP)
Creatinine level
Urinalysis
Liver function tests
Hepatitis B and C serologies
Serum cryoglobulins
Complement levels (C3, C4, and CH50)
ANCA
ANA
Cryoglobulins

Tech Vasc Interv Radiol. 2014 Dec;17(4):226-33

Clinical features: Small Vessel Involvement

Livedo reticularis

Cutaneous post-capillary venules → Palpable purpura

Glomerular capillaries → Haematuria, red cell casts in urine, proteinuria, and decline in renal function

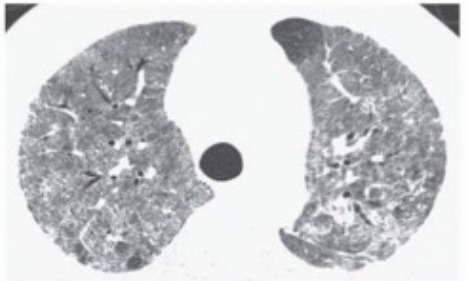
Pulmonary capillaries → Lung haemorrhage manifesting as breathlessness, haemoptysis and widespread alveolar shadowing on chest radiograph

palpable purpura

Leukocytoclastic Vasculitis

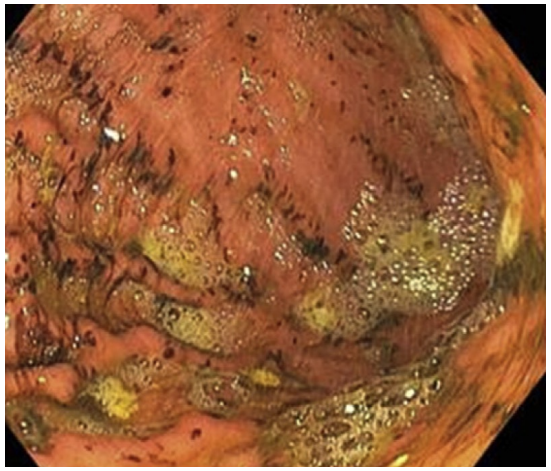
Urticarial Vasculitis

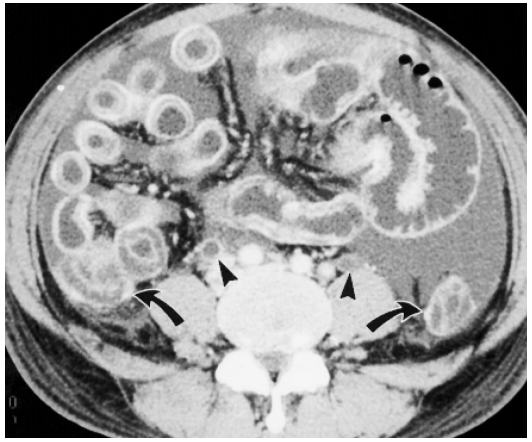
Pulmonary hemorrhag



Vasc Interv Radiol. 2014 Dec;17(4):226-33

Clinical features: Medium Vessel Involvement





Small cutaneous arteries	→	Necrotic lesions and ulcers, nail fold infarcts
Epineural arteries	→	Mononeuritis multiplex
Celiac and mesenteric	→	Abdominal pain, gastrointestinal bleeding and perforation Liver, pancreatic and splenic infarction
Renal	→	Renal infarction, hypertension, flank pain, hematuria etc
Coronaries	→	Myocardial infarction or angina, coronary artery aneurysm, ischemic cardiomyopathy
Pulmonary	→	Necrotic lesions leading to cavitating lung shadows on chest radiograph
Small arteries in ENT	→	Nasal crusting, epistaxis, sinusitis, deafness, stridor because of sub-glottic stenosis

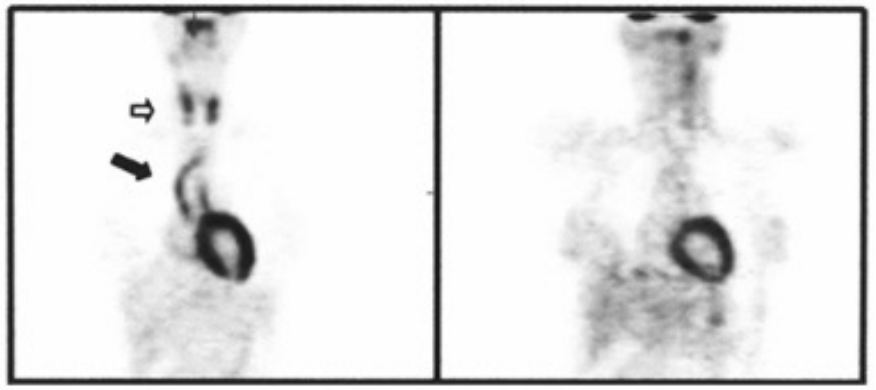
Postgrad Med J. 2006 Aug;82(970):483-8.

Clinical Features: Large Vessel Involvement

Extracranial branches of carotid artery	Temporal headache (temporal artery), blindness (ophthalmic artery), jaw claudication (vessels supplying muscles of mastication)
Thoracic aorta and its branches	Limb claudication, absent pulses and unequal blood pressure, bruits, thoracic aortic aneurysms

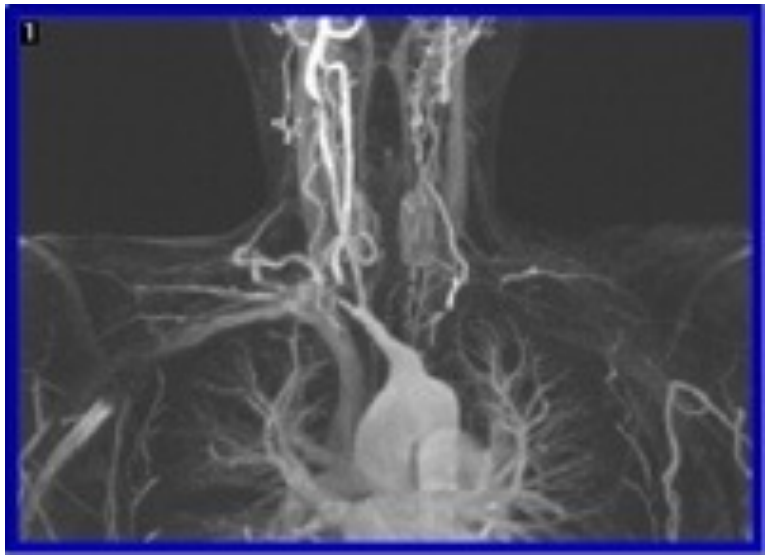
Postgrad Med J. 2006 Aug;82(970):483-8.

Step 2: Imaging



- Identify and guide treatment of vasculitides (large and medium vessel vasculitis)

- Imaging modalities:
  - Color duplex ultrasound (CDUS)
  - Computerized tomography (CT)
  - Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA)
  - Positron emission tomography (PET)

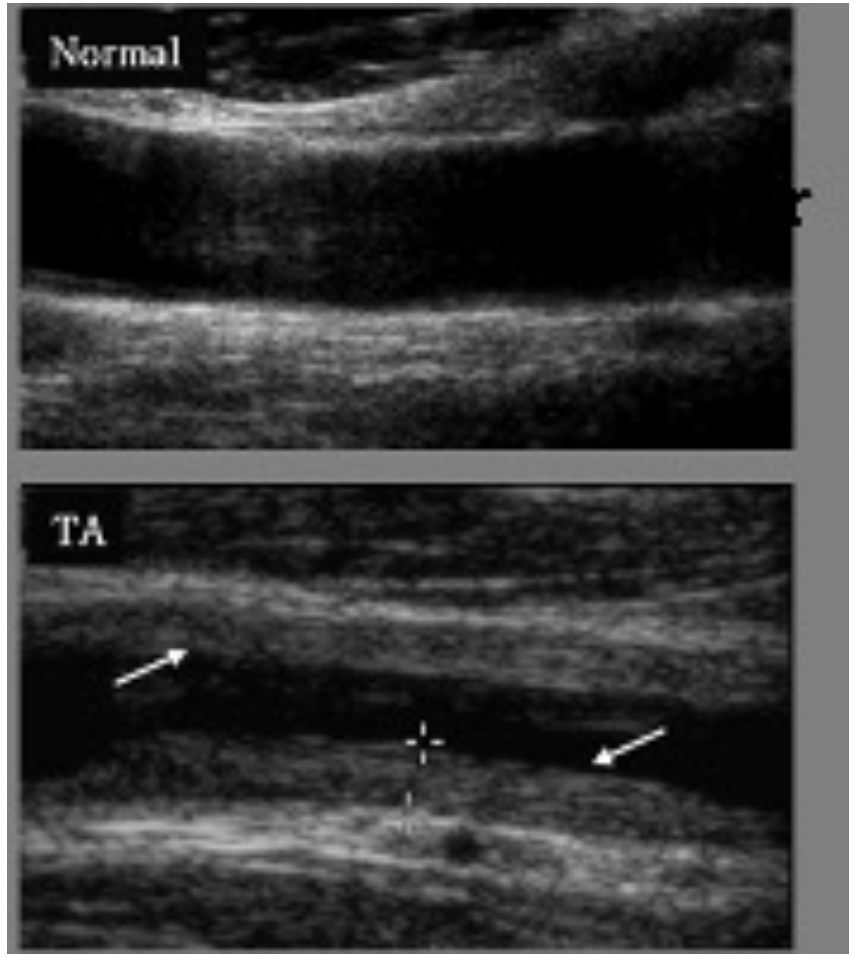


Cardinal Imaging Signs

- Vessel wall thickening
- Irregular contours
- Perivascular inflammation

- Aneurysms
- Stenosis
- Occlusion

Vascular remodeling secondary to inflammation

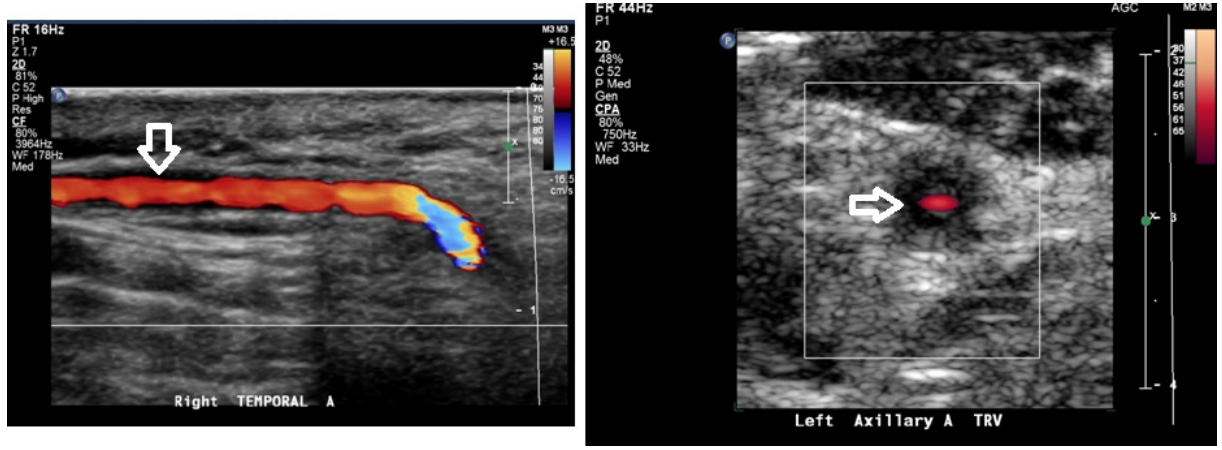


Tech Vasc Interv Radiol. 2014 Dec;17(4):226-33

Best Pract Res Clin Rheumatol. 2013 Feb;27(1):107-18

Ultrasonography

Giant Cell Arteritis

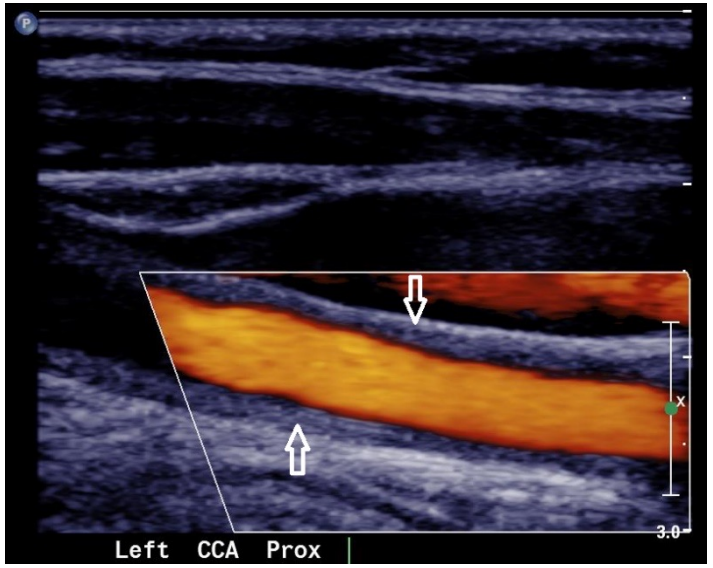


Halo sign: Hypoechoic edematous swelling in the vessel wall

- Negative predictive value → 92-96%
- Meta-analysis of 23 studies with GCA:
  - Sensitivity (87%) and specificity (96%) with CDUS
- Guide temporal artery biopsy

Images provided by Heather Gornik, MD and Esther Kim, MD

Takayasu Arteritis



"Macaroni sign": intimal thickening, irregular luminal contour and bright appearance

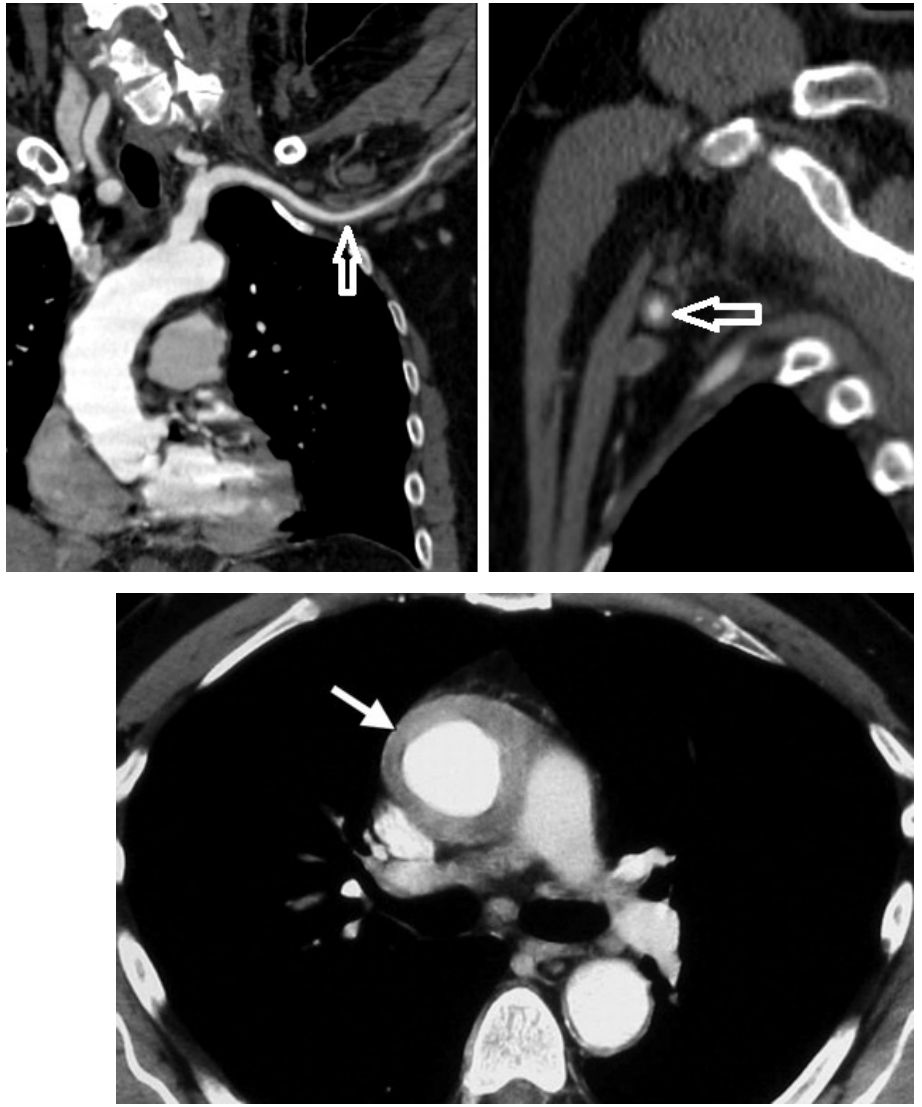
Vessel wall thickening of common carotid or subclavian artery >1.0 mm

Br J Surg, 97, 1765-71.
J Vasc Surg, 36, 1154-60.
Ann Intern Med, 142, 359-69.

Computed Tomography Angiography

- Good spatial resolution and fast scanning times

- Acute inflammatory phase: the classic “double ring” finding
  - Poorly enhancing inner ring: intimal hyperplasia
  - Brightly enhancing outer ring: active inflammation in the medial and adventitial layers of the artery
  - High specificity and sensitivity (>95 %) for large vessel vasculitis



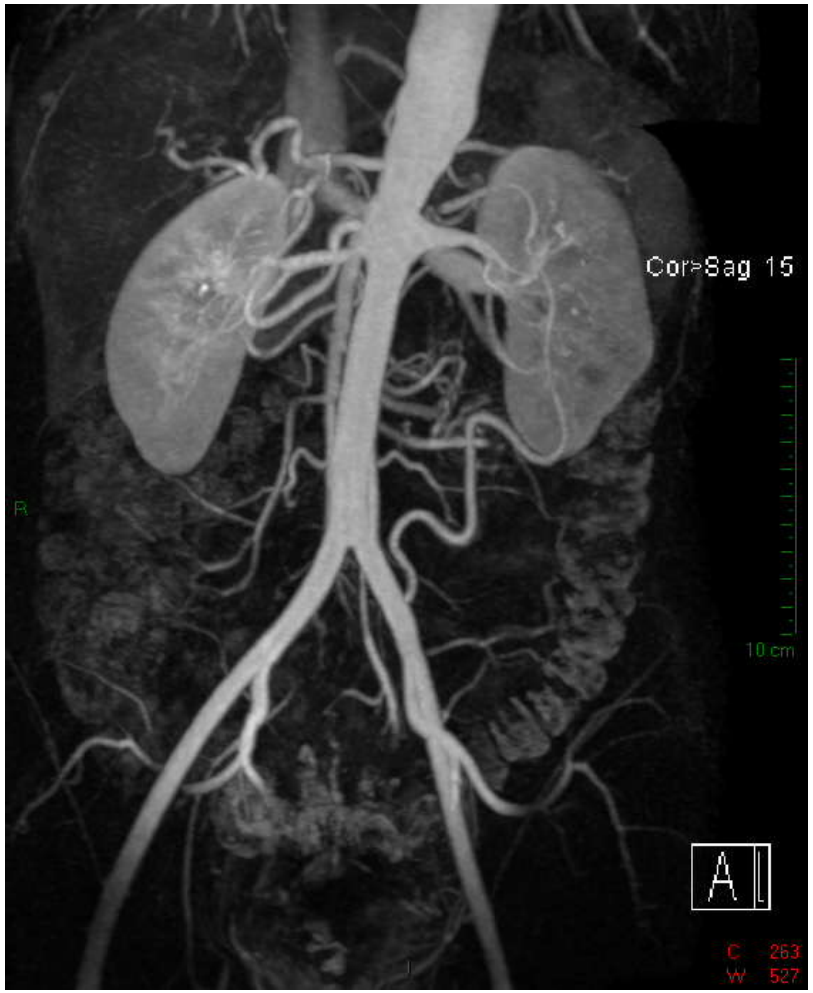
Radiographics. 2011 Mar-Apr;31(2):435-51
Insights Imaging. 2012 Dec; 3(6): 545–560.
J Am Coll Cardiol Img. 2014;7(6):605-619.

Chronic Changes

Calcification
Stenosis
Occlusions
Aneurysms

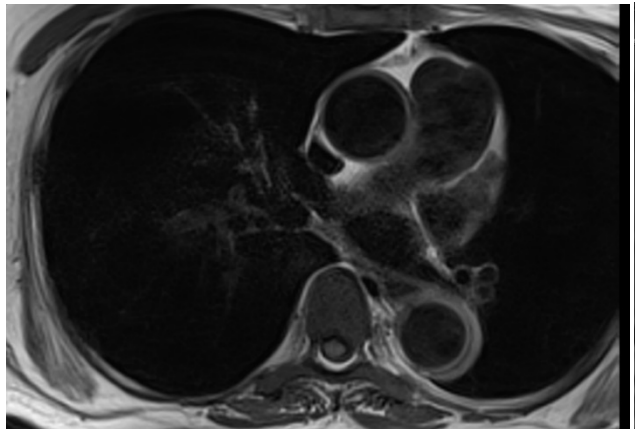
Monitor progression of aneurysm

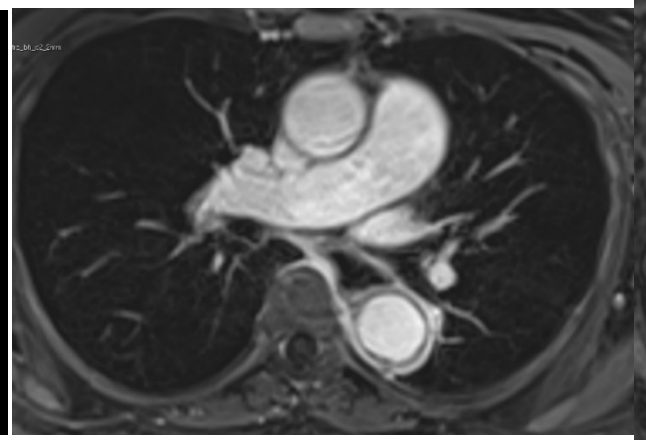
Takayasus Arteritis:

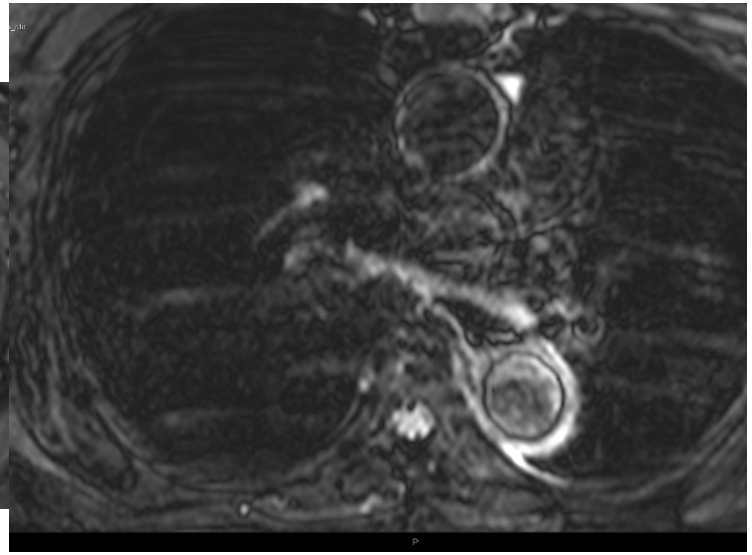


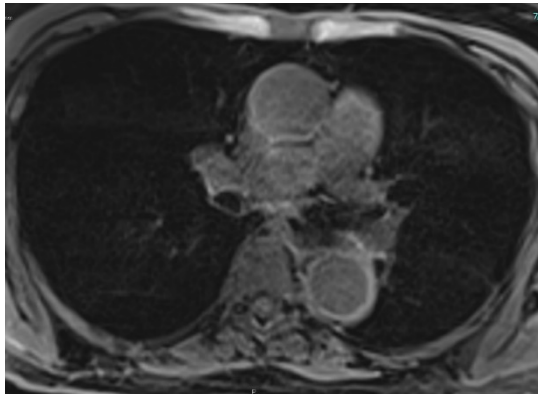
Magnetic Resonance Angiography

- Detect subtle changes in the aortic wall
- Increased wall thickness and wall edema: Fat-suppressed T2 black blood sequences
- Mural enhancement: T1-weighted sequences post contrast
  - Post contrast T1 images are superior to T2 or fat-suppressed images in detecting large-vessel inflammation and are required for detection of more subtle sign
- Periodic assessment providing luminal and vessel wall assessment
  - High signal on T2 weighted sequences and mural contrast enhancement > active disease and < initiation of immunosuppressive therapy.
- Sensitivity / specificity = 81% / 91%









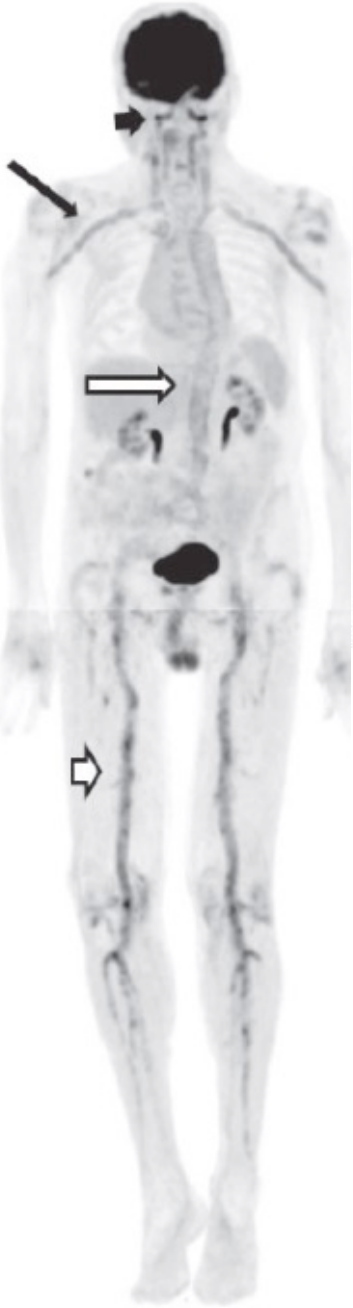
Arthritis Rheum, 46, 1634-42.
Arthritis Rheum, 58, 2574-8.
AJNR Am J Neuroradiol, 28, 1722-7.
Rheumatology (Oxford), 47, 65-7.

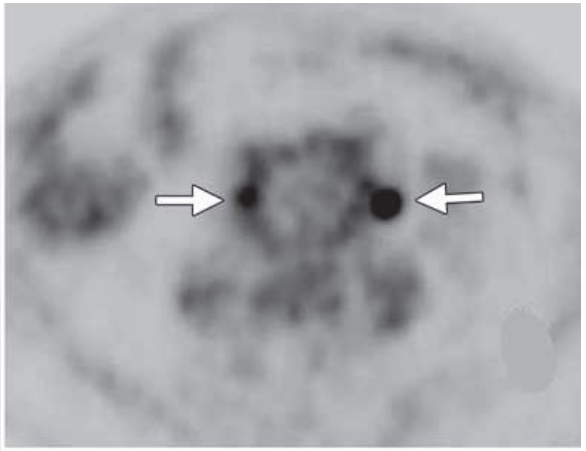
Positron Emission Tomography

- Uptake of IV radiolabeled glucose analogue FDG by activated cells in inflammatory processes

- Sensitivity / Specificity = 73% / 84%
- PPV / NPV = 82% / 77%
- Accuracy compared to clinical assessment alone = 71
- Combine with CT / MR ⬆ anatomical accuracy

- Steroid initiation: 50% accuracy





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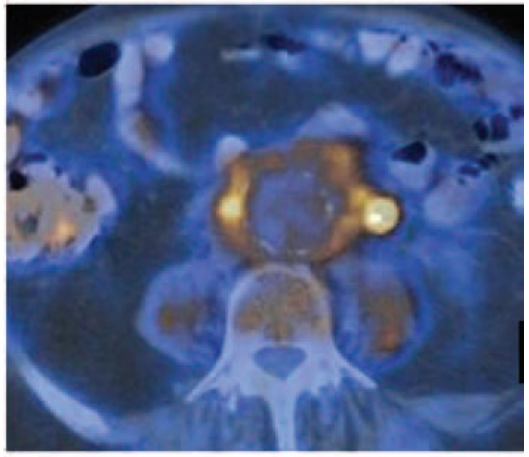
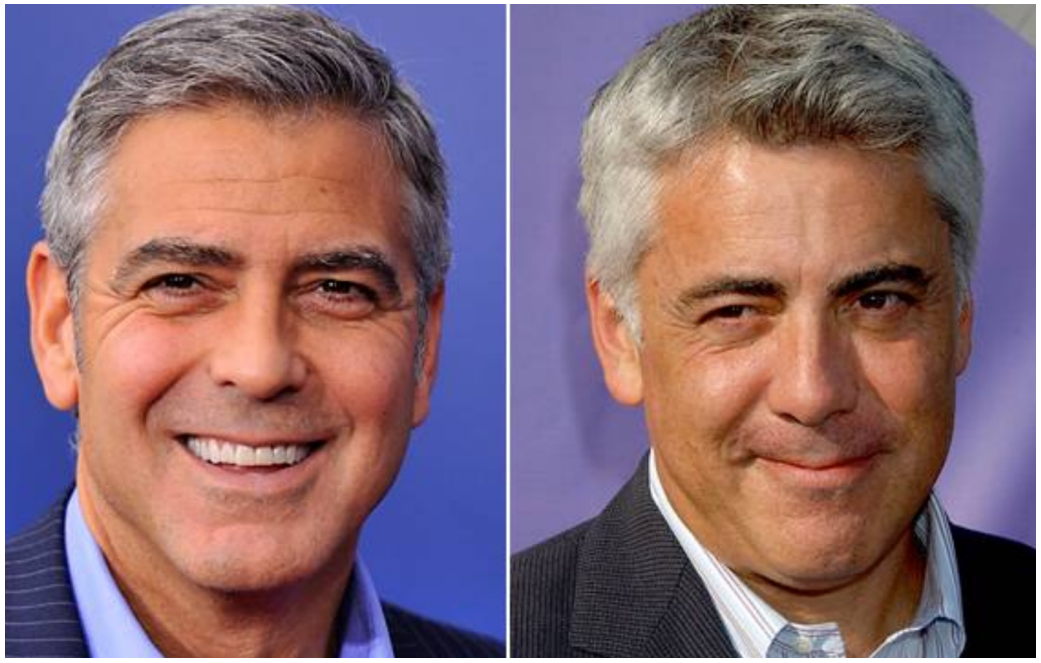


c.

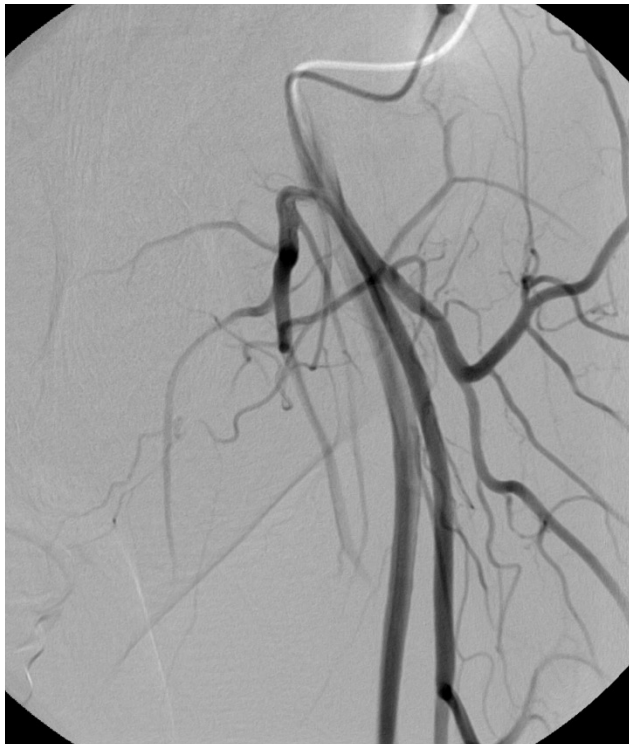
Rheumatology (Oxford), 47, 403-8.
Eur J Nucl Med Mol Imaging, 30, 730-6.
Eur J Nucl Med Mol Imaging, 39, 344-53.
Eur J Nucl Med Mol Imaging, 30, 1305-13.

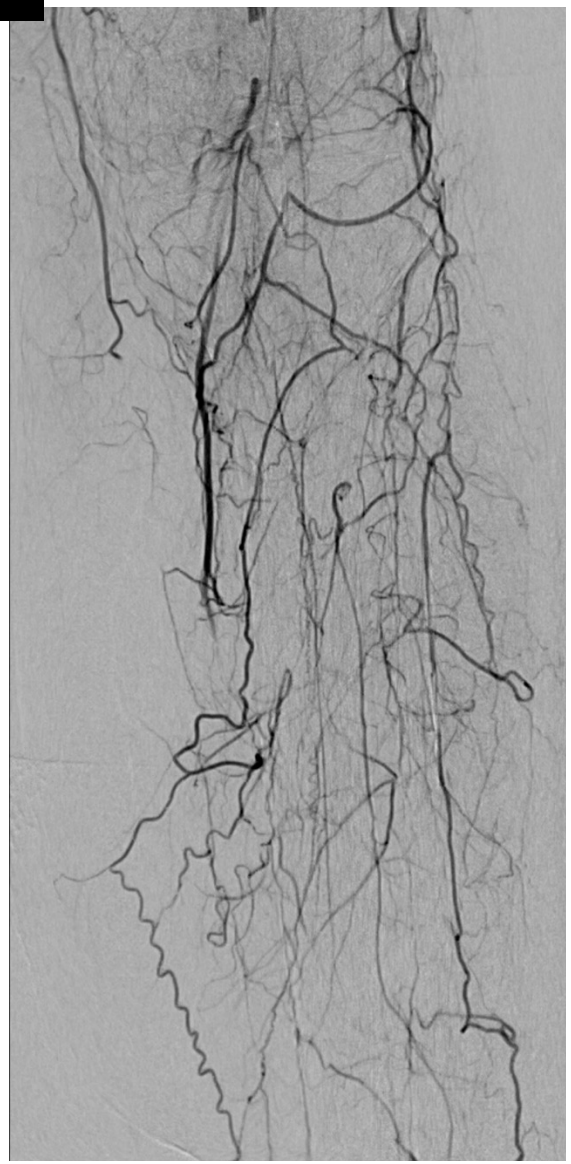
Step 3: Mimics



40 yr old with claudication

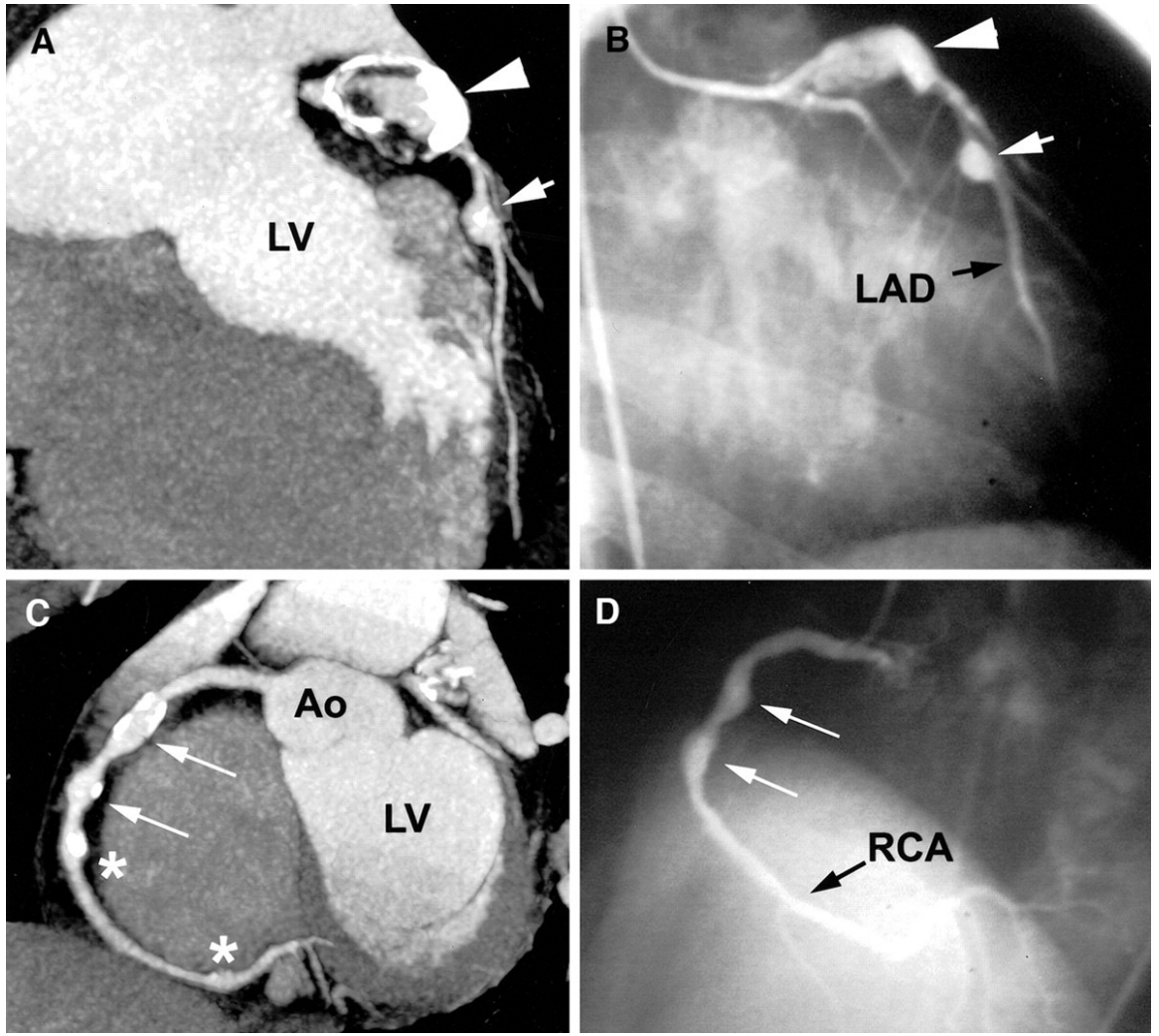
Buerger's disease





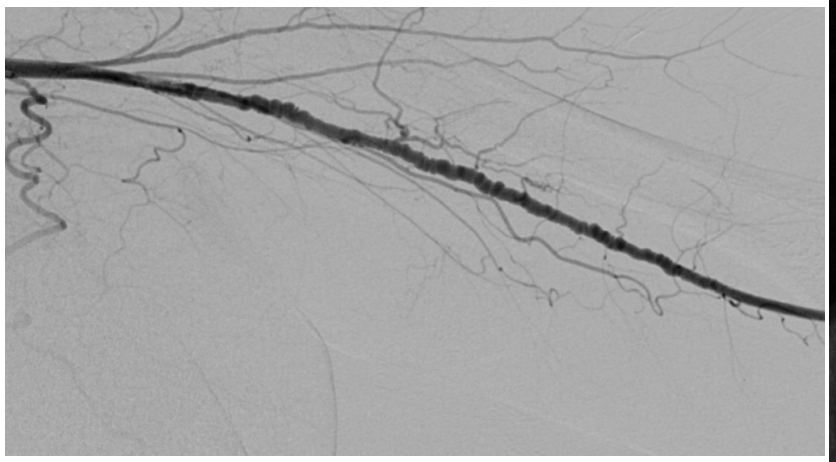
Kawasaki's Disease

40 year old with Chest pain



Mimics

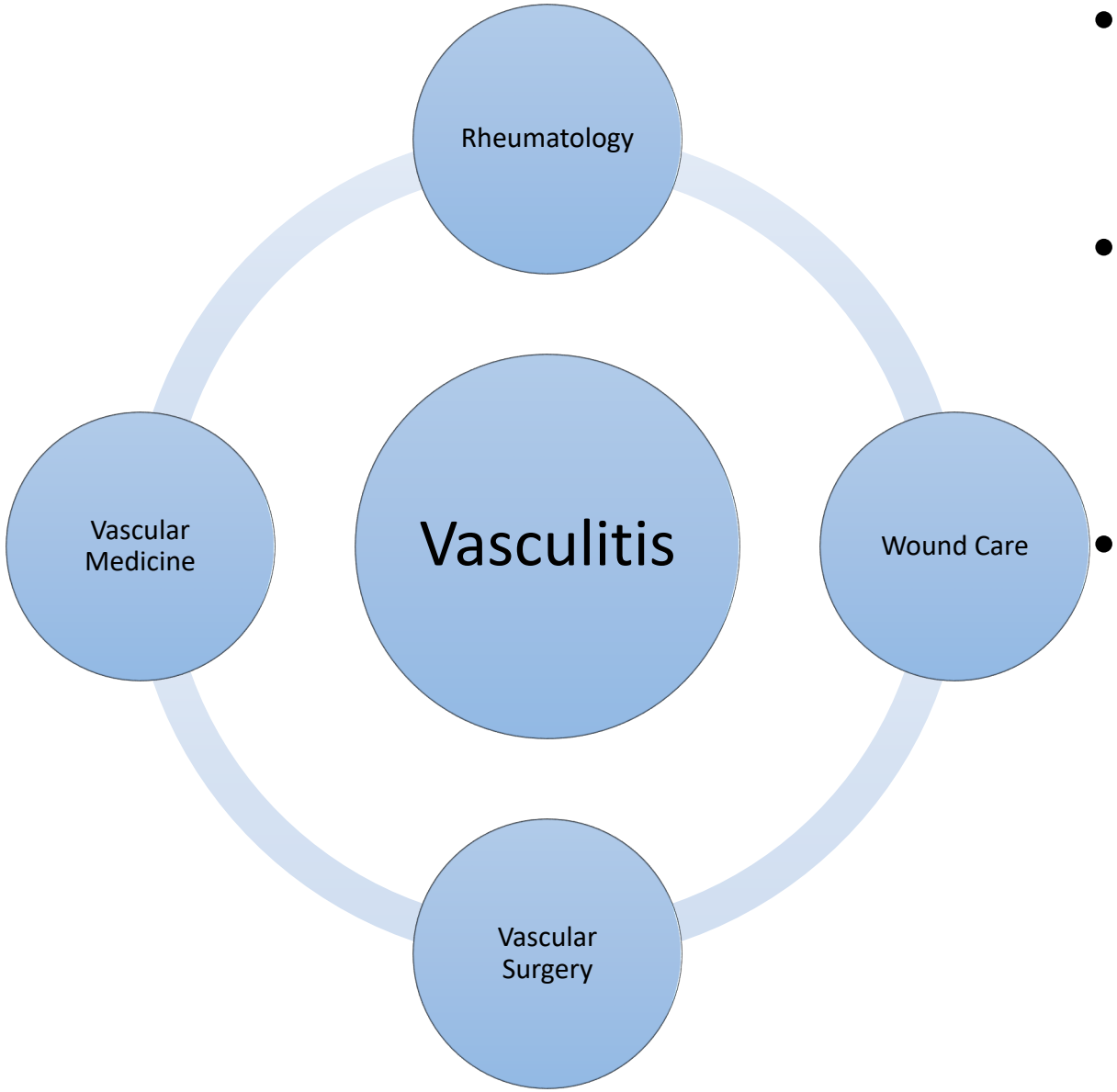
Fibromuscular Dysplasia



Connective tissue disease

Circulation: Cardiovascular Interventions. 2012;5:e9-e11

Treatment



- Goals:
  - Stop active inflammation
  - Prevent permanent damage
- Treatment approach:
  - Remission induction
  - Remission maintenance
  - Monitoring
- Treatment
  - Medical therapies
  - Surgical and endovascular therapies

- Treatment protocols are tailored to the type of vasculitis, extent of disease and clinical presentation (acute ischemia vs. claudication vs. minimal or asymptomatic)

Treatment

- Remission induction
  - Intensive with need for close monitoring
  - Delay in diagnosis and induction→ worsening morbidity and mortality
  - Glucorticoids +/- other immunosuppressive agents
- Remission maintenance
  - Maintain control of disease activity
  - Prevent disease recurrence following reduction or discontinuation of medications
  - Minimize the risks of drug toxicity
  - Management of osteoporosis, chronic infections, accelerated atherosclerosis
- Monitoring
  - Disease activity (clinical, laboratory and imaging)
  - Drug toxicity

Treatments for Inflammatory Vasculitis

Commonly Used Medications/Treatments

- Aspirin
- Glucocorticoids
- Cyclophosphamide
- Azathioprine
- Methotrexate
- Mycophenolate mofetil
- Cyclosporine and tacrolimus (FK506)
- Antiviral agents
- Plasmapheresis
- Intravenous immunoglobulin

Newer

- Rituximab (anti-CD20)
- Inhibitors of tumor necrosis factor-α
- Tociluzumab (anti–IL-6)
- Mepolizumab (anti–IL-5)
- Abatacept (CTLA4-Ig)
- Other experimental biologics

Surgical/Invasive Treatments

- Balloon angioplasty
- Intravascular stents (± drug-eluting coating)
- Vascular bypass or replacement grafts
- Reconstructive surgery

Merkel P. Vasculitis. Vascular Medicine: A Companion to Braunwald's Heart Disease

Conclusion

- Challenging group of disorders
  - Difficult to diagnose and treat
- Multidisciplinary care across multiple specialties is key
- Longitudinal follow-up is imperative