Unusual Vascular Disorders

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Unusual Vascular Disorders

These disorders may mimic other more commonly seen diseases. They are important for formulating a differential diagnosis and managing your cardiovascular patient.
Non-Atherosclerotic Arterial Vascular Disorders

- Fibromuscular Dysplasia
- Popliteal Artery Entrapment Syndrome
- Cystic Adventitial Disease
- External Iliac Artery Endofibrosis
- Thromboangiitis obliterans
- Segmental Arterial Mediolysis
- Uncommon Arteriopathies

When should you think of these disorders?
- Younger patients
- Patients with no traditional risk factors for atherosclerosis
A 44 year old female presents to your office complaining of a whooshing noise in her ears. She has a history of hypertension but otherwise is in good health. You suspect she might have…….

a). Fibromuscular dysplasia
b). Meniere's disease
c). An acoustic neuroma
d). A glomus tumor
e). A stroke
Fibromuscular Dysplasia or FMD

- Affects small to medium sized vessels
- Nonatherosclerotic, noninflammatory vascular disease
- Affects young to middle aged women
- Results in arterial stenosis, occlusion, aneurysm formation or dissection

<table>
<thead>
<tr>
<th>Renal</th>
<th>Cerebrovascular</th>
<th>Visceral</th>
<th>Extremities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertension</td>
<td>Asymptomatic or headache, pulsatile</td>
<td>Abdominal pain, weight loss,</td>
<td>Intermittent claudication,</td>
</tr>
<tr>
<td></td>
<td>tinnitus, vertigo, syncope, TIA, CVA,</td>
<td>epigastric bruit</td>
<td>critical limb ischemia,</td>
</tr>
<tr>
<td></td>
<td>intracranial aneurysm</td>
<td></td>
<td>embolization</td>
</tr>
</tbody>
</table>
Fibromuscular Dysplasia

Predominately affects renal and internal carotid arteries - (65% of the Cases)

NEJM 2004; 350: 1862-1871
Circulation 2014;129: 1048-1078
Fibromuscular Dysplasia

Reported in virtually every Arterial Bed
FMD and Coronary disease

LAD artery disease and right renal artery FMD
FMD of coronary arteries reported as a cause of MI
(7 cases of renal FMD with well-demarcated obliterative CAD)

Cath and Cardiovasc Interv 2005; 64: 138
Epidemiology of FMD

- Prevalence in the general population is not known
- More common in women than men (9:1 ratio)
- Prevalence of renal artery FMD estimated - 1% to 5.7%
- Prevalence of carotid, vertebral and intracranial FMD is unknown but estimated - 0.3% to 3.2%

Circulation 2014’ 129:1048-1078
Orphanet J Rare Dis. 2007;2:28
Stroke 1982;13:53-58
### Fibromuscular Dysplasia – Older Classification

<table>
<thead>
<tr>
<th>TYPES OF FMD</th>
<th>CHARACTERISTICS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Medial fibroplasia</td>
<td>Most common, “string of beads” <em>(beads larger than caliber of vessel)</em> located middle to distal artery 75 to 80% of all cases</td>
</tr>
<tr>
<td>Perimedial fibroplasia</td>
<td>Found in &lt;10% of cases. Usually young girls, beads smaller than caliber of vessel, focal stenoses</td>
</tr>
<tr>
<td>Medial hyperplasia</td>
<td>Found &lt;1% and is often indistinguishable from intimal fibroplasia</td>
</tr>
<tr>
<td>Intimal fibroplasia</td>
<td>Under 10%, may mimic Takayasu’s or GCA with long, smooth narrowing of the vessel</td>
</tr>
<tr>
<td>Adventitial (periarterial) hyperplasia</td>
<td>Rarest type</td>
</tr>
</tbody>
</table>
## 2014 American Heart Association
### Classification of Fibromuscular Dysplasia

<table>
<thead>
<tr>
<th></th>
<th>Multifocal</th>
<th>Focal</th>
</tr>
</thead>
<tbody>
<tr>
<td>Angiographic appearance</td>
<td>Alternating dilatation and constriction of the vessel (string of beads) Areas of dilatation are larger than the normal caliber of the artery Occurs in the mid and distal portion of the renal, internal carotid, and vertebral arteries May occur in any other artery in the body†</td>
<td>Focal concentric or tubular stenosis*</td>
</tr>
<tr>
<td>Typical histology</td>
<td>Medial fibroplasia (most common)</td>
<td>Intimal fibroplasia (most common)</td>
</tr>
<tr>
<td></td>
<td>Perimedial fibroplasia (rare)‡</td>
<td>Adventitial (periarterial) fibroplasia (rare)</td>
</tr>
<tr>
<td>Associated features</td>
<td>Aneurysm, dissection, and vessel tortuosity of medium-sized arteries may be present; multifocal and focal lesions may coexist in the same patient</td>
<td>Medial hyperplasia (rare)</td>
</tr>
</tbody>
</table>

*Lesions are not necessarily confined to the mid or distal portion of the artery (ie, can occur in any arterial segment).
†There are no cases of aortic fibromuscular dysplasia that are well documented pathologically.
‡This rare form of fibromuscular dysplasia typically occurs in young girls (eg, those 5 to 15 years of age). Although there is a beaded appearance to the renal arteries, the beads are smaller than the normal renal artery and less numerous. There is often collateralization around the area of stenosis (Figure 3).
Differential Diagnosis of FMD

- **Standing waves or stationary waves** (associated with a catheter or contrast-induced spasm of the artery)
- **Atherosclerosis** (FMD usually occurs at mid to distal portion of the arteries)
- **Vasculitis** (FMD is noninflammatory)
- **Segmental arterial mediolysis (SAM)** (may be difficult to differentiate as radiographic findings are similar - SAM more likely to affect visceral arteries than FMD)
Standing waves. Note the regular oscillations in the superficial femoral artery (arrows, A) and tibial arteries (arrows, B). This is often mistaken for FMD.
Cause of Fibromuscular Dysplasia

Pathogenesis: Unknown

- Hormonal effects (such as estrogen)
- Cigarette smoking
- Mechanical stress on vessel walls
- Genetic factors (but no genes for FMD have yet been identified)
Fibromuscular Dysplasia - Diagnosis

- History and physical examination
- Duplex ultrasound
- CTA, MRA
- Catheter-based angiography
### Presenting Signs and Symptoms
(Patients in the US Registry for FMD – 447 patients)

<table>
<thead>
<tr>
<th>Symptoms/Signs</th>
<th>(%) of 447 patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertension</td>
<td>63.8</td>
</tr>
<tr>
<td>Headache (current, history of)</td>
<td>52.4, 30.2, 38.7</td>
</tr>
<tr>
<td>Pulsatile tinnitus</td>
<td>27.5</td>
</tr>
<tr>
<td>Dizziness</td>
<td>26</td>
</tr>
<tr>
<td>Cervical bruit</td>
<td>22.2</td>
</tr>
<tr>
<td>Neck pain</td>
<td>22.2</td>
</tr>
<tr>
<td>Chest pain or SOB</td>
<td>16.1</td>
</tr>
<tr>
<td>Flank/abdominal pain</td>
<td>15.7</td>
</tr>
<tr>
<td>Aneurysm</td>
<td>14.1</td>
</tr>
<tr>
<td>Cervical dissection</td>
<td>12.1</td>
</tr>
<tr>
<td>Epigastric bruit</td>
<td>9.4</td>
</tr>
<tr>
<td>TIA</td>
<td>8.7</td>
</tr>
<tr>
<td>Postprandial abdominal pain</td>
<td>7.8</td>
</tr>
<tr>
<td>Stroke</td>
<td>6.9</td>
</tr>
<tr>
<td>Claudication</td>
<td>5.2</td>
</tr>
</tbody>
</table>
# Prevalence and Vascular Distribution of Arterial Aneurysm and Dissection in the US Registry for FMD

<table>
<thead>
<tr>
<th></th>
<th>N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Aneurysm</strong></td>
<td></td>
</tr>
<tr>
<td>Renal</td>
<td>25/76 (32.9%)</td>
</tr>
<tr>
<td>Carotid</td>
<td>16/76 (21.1%)</td>
</tr>
<tr>
<td>Aorta</td>
<td>15/76 (19.7%)</td>
</tr>
<tr>
<td>Ascending</td>
<td>6/76 (7.9%)</td>
</tr>
<tr>
<td>Descending</td>
<td>4/76 (5.3%)</td>
</tr>
<tr>
<td>Abdominal</td>
<td>5/76 (6.6%)</td>
</tr>
<tr>
<td>Celiac</td>
<td>12/76 (15.8%)</td>
</tr>
<tr>
<td>Cerebral</td>
<td>9/76 (11.8%)</td>
</tr>
<tr>
<td>Mesenteric</td>
<td>5/76 (6.6%)</td>
</tr>
<tr>
<td>Basilar</td>
<td>5/76 (6.6%)</td>
</tr>
<tr>
<td>Vertebral</td>
<td>2/76 (2.6%)</td>
</tr>
<tr>
<td>Subclavian</td>
<td>2/76 (2.6%)</td>
</tr>
<tr>
<td>Popliteal</td>
<td>2/76 (2.6%)</td>
</tr>
<tr>
<td><strong>Dissection</strong></td>
<td></td>
</tr>
<tr>
<td>Carotid</td>
<td>68 (75)</td>
</tr>
<tr>
<td>Renal</td>
<td>19 (22)</td>
</tr>
<tr>
<td>Vertebral</td>
<td>15 (17)</td>
</tr>
<tr>
<td>Mesenteric</td>
<td>4 (4.5)</td>
</tr>
<tr>
<td>Coronary</td>
<td>3 (3.4)</td>
</tr>
<tr>
<td>Celiac</td>
<td>2 (2.3)</td>
</tr>
<tr>
<td>Iliac</td>
<td>2 (2.3)</td>
</tr>
</tbody>
</table>

Circulation 2014;129:1048-1078
Features suggesting FMD include elevated velocities, turbulence of color or spectral Doppler flow and tortuosity in the mid and distal segment of the renal or carotid arteries.

- Beading may be seen.
- Doppler criteria used for atherosclerotic renal or carotid artery disease cannot be used to determine the severity of FMD.

U/S of Right ICA “beading of vessel”
Color Doppler showing turbulence and spectral analysis demonstrating high peak systolic (419 cm/s) and end-diastolic velocities (186 cm/s). Color power angiography demonstrating severe tortuosity and redundancy (S curve) of the internal carotid artery.
Fibromuscular Dysplasia - CTA or MRA

CTA images demonstrating “string of beads”

MR imaging demonstrating “string of beads”
Perimedial fibroplasia of the renal artery.
The beads (arrow) are smaller and less numerous than in medial fibroplasia.

Typical arteriographic findings of multifocal fibromuscular dysplasia in the carotid and renal arteries according to the American Heart Association classification system. Angiographic pattern is indicative of medial fibroplasia with multiple areas of alternating stenosis and dilatation (string of beads).

Arterial tortuosity is frequently encountered in patients with fibromuscular dysplasia.

Perimedial fibroplasia of the renal artery. The beads (arrow) are smaller and less numerous than in medial fibroplasia.
Treatment of Fibromuscular Dysplasia

- Medical therapy
  - antiplatelet and antithrombotics
  - antihypertensive therapy
  - cardiovascular risk factors and lifestyle modification
- Endovascular therapy
  - angioplasty
  - stenting (dissections)
  - coils, stents (aneurysms)
- Surgery
Case Report

A 28 year old male is complaining of left calf pain while running. He denies any recent trauma or injury. He has no cardiovascular risk factors. You suspect he has……..

a). Fibromuscular dysplasia
b). Popliteal artery entrapment syndrome
c). A torn meniscus
d). Anterior cruciate ligament injury
Popliteal Artery Entrapment Syndrome (PEAS)

- A group of conditions in which compression of the popliteal artery, vein, and tibial nerve (singly or in combination) in the popliteal fossa by surrounding musculoskeletal structures causes vascular and neurogenic symptoms.
PEAS

- Rare, but not considered a benign condition as progressive injury to the popliteal artery can lead to limb loss
- Median age 29 (16-47)
- Athletic young males (83%)
- Prevalence is unknown (reports range from 0.2% in Greek recruits to 3.5% in postmortem study of patients with vascular disease)
- Often misdiagnosed or simply overlooked

J Vasc Surg 2012;55:252-262
Vascular and Endovascular Surgery 2013;47:513-518
**Symptoms:**

- Exercise-induced intermittent claudication (running, cycling)
- Affects calf and foot (69% of all patients)
- Onset of symptoms often sudden
- Acute limb ischemia including distal limb thromboemboli or CLI (rest pain, ulcers)
- Bilateral PAES (30% to 67%)
- Limb swelling *(popliteal vein entrapment)*
- Nocturnal cramps and paresthesias *(tibial nerve entrapment)*
Poqliteal Vascular Entrapment Forum Classification for PEAS

<table>
<thead>
<tr>
<th>Type</th>
<th>Involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I</td>
<td>Popliteal artery running medial to the medial head of the gastrocnemius</td>
</tr>
<tr>
<td>Type II</td>
<td>Medial head of gastrocnemius laterally attached</td>
</tr>
<tr>
<td>Type III</td>
<td>Accessory slip of gastrocnemius/fibrous band arising from medial head of gastrocnemius</td>
</tr>
<tr>
<td>Type IV</td>
<td>Popliteal artery passing below popliteus muscle/fibrous bands</td>
</tr>
<tr>
<td>Type V</td>
<td>Primarily venous entrapment (more common in females)</td>
</tr>
<tr>
<td>Type VI</td>
<td>Other variants</td>
</tr>
<tr>
<td>Type VII</td>
<td>Functional entrapment defined as compression due to muscular hypertrophy</td>
</tr>
</tbody>
</table>

Congenital abnormality between popliteal artery and medial head of gastrocnemius muscle

Stenosis, occlusion, thromboembolism or aneurysmal formation can occur

**Diagnosis:**

- History and physical exam
- Examine patient with passive dorsiflexion of the foot; plantar flexion against active resistance
PEAS - Pulse Volume Recordings

ABI rest 1.13

Dorsiflexion of right foot
Pulse not Dopplerable

Diagnosis:
• PVR’s with exercise followed by flexion maneuvers
Duplex ultrasound scanning of the popliteal artery (healthy volunteer) in rest showing triphasic signal. B, Duplex ultrasound scanning of the popliteal artery (same person) during plantar flexion unchanged normal flow and unchanged max velocity.
**PEAS - Duplex Ultrasound**

**Duplex scanning**: longitudinal view of popliteal artery. In *neutral position*, normal typical triphasic waveform is noticed.

**Duplex scanning**: longitudinal view of popliteal artery. During *dorsiflexion* of the ankle, a biphasic waveform with altered flow velocity is recorded.

Clin J Sport Med 2004; 14: 8-12
Allows visualization of the common vascular (arterial stenosis, occlusion, aneurysmal change and deviated course) and abnormal muscle attachments and muscular hypertrophy
Bilateral popliteal artery entrapment demonstrating medial displacement of the popliteal artery (left) (asymptomatic) and near occlusion of the popliteal artery on the right (symptomatic).

Angiography (provocation and static) (deviation of the popliteal artery, may appear narrowed, poststenotic dilatation, occlusion or aneurysmal formation)
**PEAS - Treatment**

- **Catheter-directed thrombolysis and endovascular techniques** – "complementary treatment strategies"
- **Surgery for PAES** - (myotomy)
  - release muscle to free entrapped artery if no arterial injury.
  - If arterial damage, revascularization surgery is needed (bypass or interposition grafts with reversed saphenous vein)
- **Surgery for PVES** – fasciotomy with or without musculotendinous section ± popliteal vein reconstruction
- Avoid PTA or stenting as the only treatment

*References*

Cystic Adventitial Disease

- Rare condition in which mucinous cysts form within the adventitia of arteries and veins
- Can lead to lumen occlusion
- Affects young to middle-aged men most often
- Popliteal artery most commonly involved site

J Vasc Surg 2014;60:235-245
Cystic Adventitial Disease

- Incidence 1:1200
- Males > female (4:1)
- Typical age mid 40’s (reported in pediatric patients ages 5-15)
- Sudden onset of intermittent claudication (days to weeks)
- Generally unilateral
- May also present as painless swelling (venous cyst formation)

Intermittent claudication may wax and wane and takes a longer time to subside than ASO
Anatomic locations:

- Popliteal artery (PCAD)
- External iliac artery
- Common femoral artery
- Venous *(saphenous)*
- Radial or ulnar artery
- Axillary artery
- Brachial artery
**Pathogenesis**

- Development of mucoid filled cyst(s) in the adventitial layer
- Luminal narrowing compresses the artery leading to arterial or venous insufficiency

**Theories:**

- **Repetitive Trauma** - leading to chronic degeneration?
- **Systemic disorder** – part of a connective tissue disorder?
- **Ganglion cysts** enlarge and expand into the adventitia?
- **Embryological error** – mucin secreting mesenchymal cells are incorrectly placed in the adventitia?
Cystic Adventitial Disease – History and Physical Examination

• Palpate pulses
• Obliteration of pedal pulse on flexion of the knee (Ishikawa’s sign)
Cystic Adventitial Disease – PVR’s

- ABI with exercise
  - Right ABI rest 1.17
  - Right ABI with exercise 0.34
Ultrasound imaging of the right knee. Sagittal Doppler images confirm a focal stenosis within the popliteal artery (arrow) with post-stenotic turbulent flow, seen as multicolor flow on color Doppler. There is absence of Doppler flow within the hypoechoic structures surrounding the popliteal artery representing the cystic lesions (arrowheads).
Cystic Adventitial Disease - MRI

MRI image shows fusiform mass of high signal intensity at the posterior aspect of the popliteal artery

MRI image shows intramural crescentic Mass of low signal intensity compressing the arterial lumen

Cystic Adventitial Disease - Angiography

Angiogram at rest

Flexion of knee against resistance

Scimitar sign

Smooth, gradually tapering stenosis (Scimitar or Hourglass signs) without poststenotic dilation.

Eur J Vasc Endovasc Surg 2005; 29:93-96
Cystic Adventitial Disease - Treatment

- **Nonsurgical**
  - percutaneous cyst aspiration
  - open cyst aspiration
  - angioplasty – usually unsatisfactory as intima is normal
  - Stent (few case reports)

- **Surgical**
  - evacuate the cyst, resect affected arterial segment and reconstruct with a venous graft or synthetic graft

- **Thrombolytic therapy if artery occluded**
Cystic Adventitial Disease

Attempt at aspiration – usually results in early recurrence

Eur J Vasc Endovasc Surg 2005; 29:93-96
Cystic Adventitial Disease - Endovascular Treatment

CTA – Pre intervention

6 x 40 mm bare-metal, Nitinol self expanding stent

CTA - 24 mos. after reconstruction

Ann Vasc Surg 2013;27;1.e1-1185.e3185
Cystic Adventitial Disease - Surgery

J Vasc Surg 2014;60:235-245
External Iliac Artery Endofibrosis - EIAE

- Affects highly trained athletes (cyclists, endurance runners, rugby players, speed skaters)
- Most common in men – women account < 7% of cases
- Most common (left) external iliac artery (~90%) but also reported in common and internal iliac and femoral arteries
- Unilateral (85%), rarely bilateral (15%)

Helv Chir Acta 1985; 51:793-795
Acta Chir Belg 2004; 104: 635-640
Symptoms:

- Exercise-induced leg pain (thigh) or feeling of a “lack of power”
- Swollen thigh, cramping
- Numbness or paresthesias
- Symptoms relieved with rest
- Results in compromised training, early retirement
External Iliac Artery Endofibrosis

**Physical exam:**
- Usually normal at rest
- May have some muscle wasting in the thigh
- Absent DP or PT pulses are rarely found
- Post exercise (may hear an arterial bruit)
External Iliac Artery Endofibrosis

Complications

• Thrombosis
• Dissection
• Secondary atheroma

• Pathophysiology (anatomic or mechanical factors)
  – arterial kinking with hip flexion exacerbated by psoas muscle hypertrophy and excess arterial length
  – high blood flow causing arterial wall stress and reactive intimal hypertrophy

Vascular and Endovascular Surgery 2011;45:761-768.
External Iliac Artery Endofibrosis

**Diagnosis:**
- ABI before, after maximal exercise
- Use bicycle ergometer
- ABI <0.5 in 85% of individuals
- Other useful tests:
  - Duplex ultrasound may demonstrate artery wall thickening, artery kinking and high flow velocities in the external iliac artery
  - Conventional angiography, MRI
Digital subtraction angiogram demonstrates normal aorta and peripheral run-off vessels

DSA shows left external iliac artery occlusion with collateral flow to the common femoral artery (arrow)
CT scan reveals left psoas muscle (LPM) hypertrophy, external iliac artery occlusion (arrow), and a patent hypogastric artery (dotted arrow).
External Iliac Artery Endofibrosis - Treatment

*Treatment:*

- Conservative = change sports activity but patients often demanding and expect nothing but full recovery
- PTA (recoil an issue)
- PTA with stenting
- Endarterectomy with vein patch
- Resect and bypass
Case Report

- This 35 year old male is seen for ischemic ulcers on his right hand. He denies drug abuse and trauma but has smoked two packs of cigarettes daily for 17 years. He has ……
  a). Atheroemboism
  b). Thoracic outlet syndrome
  c). Thromboangiitis obliterans
  d). A myxoma
Thromboangiitis Obliterans - Buerger’s Disease

- Nonatherosclerotic, segmental inflammatory disease affecting
  - small and medium-sized arteries, veins, and nerves
- Males > females
- Etiology unknown but strong association with tobacco use
- 2/3 have severe periodontal disease

Circulation. 2010;121:1858-1861.
NEJM 2000;343:864-869
Thromboangiitis Obliterans

- More prevalent in the Middle East, Mediterranean, and Asia
- Young tobacco smokers (chewers)
- Reported with cigars, smokeless tobacco
- Claudication of the foot (arch), legs, occasionally arms and hands
- Distal ischemic ulceration, gangrene
- Superficial thrombophlebitis, Raynaud’s phenomenon
- Two or more limbs generally involved
A vasculitis characterized by a highly cellular inflammatory thrombus with sparing of the vessel wall.
Thromboangiitis Obliterans – Diagnosis
History and Physical Exam

Allen test
Thromboangiitis Obliterans - Diagnosis

• Lab tests (exclude other disorders)
  - LAC, ACA, WSR, C-RP, cold agglutinins, cryoglobulins, ANA, anticentromere antibodies, anti-SCL-70, anti-endothelial antibodies

• Echocardiography

• Consistent arteriographic findings:
  segmental occlusions around areas of normal vessels and “corkscrew collaterals”

Circulation 2010; 121: 1858-1861
Thromboangiitis Obliterans - Treatment

- Stop smoking
- Avoid nicotine replacement therapy
- Palliative therapy
- Iloprost infusion
- Calcium channel blockers
- Sympathectomy
- Spinal cord stimulator
- Surgery
- Therapeutic angiogenesis
- Foot and/or hand care

Circulation 2010; 121: 1858-1861
"I put the nicotine patches over his eyes Doctor, and they seem to be working remarkably well"
Young smoker with a compatible clinical presentation

Detailed cardiovascular examination including ankle:brachial indices and Allen test.

Alternative etiology suggested by physical examination or testing?

YES
Evaluate alternative diagnosis

NO
Angiography consistent?

YES
Treat for Thromboangiitis obliterans

NO
Evaluate for other etiology
Suggested Diagnostic Approach to Non-atherosclerotic Arterial Lower Extremity Vascular Disorders

Exercise-induced Leg pain

Iliac artery Distribution

Consider: FMD EIAE

Femoropopliteal Distribution

Consider: PCAD PAES

Foot or arch Distribution

Consider: TAO
Uncommon Arteriopathies

- Segmental Arterial Mediolysis
- Radiation Arteritis
- Pseudoxanthoma elasticum
- Drug-induced
- Drug-abuse
- Ergotism
Segmental Arterial Mediolysis

- Nonatherosclerotic, noninflammatory
- Sudden abdominal pain, distension, massive intra-abdominal hemorrhage, shock
- Dissections, aneurysms (single or multiple), stenoses or occlusion of visceral arteries
- Also affects carotid and vertebral arteries
- Diagnosis: clinical and imaging findings with absence of lab abnormalities or alternative diagnoses
- Treatment supportive, surveillance imaging
- Mortality very high if acute rupture

AJR. 2006;187:1463-1469.
Radiation Arteritis

- Affects major vessels in irradiated fields
  - Cervicocranial stenosis (head and neck cancers)
  - Visceral vessels (lymphoma)
  - Iliac artery (cervical cancer)
- Latency period >10 yrs before symptoms
- Extensive radiation-accelerated ASO
- Stenosis or occlusion of arteries (aneurysm)
- Symptoms similar to ASO
- Angiographic - mimics large vessel vasculitis (tapered narrowing)
- Endovascular techniques vs. surgical
Pseudoxanthoma Elasticum (PXE)

- Rare, autosomal recessive, systemic disease (female-to-male ~ 2:1)
- Xanthomas (pebbly skin) “plucked chicken skin” affects posterior neck, axillae, groin
- Retinal hemorrhage and blindness
- Accelerated atherosclerosis: angina, heart failure due to restrictive cardiomyopathy, HTN, CVA, intermittent claudication
- GI bleeding (avoid antiplatelets, anticoagulants)
- Small to medium-sized arteries more than aorta
- Loss of function mutations in ABCC6 gene
- Genetic counseling, risk factor modification, avoid contact sports

Drug –Induced Arteriopathies

- Clinical manifestations:
  - Vasospasm, digital necrosis, myocardial infarction, Raynaud’s phenomenon, stroke, venous thrombosis, mesenteric ischemia
- Drugs most commonly associated include: sympathomimetic amines (phenylephrine, epinephrine, norepinephrine, high-dose dopamine, methamphetamines, cocaine)
- Inadvertent arterial injection
Drug–Induced Arteriopathies