INTERESTING CASE

Sirinthip Kittivisuit, MD.
Fellowship in pediatric rheumatology, Ramathibodi hospital

A 12-year-old girl

- CC: Blurred vision 1 day

History taking

A 12-year-old girl

15/9/17

- Fever 3 days with headache and vomit
  >> Admit at private hospital
- No neuro deficit, no stiffness of neck
- VA: Rt $20/20^{-1}$, Lt $20/30^{-1}$

- CBC: Hb 13 g/dL, Hct 40%, WBC 11,230/µL, Platelet 150,000/µL
- Dengue: negative

- Try treat as sinusitis:
- Rx: Augmentin IV and Azithromycin

18/9/17

- Bilateral conjunctival injection
- High grade fever
- Headache PS 4/10
- Vomit 1 times/day

- Rx: Ceftriaxone 22-26/9/17
  Clindamycin 22/9/17

A 12-year-old girl

23/9/17

- Severe headache, night awakening pain

- CT brain: normal
- Lumbar puncture:
  OP/CP 17/- cmH_2O,
  WBC 8, RBC 250/HPF,
  protein 37 g/dL, sugar 89 mg%
- CSF culture: no growth

- Add Doxycycline 23-26/9/60
- Dexamethaxone 4 mg IV x 1 dose

25/9/17

- Fever subsite but still had headache
- Discharge 26/9/17

A 12-year-old girl

28/9/17

- Headache
- Dizziness and tinnitus both ears Left > Right side
- Blurred vision, eye pain and photophobia both eyes

>> Admit government hospital

- CBC: Hb 10.9 g/dL, Hct 32%, WBC 15,610/µL, N 76%, L 12%, M 11%, Eo 1%, Platelet 564,000/µL
- ESR 117 mm/h, CRP 150 mg/L
- UA: protein negative, WBC and RBC 0-1/HPF

- Eye exam: Bilateral non-granulomatous anterior uveitis with secondary increase IOP
- No record of VA test result
- Audiogram: Bilateral moderate SNHL
- MRI brain: normal

A 12-year-old girl

28/9/17 — **14/12/17** — **Legs pain**

- Dx: Incomplete Vogt-Koyanagi-Harada syndrome

- Rx: Pulse methylprednisolone 1 gm IV x 3 days (6-8/10/17) then prednisolone (5) 3x2

>> Discharge 10/10/17

>> Refer to Ramathibodi hospital

- Left knee, bilateral ankles pain
- Bilateral thighs and legs pain
- Progression over 2 months with severe pain and night awakening pain within 2 weeks
- Precipitous by more walking distance
- No morning stiffness, no joint swollen/warmth/erythema, no limit ROM

Past history

- No history of eye trauma
- No history of anorexia/weight loss
- No history of photosensitivity rash/hair loss
- No underlying disease
- No history of drug allergy
- Normal development

A 12-year-old girl

Physical examination

Physical examination

- GA: active, cushingoid appearance
- V/S: BT 36.7°C, PR 118/min, RR 20/min
- BP 4 extremities:

131/67 mmHg	131/69
105/48	111/54

- Measurement: Weight 47 kg ($P75^{th}$), Height 156 cm ($P97^{th}$)
- HEENT: no pallor, anicteric sclera, no carotid bruit

Physical examination

- LNs: negative
- Heart: no heave/thrill, normal S1S2, no murmur
- Lungs: clear both lungs
- Abdomen: soft, not tender, no hepatosplenomegaly, no palpable mass, no abdominal bruit
- Extremities: no pallor/ulcer
  Upper extremities pulse 2+ with capillary refill 1 s
  Lower extremities pulse 1+ with capillary refill 2 s

Physical examination

- Neuro signs:
  - Good consciousness
  - Intact CN functions
  - Normal tone, Motor power gr.V all extremities
  - Sensory intact
- DTRs 2+ all extremities
- BBK absent
- Clonus negative

A 12-year-old girl

Problem list

Problem list

- Hypertension with arterial insufficiency: Lower limb claudication with different BP between upper and lower extremities
- Bilateral anterior uveitis
- Bilateral SNHL

A 12-year-old girl

Differential diagnosis

Differential diagnosis

- Takayasu arteritis
- Cogan’s syndrome
- Vogt-Koyanagi-Harada syndrome

A 12-year-old girl

Investigation

Investigation










Complete blood count

- Hb 13.6 g/dL, Hct 43.3%
- WBC 13,300/µL, N 75%, L 15%, M 9%, Eo 1%
- Platelet 260,000/µL

Blood chemistry

- BUN 19, Cr 0.35 mg/dL
- Na 142, K 3.93, Cl 107, HCO_3 22.2 mmol/L
- TB 0.3, DB 0.1 mg/dL
- AST 15, ALT 45, GGT 22 U/L
- Albumin 32.9 g/L

Inflammatory markers

- ESR 8 mm/hr
- CRP < 1 mg/L

Urinalysis

- Sp. gr. 1.019, pH 5, protein negative, blood negative
- WBC 0-1, RBC 0-1 /HPF

Immunology

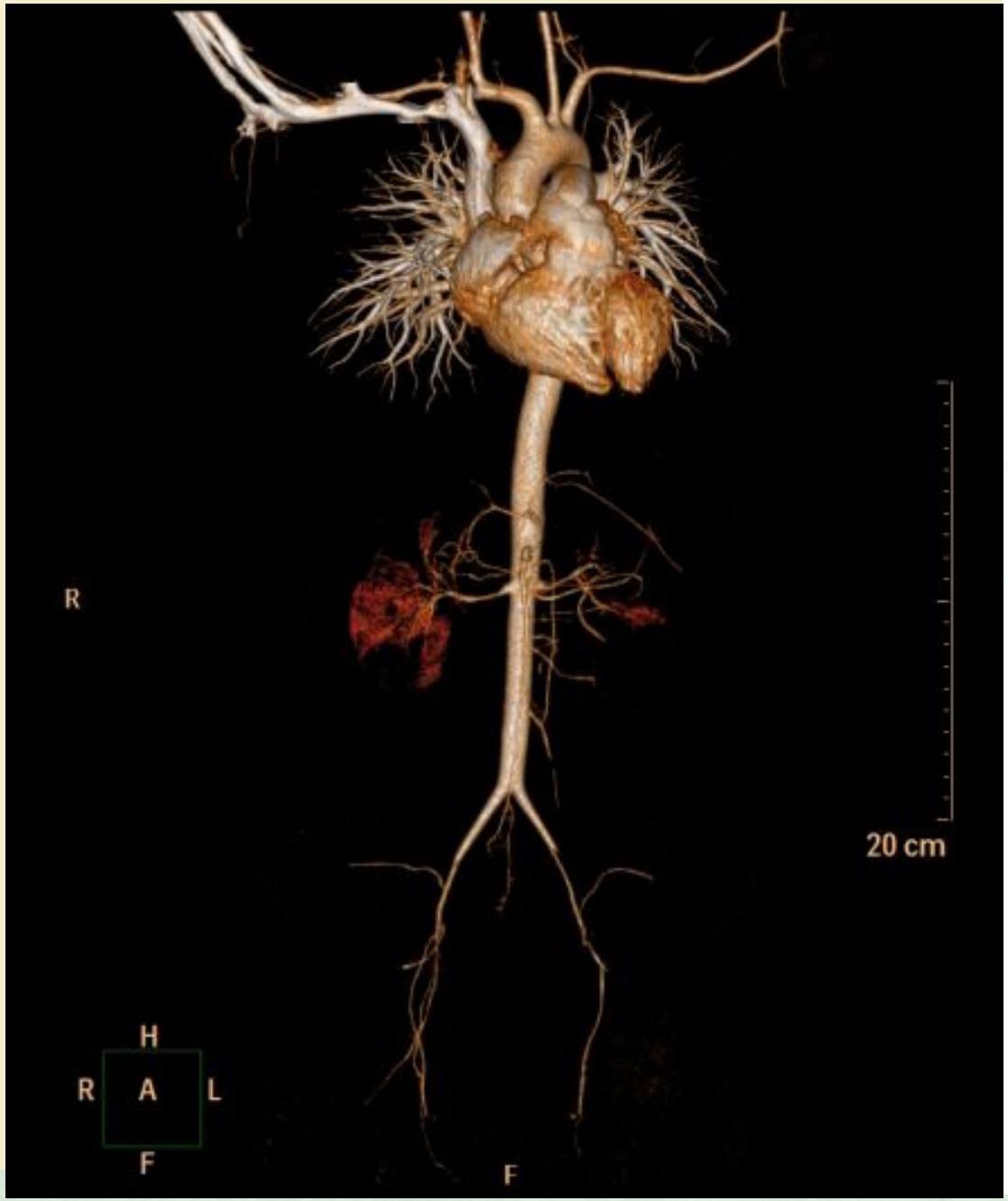
- ANA negative

CTA whole aorta

- Diffuse mild to moderate irregular luminal narrowing involving **celiac trunk**, **SMA**, **mid part of bilateral renal arteries**, **distal part of bilateral common iliac arteries**, **bilateral external iliac arteries**, **left sacral arteries** and **bilateral femoral arteries** included their branches.

CTA whole aorta

- Surrounding soft tissue thickening with delayed mural enhancement at bilateral external iliac arteries with extended to bilateral femoral arteries.
- Suspicious irregularity of **mid part of bilateral subclavian arteries**
- No definite thoracic aortic branches or pulmonary artery involvement.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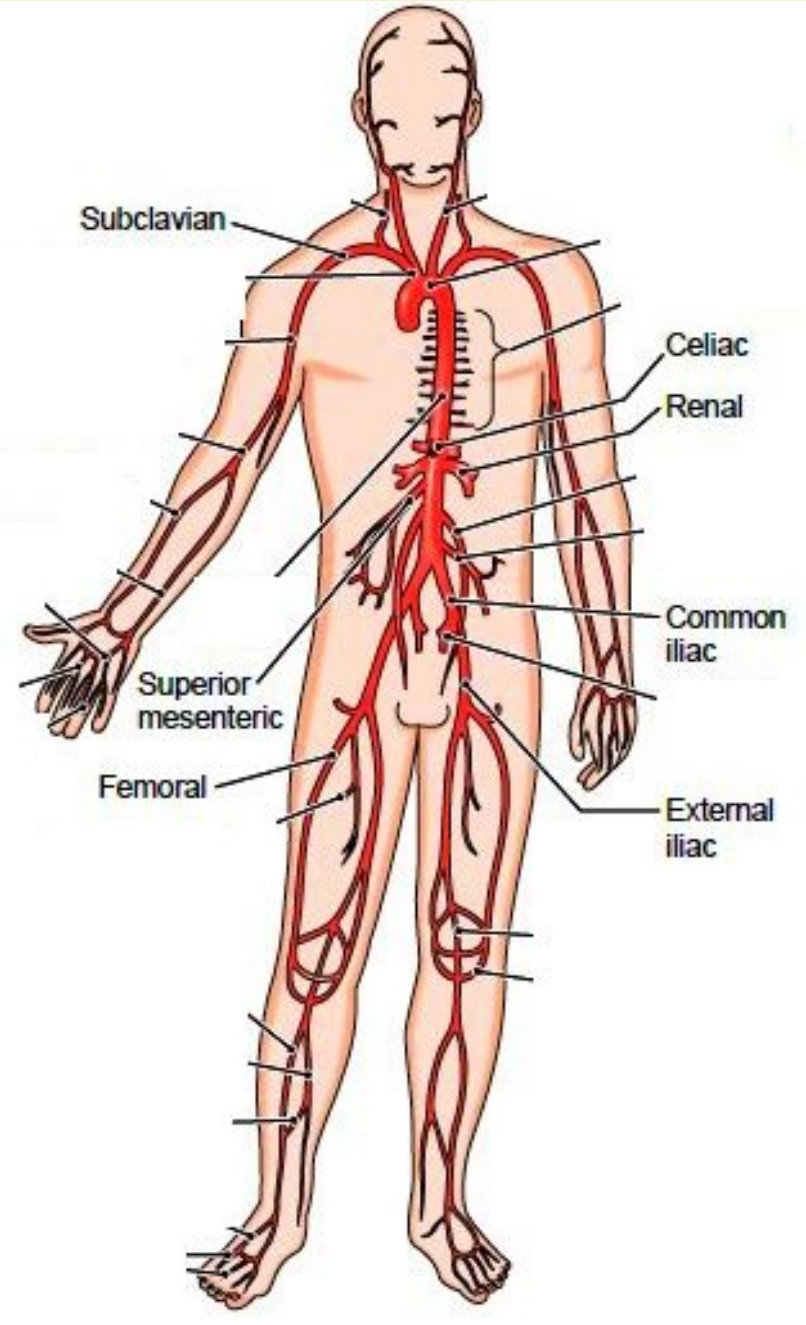
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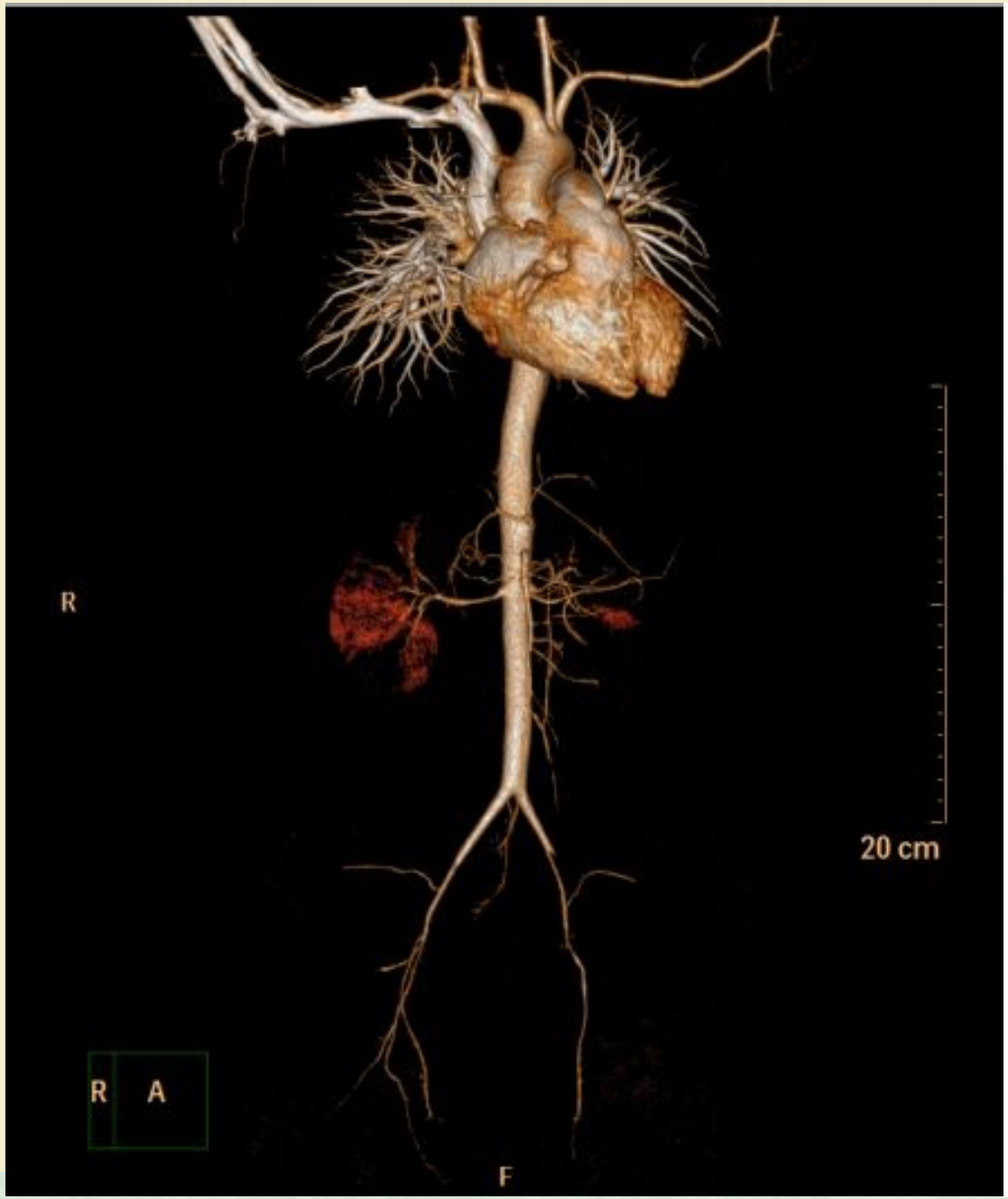
Superior mesenteric
Femoral
External iliac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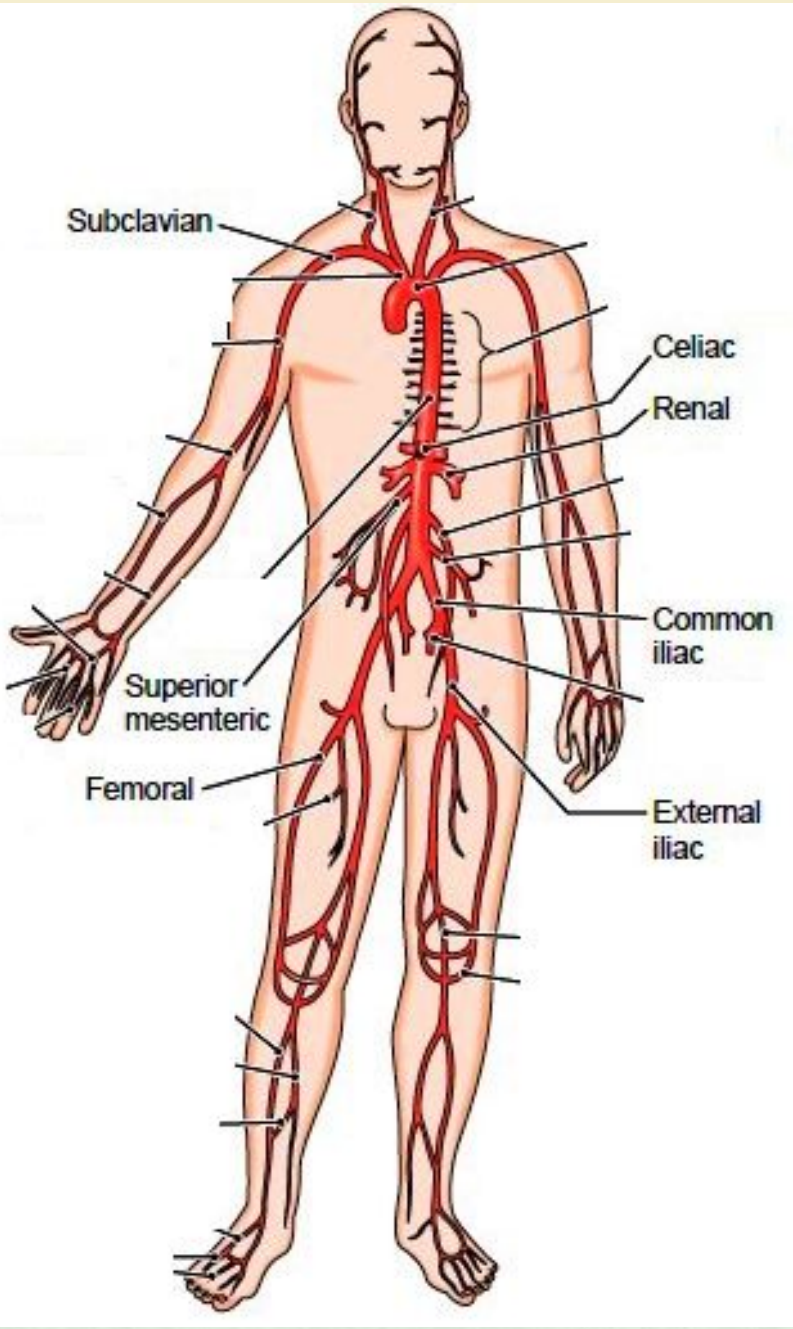
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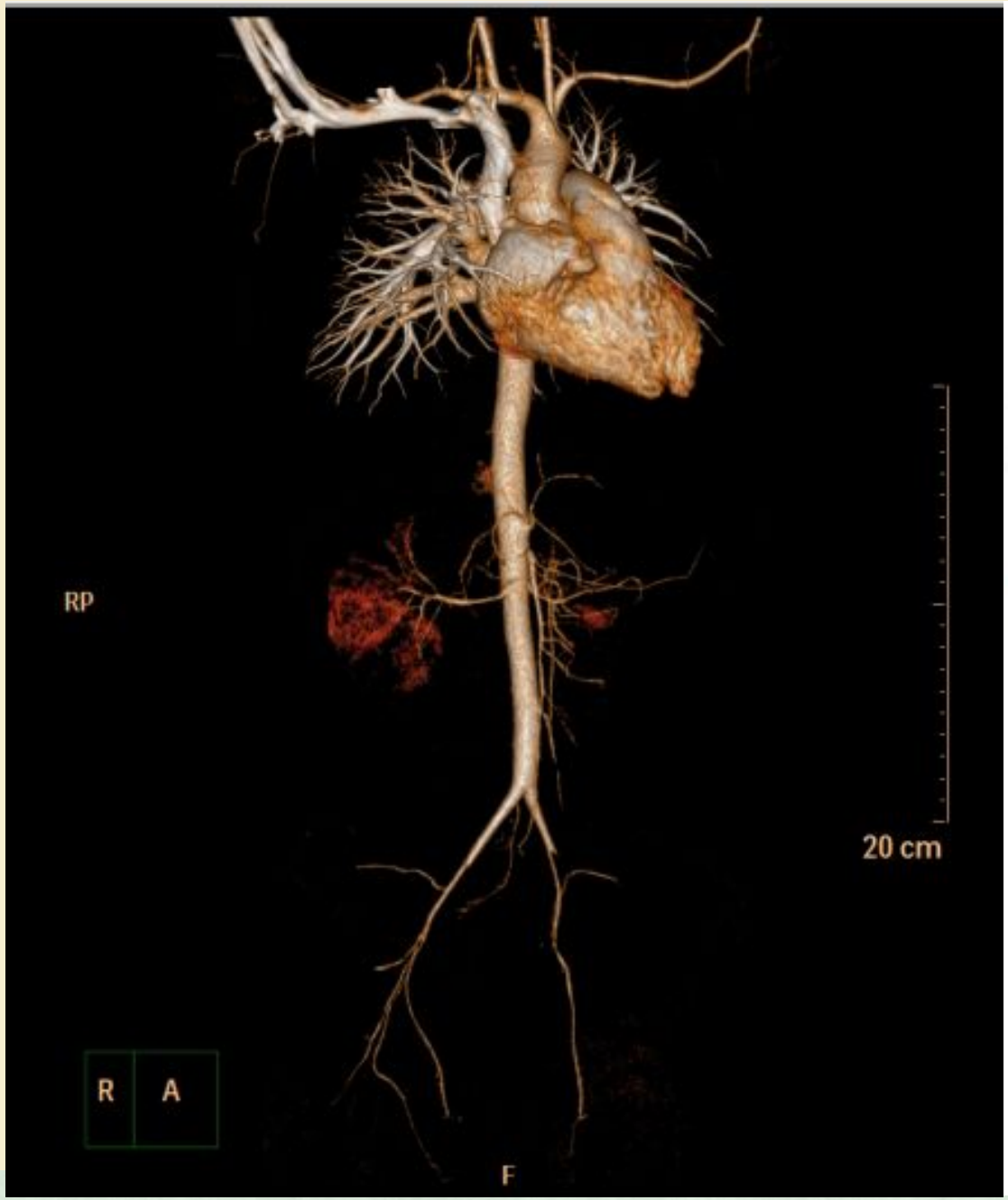
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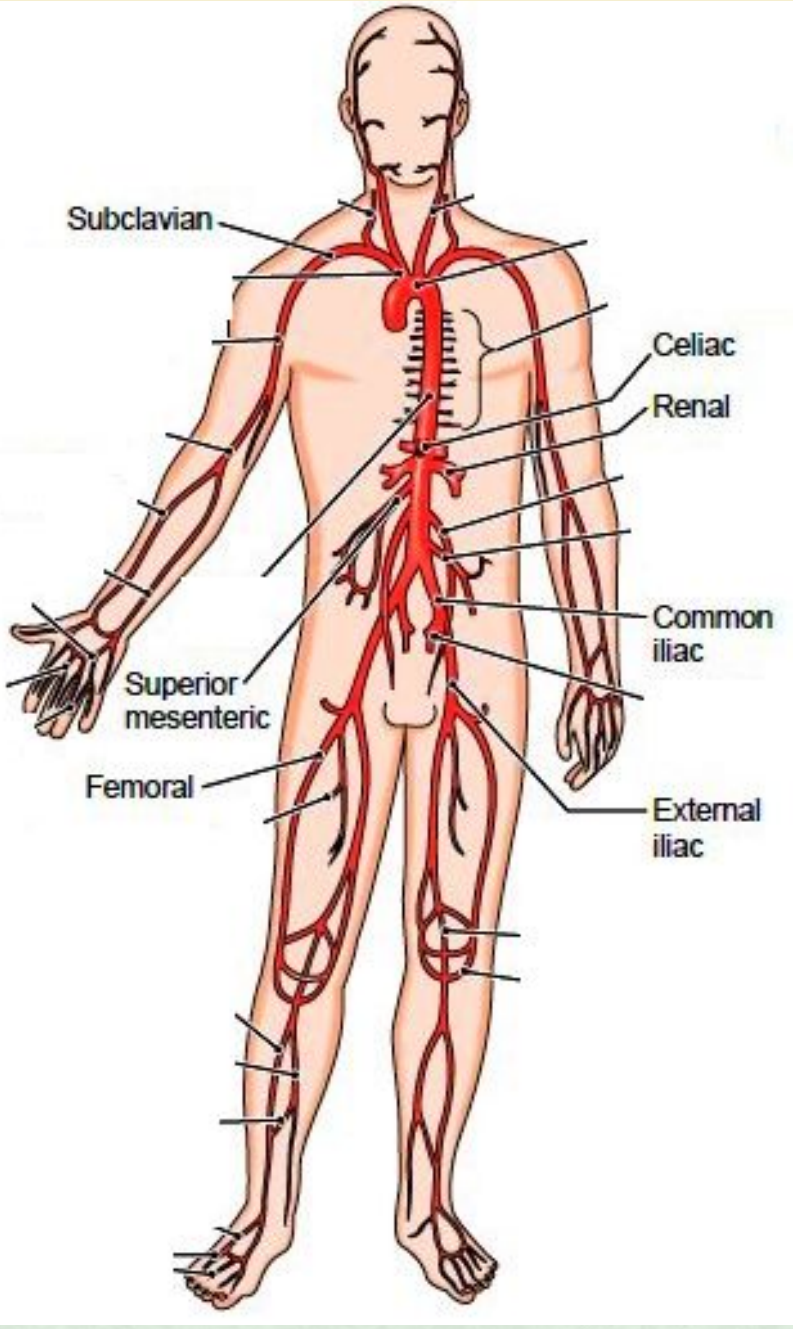
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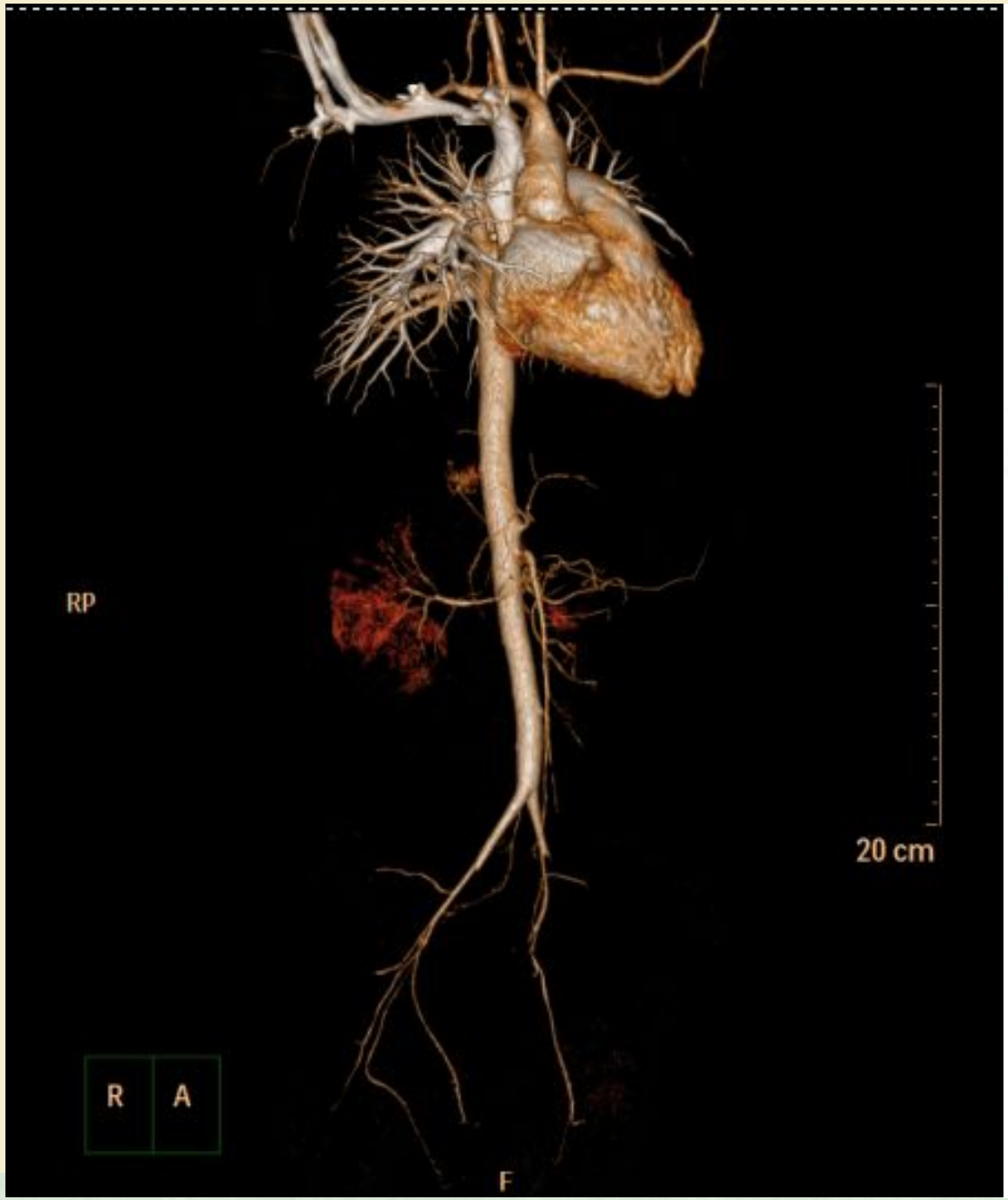
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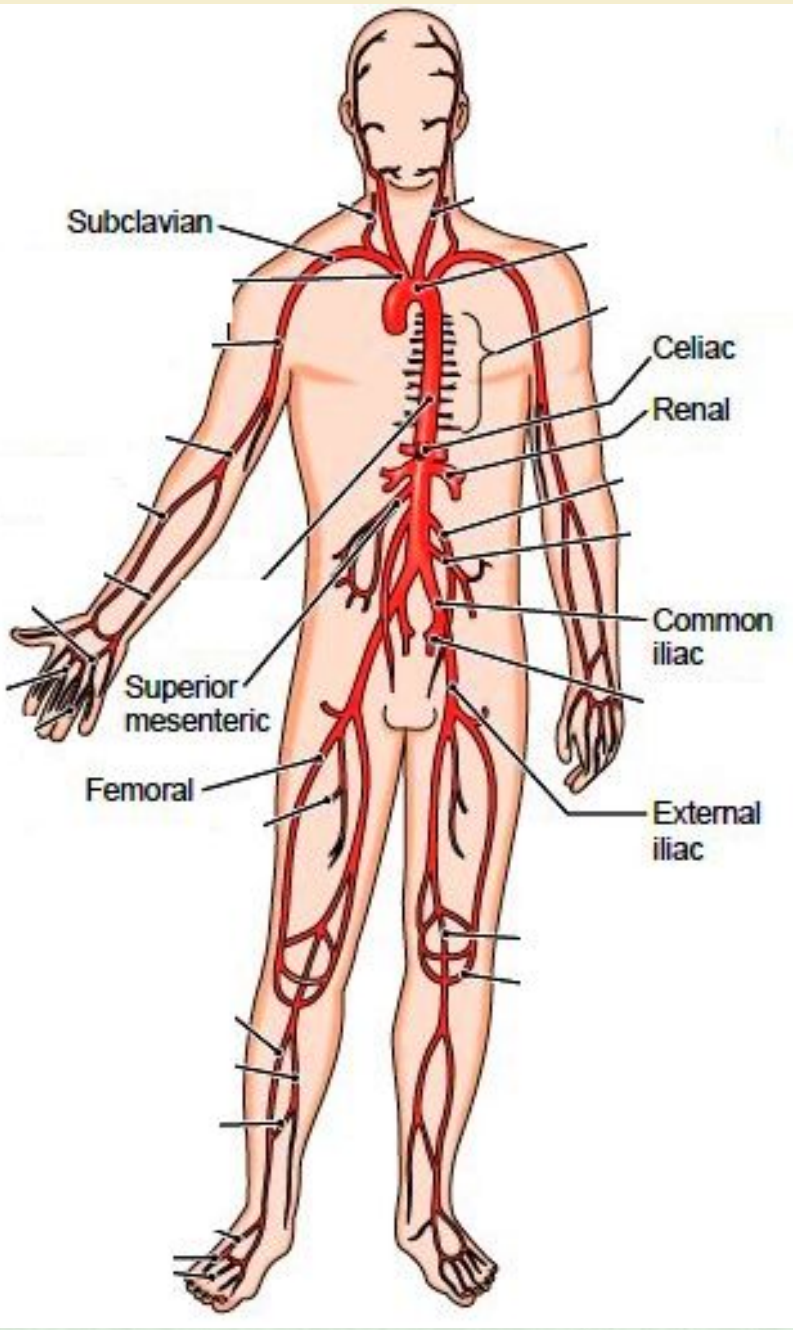
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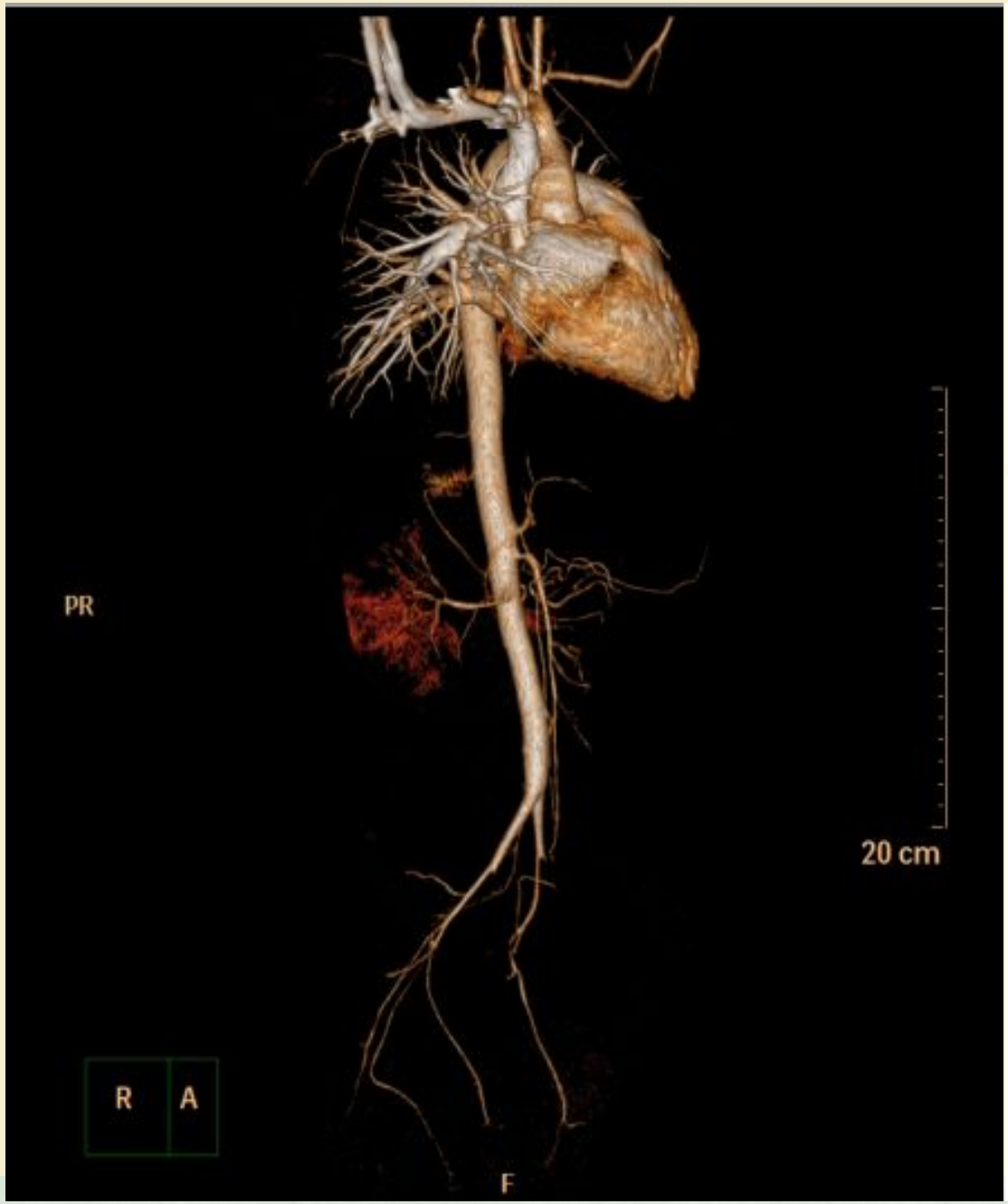
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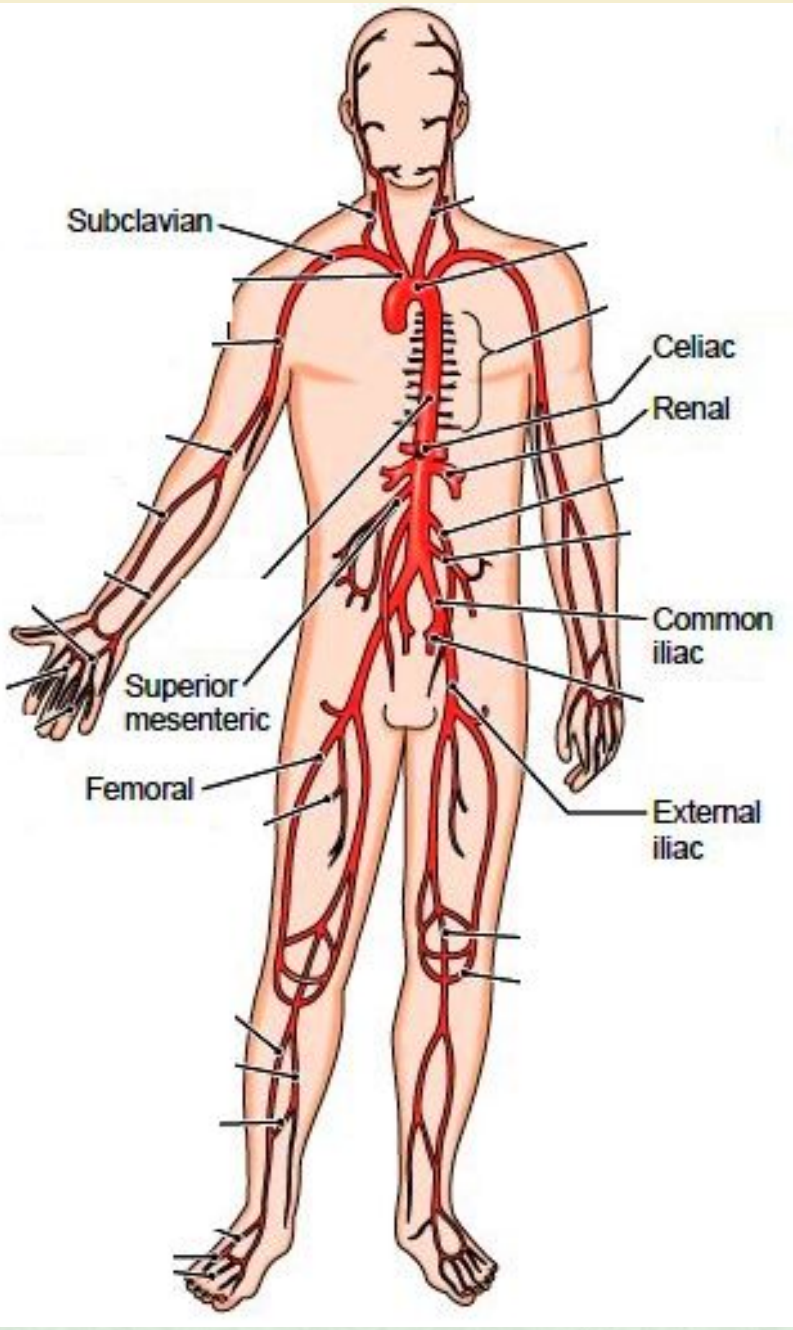
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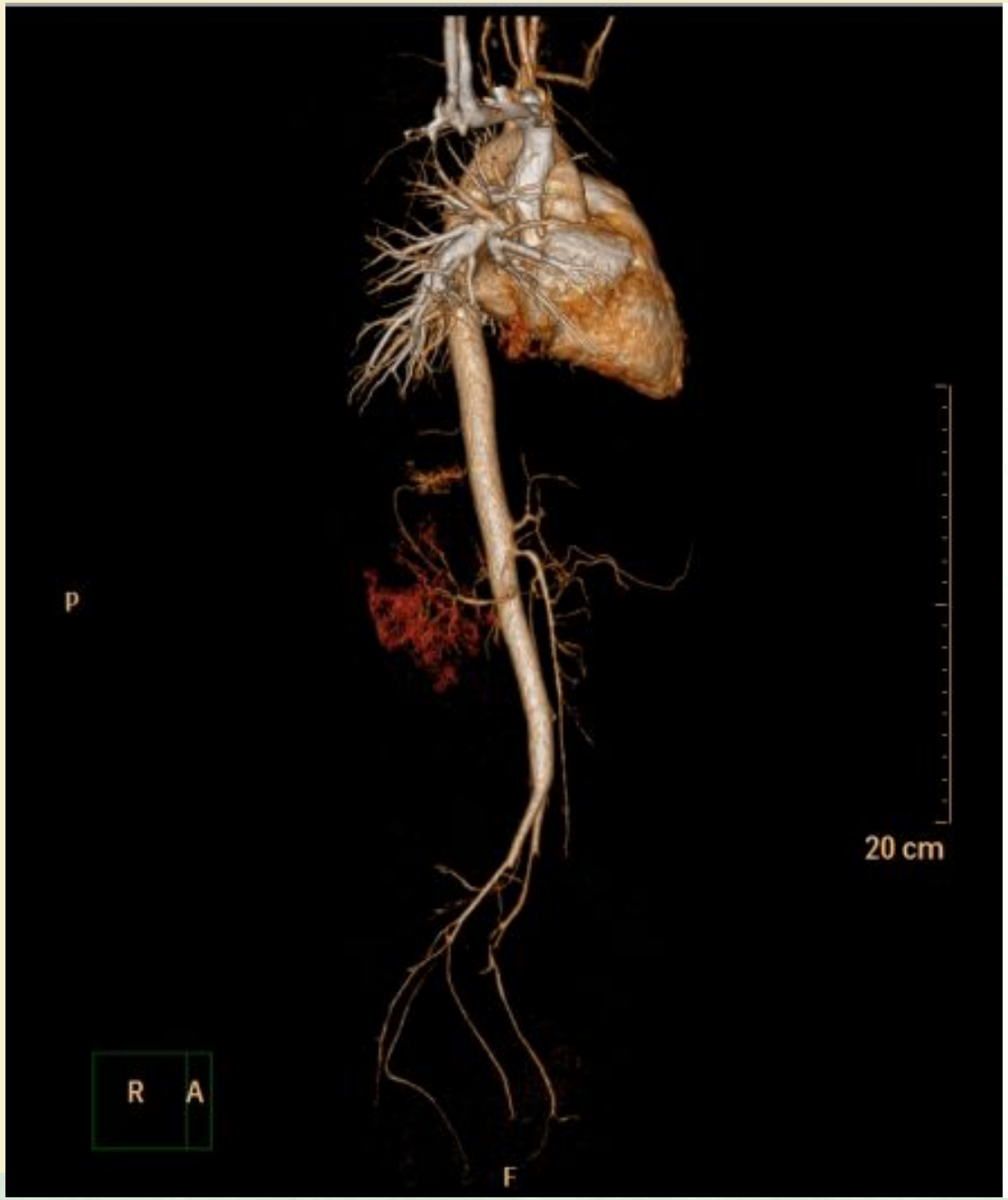
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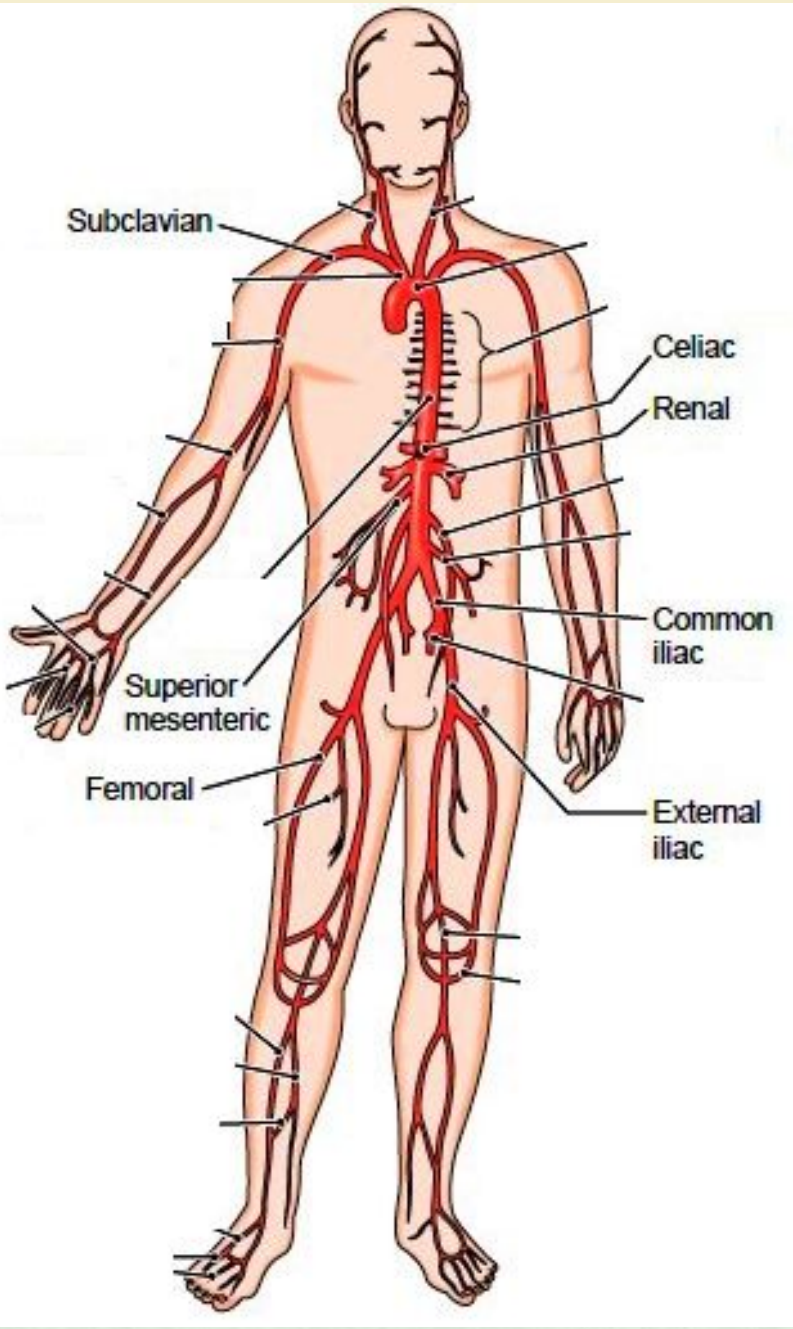
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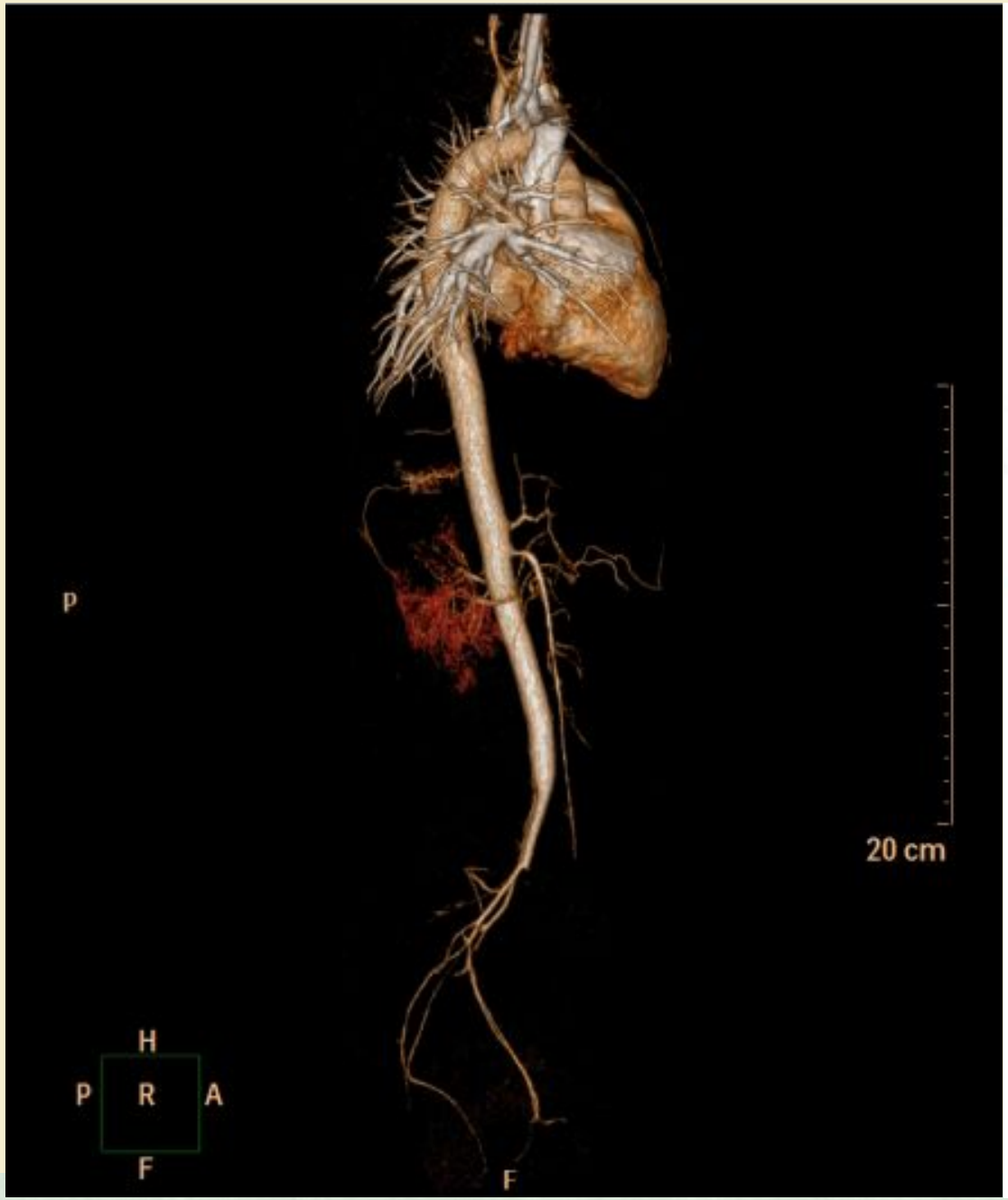
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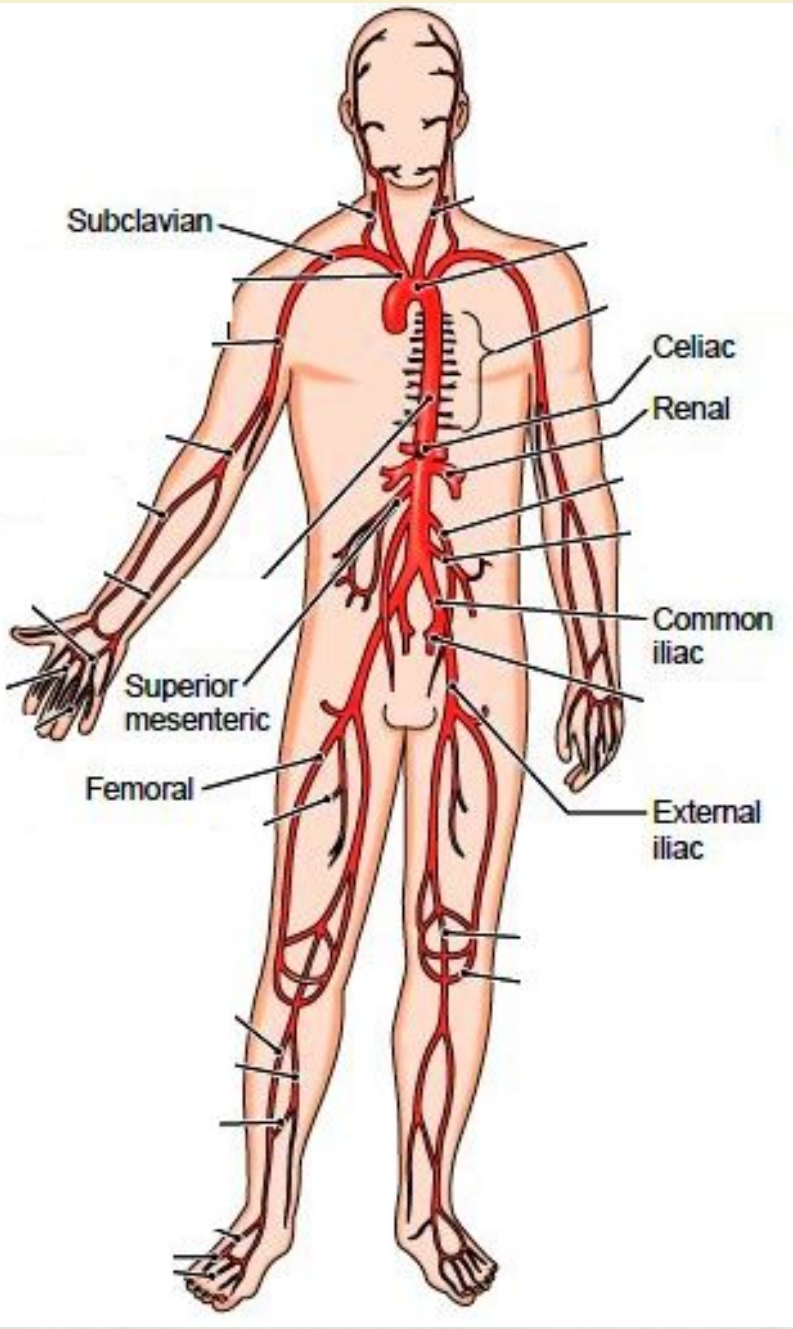
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Electrocardiogram

- Normal sinus rhythm, rate 110/min, normal axis
- No chamber enlargement

Echocardiogram

- Normal cardiac function
- Trivial to mild MR and AR
- No coarctation of aorta or aortic root dilatation

Eye examination

- VA: Rt 20/80, 20/50 with PH
  Lt 20/50, 20/40 with PH
- IOP Rt 15, Lt 11 mmHg (12-22)
- RAPD negative both eyes
- Mutton-fat keratic precipitates both eyes
- Anterior chamber cell 3^{+} both eyes. No vitritis. No retinitis

>> **Granulomatous anterior uveitis** both eyes; compatible with "Incomplete Vogt-Koyanagi-Harada syndrome

Audiogram

- Moderately severe sensorineural hearing loss both ears

A 12-year-old girl

Differential diagnosis

Differential diagnosis

- Takayasu arteritis
- Cogan’s syndrome
- Vogt-Koyanagi-Harada syndrome

Differential diagnosis

	TA	Cogan's	VKH
Limb claudication	/	/	X
Different BP	/	/	X
Headache	/	/	/
Anterior uveitis	Rare	Not common	/
Audiovestibular symptoms	X	/	/
SNHL	Rare	/	/
Elevated ESR and CRP	/	/	X
Imaging	Rare	/	X



Takayasu arteritis (TA)

- Arteritis, often granulomatous
- Predominantly affecting the aorta and/or its major branches

Interesting points

- Diagnosis
- Ocular manifestations in TA
- SNHL in TA
- TA without involvement of aorta

Arthritis Rheum 2013;65:1-11.

EULAR/PRINTO/PRES classification criteria of childhood TA

- Angiographic abnormalities plus 1 of 5 following criteria (sens 100%, spec 99.9%)

1. Pulse deficit or claudication
2. Four limbs blood pressure discrepancy > 10 mmHg
3. Bruit
4. Hypertension >P95th
5. Acute phase reactant

- Angiography (conventional, CT, or MRI) of the aorta or **its main branches** and pulmonary arteries showing aneurysm/ dilatation, narrowing, occlusion or thickened arterial wall not due to fibromuscular dysplasia, or similar causes; changes usually focal or segmental

Ann Rheum Dis 2010;69:798-806.

Ocular manifestations in TA

- Prevalence is varied from 8.1% - 68%

Disease related	Treatment related
Takayasu retinopathy	Increased intraocular pressure
Hypertensive retinopathy	Posterior subcapsular cataract
Ocular ischemic syndrome (OIS)	Ocular infections
	Mydriasis
	Scleral melting
	Eyelid skin atrophy

Retina 2011;31:1170-8.

Ocular manifestations in TA

- Retrospective cohort 78 TA patients, Korea
- Cross-sectional study 61 TA patients, India

Symptoms	Korea	India
Ocular symptoms	44.9%	29.5%
Decrease in vision	29.5%	29.5%
Amaurosis fugax	25.6%	1.6%
Ocular pain	11.5%	0%

Retina 2001;21:132-40.
Retina 2011;31:1170-8.

Ocular manifestations in TA

- Retrospective cohort 78 TA patients, Korea
- Cross-sectional study 61 TA patients, India

Ocular manifestations	Korea	India
Takayasu retinopathy	13.5%	13.1%
Hypertensive retinopathy	30.8%	16.4%
Ocular ischemic syndrome	NA	6.7%
Anterior ischemic optic neuropathy	NA	3.3%
Uveitis	NA	1.6%

Retina 2001;21:132-40.
Retina 2011;31:1170-8.

SNHL in TA

- Few case reports of SNHL associated with TA
- Many reports showed the beneficial effects of steroid but does not always reverse the hearing deficit
- Sometimes progressive and fluctuates during course of treatment, and severe hearing loss may persist in spite of steroid therapy

Laryngoscope 1987;97:797-800.
Br J Rheumatol 1998;37:369-72.
ORL J Otorhinolaryngol Relat Spec 1990;52:86-95.
Intern Med 2005;44:124-8.

SNHL in TA

- The cause of the hearing impairment associated with TA is unknown, may be part of a systemic autoimmune disease for the following reasons:
  (a) there may be elevation of serum immune complexes
  (b) there is an elevation of CRP and ESR preceding deterioration of hearing
  (c) steroid therapy reduces hearing loss as well as disease activity and inflammatory activity

Intern Med 2005;44:124-8.

SNHL in TA

- The mechanisms of the hearing loss in TA are reversible circulatory disturbances due to vasculitis and/or some autoimmune pathogenesis in the inner ear, especially in hair cells
- Fujino et al. proposed the possibility of inner ear dysfunction because of the vasculitis caused by the adhesion of immune complex to the vessel wall

Practica Otologica 1985;78:2313-22.
Intern Med 2005;44:124-8.

TA without involvement of aorta

- Retrospective review 85 CT angiography in TA patient, 1994-2003, Korea
- 95% (81/85) Aortic involvement with or without major aortic branch vessel involvement
- 5% (4/85) Only aortic branch involvement
  - 3/85 Only left subclavian
  - 1/85 Innominate artery, both common carotid artery and superior mesenteric artery

J Vasc Surg 2007;45:906-14.

Cogan's syndrome (CS)

- Characterized by **ocular inflammatory lesions**, including interstitial keratitis, **uveitis**, and episcleritis, and **inner ear disease**, including sensorineural hearing loss and vestibular dysfunction

Interesting points

- Diagnosis
- Ocular manifestations in CS
- Audiovestibular manifestations in CS
- Vasculitic manifestations in CS

Arthritis Rheum 2013;65:1-11.

Typical CS

- Defined by following 3 conditions:

  (1) Ocular symptoms typically an isolated non-syphilitic interstitial keratitis that could be associated with conjunctivitis, conjunctival or subconjunctival bleeding or iritis

- ☑ (2) Audiovestibular symptoms usually progressing to deafness in 1-3 months

- ☑ (3) Interval between the onset of ocular and audiovestibular manifestations of less than 2 year

Arch Ophthalmol 1945;33:144-9.

Atypical CS

- Any of following conditions:

(1) Inflammatory ocular manifestations including episcleritis, scleritis, retinal artery occlusion, choroiditis, retinal hemorrhage, papilloedema, exophthalmos with or without interstitial keratitis; patients with isolated conjunctivitis, subconjunctival hemorrhage or iritis with Ménière's episodes within interval of 2 year

(2) Typical ocular manifestations associated within 2 year with audiovestibular symptoms different from of Ménière-like episodes

(3) Delay of more than 2 year between onset of typical ocular and audiovestibular manifestation

Medicine 1980;59:426-41.

Ocular manifestations in CS

- 80% Interstitial keratitis, mostly bilateral involvement; inflamed small blood vessels invade the adjacent normally avascular corneal stroma
- Target for inflammation is the small vessels in the vascularized layers of the anterior globe: conjunctivitis, episcleritis, scleritis, uveitis
- Retinitis, optic neuritis, glaucoma, papillary edema, cataracts, ocular motor palsy, exophthalmia, central retinal artery occlusion, xerophthalmia, ptosis

Rheumatology 2004;43:1007-15.
Arthritis Rheum 2013;65:1-11.
Autoimmune Rev 2014;13:351-4.

Audiovestibular manifestations in CS

- Sudden onset of hearing loss, vertigo, tinnitus, nausea, vomiting
- Often resolving after several days but followed by progressive hearing loss of variable severity
- Developed at any time during the course of disease
- Hearing loss often bilateral from onset but may be unilateral initially and become bilateral later

Rheumatology 2004;43:1007-15.
Autoimmune Rev 2014;13:351-4.

Vasculitic manifestations in CS

- May include arteritis (affecting small, medium, or large arteries), aortitis, aortic aneurysms, and aortic and mitral valvulitis
- Aortitis with aortic insufficiency occurs 10% of patients
- Indistinguishable from Takayasu arteritis
- 30-50% Systemic symptoms
- Retrospective review 60 CS, 1940-2002, USA:
  40% Headache 35% Arthralgia
  27% Fever 23% Arthritis
  22% Myalgia

Mayo Clin Proc 2006;81:483-8.
Intern Med 2009;48:1093-7.
Autoimmune Rev 2011;11:77-83.
Arthritis Rheum 2013;65:1-11.
Autoimmune Rev 2014;13:351-4.

Vogt-Koyanagi-Harada syndrome (VKH)

- Systemic autoimmune disease; main target is melanin-containing-cells present in the eye, meninges, ear and skin
- Characterized by bilateral chronic diffuse granulomatous uveitis, neurological, audiovestibular and dermatological systems

Interesting points

- Diagnosis
- Ocular manifestations in VKH
- Audiovestibular manifestations in VKH
- Vasculitis in VKH

Autoimmun Rev 2014;13:550-5.
Autoimmun Rev 2016;15:809-19.

Clinical course of VKH

- **Prodromal phase:** Few days prior to ocular symptoms, predominately neurological and auditory manifestations (severe headache, nausea, meningismus, dysacusia, tinnitus, fever, orbital pain, photophobia, pleocytosis of CSF)
- **Acute uveitic phase:** Bilateral vision blurring with choroiditis, vitritis and papillitis which inflammatory cell infiltration into choroid is hallmark. Mutton-fat keratic precipitates may be found
- **Convalescent phase:** Gradual abatement of uveitis; depigmentation of skin, hair, choroid
- **Chronic recurrent phase:** predominately anterior uveitis

Semin Ophthalmol 2005;20:183-90.

Revise diagnostic criteria of VKH

- Complete disease; criteria 1 to 5 must be present
- Incomplete disease; criteria 1 to 3 and either 4 or 5 must be present
- Probable disease (Isolated ocular disease); criteria 1 to 3 must be present

Am J Ophthalmol 2001;131:647-52.

Revise diagno

- (1) No history of penetrating
  onset of uveitis
- (2) No clinical or laboratory
- (3) Bilateral ocular involvem
  the stage of disease)
  a. Early manifestations
  b. Late manifestations
- (4) Neurological/audiotory fi
- (5) Integumentary finding (r
  alopecia or poliosis or vit

Early manifestations:

- Diffuse choroiditis (focal areas of subretinal fluid, bullous serous retinal detachment)
- OR, characteristics fluorescein angiography findings AND echography evidence of diffuse choroidal thickening

Late manifestations:

- History of suggestive of prior uveitis with the above described characteristics
- AND ocular depigmentation
- AND other ocular signs (nummular chorioretinal depigmented scars or recurrent or chronic anterior uveitis)

Am J Ophthalmol 2001;131:647-52.

Ocular manifestations in VKH

- The posterior manifestation is the hallmark, demonstrating vitreous cells with bilateral exudative retinal detachment.
- Some cases might initially present with swollen reddish disc before developing into full-blown exudative retinal detachment.
- The presence of choroidal thickening is common
- The anterior chamber inflammation may present as granulomatous or non-granulomatous

Autoimmun Rev 2016;15:809-19.

Ocular manifestations in VKH

- Signs of depigmentation:
  - Sunset glow: changes of fundus represent choroidal depigmentation showing luminously bright orange-red reflex.
  - Dalen-Fuchs nodule: small discrete atrophic lesions, mainly composed of altered and/or proliferated retinal pigment epithelium (RPE) cells admixed with inflammatory cells at the posterior pole or periphery.
  - Sugiura sign: perilimbal depigmentation

Autoimmun Rev 2016;15:809-19.

Audiovestibular manifestations in VKH

- SNHL (27-50%), tinnitus (34-43%) and vertigo/dizziness (4-25%)
- Typically bilateral and mild HL, mainly in the high frequency range but can cause profound HL (rare)
- May be delayed, fluctuation or progressive HL
- Often appear in prodromal phase or concomitant with active uveitis

Autoimmun Rev 2016;15:809-19.

Vasculitis in VKH

- Case report of a 44-year-old female in Japan, developed VKH after diagnosed aortitis syndrome for 20 years
- Possible association between VKH and aortitis syndrome but may be coincidental

Nippon Ganka Gakkai Zasshi 1966;100:326-31.

A 12-year-old girl

Management

Management

TA	CS	VKH
• Corticosteroid • Immunosuppressive: Cyclophosphamide, MTX, AZA, MMF	• **Mild ocular:** topical steriod and cycloplegic agents	• Corticosteroid • Immunosuppressive: Cyclophosphamide, Cyclosporin A, AZA, Tacrolimus, MMF, MTX • Biologic: Anti-TNF alpha, Rituximab
• Surgery vs Vascular intervention (Balloon angioplasty with or with stent)	• **Inner ear, severe ocular, vasculitis** • Corticosteroid • Immunosuppressive: Cyclophosphamide, MTX, AZA, Cyclosporin A • Biologic: Anti-TNF alpha, Rituximab, Tocilizumab • Cochlear implantation	

J Cliln Pathol 2002;55:481-6.
Autoimmun Rev 2016;15:809-19.
J Multidiscip Healthc 2017;11:1-11.

Management in this patient

1/11/17

- VA: Rt 20/80, 20/50 with PH
  Lt 20/50, 20/40 with PH
- AC cell 2+, mutton-fat BE

- Prednisolone (5) 4x2 [1 MKD]
- MTX (2.5) 5 tab PO weekly
  titrate to 4 tab PO twice a week
  [15 mg/m^2/wk]
- Folic acid (5) 0.5x1
- 1% Pred forte 1 drop BE q 1 h
- 1% Atropine 1 drop BE bid
- 0.5% Glauco-oph 1 drop BE bid

14/12/17

- VA: Rt 20/40^{-1}, Lt 20/25^{-2} with PH
- AC trace, positive KP
- Audiogram: Rt mild, Lt mod SNHL

- Pulse methylprednisolone 1 gm
- Prednisolone (5) 3x2 [0.6 MKD]
- IV Cyclophosphamide [1st dose]
- ASA (81) 1x1, Amlodipine (10) 1x1
- CaCO3, Vitamin D
- 1% Pred forte 1 drop BE qid
- 1% Atropine 1 drop BE OD
- 0.5% Glauco-oph 1 drop BE bid

Management in this patient

11/1/18

- VA: Rt 20/20
  Lt 20/20
- AC clear, IOP 18 mmHg

116/64	113/56
119/54	103/64

- IV Cyclophosphamide [2nd dose]
- Prednisolone (5) 2x2 [0.4 MKD]
- ASA (81) 1x1
- Amlodipine (10) 1x1
- CaCO3, Vitamin D
- Off Pred forte and Atropine eye drop
- 0.5% Glauco-oph 1 drop BE bid

14/2/18

- VA: Rt 20/20
  Lt $20/20^{-3}$
- AC clear

111/59	111/63
96/52	101/59

- IV Cyclophosphamide [3rd dose]
- Prednisolone (5) 3x1 [0.3 MKD]
- ASA (81) 1x1
- Off Amlodipine
- CaCO3, Vitamin D
- 0.5% Glauco-oph 1 drop BE bid
- Plan follow up CTA 12/3/18

Take home message

- **Takayasu arteritis:**
  - Uveitis is uncommon ocular manifestation
  - Isolated branch vessel involvement is possible but less common
- **Cogan's syndrome:** ocular inflammatory lesions, inner ear disease and variable vessel vasculitis
- **Vogt-Koyanagi-Harada syndrome:** bilateral chronic diffuse granulomatous uveitis with neurological, audiotory and integumentary features

Thank you

Rheumatic diseases with reported aortic involvement

Rheumatic diseases with high (10% and more) prevalence of aortic involvement.	Rheumatic diseases with uncommon, but well reported, aortic involvement.	Rheumatic diseases with isolated case reports of aortic involvement
Takayasu's arteritis	Rheumatoid arthritis	Sarcoidosis
Giant cell (temporal) arteritis	Spodyloarthropathies	Wegener's granulomatosis
Long-standing ankylosing spondylitis	Behçet's disease	Polyarteritis nodosa
Cogan's syndrome	Systemic lupus erythematosus	Juvenile Idiopathic Arthritis
Relapsing polychondritis		ANCA-associated Aortitis

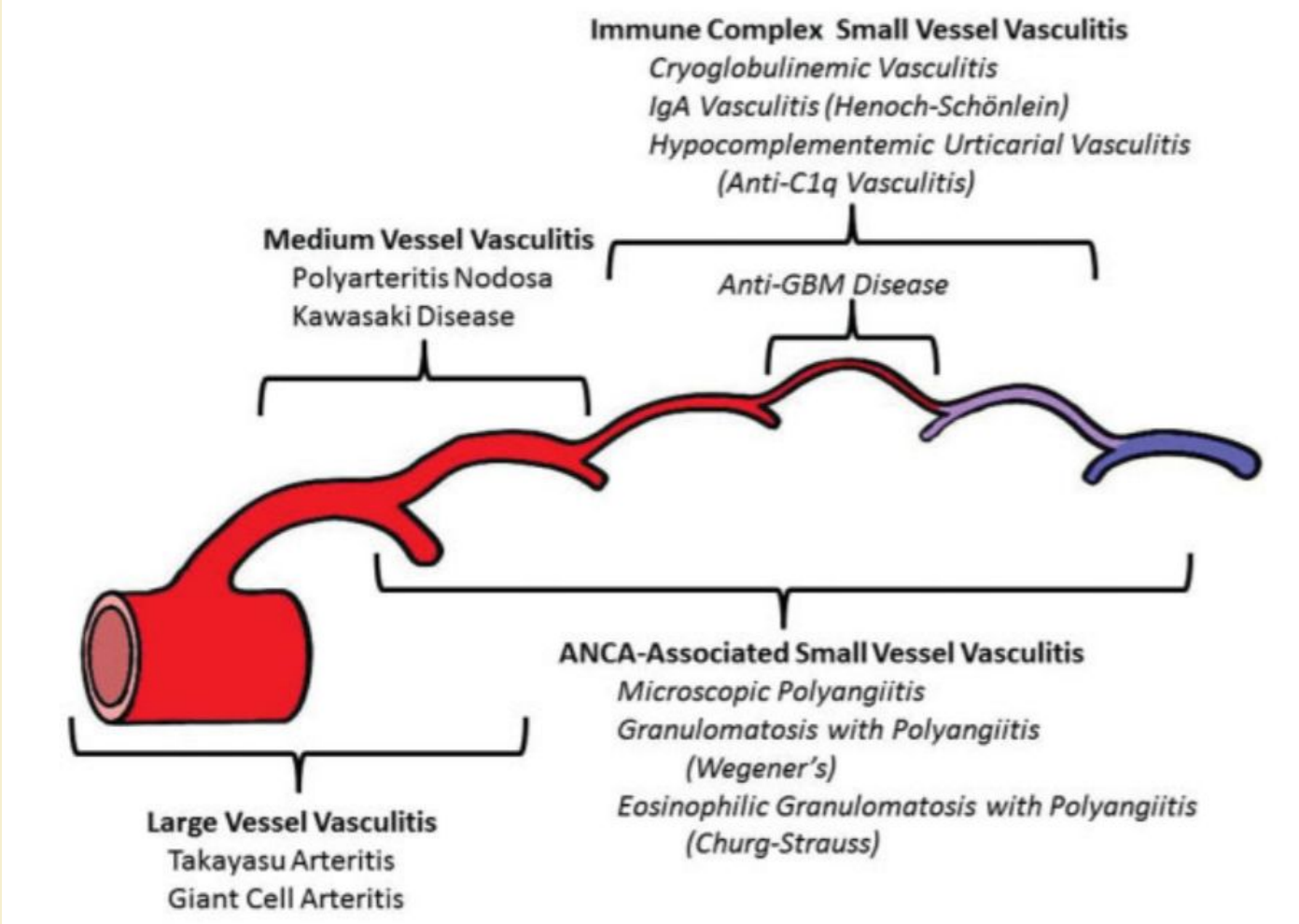
Clin Exp Rheumatol 2006;24:S41-7.

Vasculitis

- Inflammation of blood vessel walls
- The inflammatory infiltrate may be one that is predominantly neutrophilic, eosinophilic, or mononuclear
- Variable features can be used for categorization:
  - Etiology
  - Pathogenesis
  - Type of vessel affected
  - Type of inflammation
  - Favored organ
  - Clinical manifestations
  - Genetic predispositions
  - Demographic characteristics

Arthritis Rheum 2013;65:1-11.

Vasculitis



Arthritis Rheum 2013;65:1-11.

Vasculitis

Types	Diseases
Large vessel vasculitis (LVV)	Takayasu arteritis (TA) Giant cell arteritis (GCA)
Medium vessel vasculitis (MVV)	Polyarteritis nodosa (PAN) Kawasaki disease (KD)
Small vessel vasculitis (SVV)	ANCA associated vasculitis: GPA, EGPA, MPA Immune complex SVV: Anti-GBM disease, Cryoglobulinemic vasculitis, IgA vasculitis, anti C1q vasculitis
Variable vessel vasculitis	Behçet's disease (BD) Cogan's syndrome (CS)

Arthritis Rheum 2013;65:1-11.

Vasculitis

Types	Diseases
Single organ vasculitis (SOV)	Cutaneous leukocytoclastic angiitis Cutaneous arteritis Primary central nervous system vasculitis Isolated aortitis Others

Arthritis Rheum 2013;65:1-11.

Vasculitis

Types	Diseases
Vasculitis associated with systemic disease	Lupus vasculitis Rheumatoid vasculitis Sarcoid vasculitis Others

Arthritis Rheum 2013;65:1-11.

Vasculitis

Types	Diseases
Vasculitis associated with probable etiology	HCV-associated cryoglobulinemic vasculitis HBV-associated vasculitis Syphilis-associated aortitis Drug-associated immune complex vasculitis Drug-associated ANCA-associated vasculitis Cancer-associated vasculitis Others

Arthritis Rheum 2013;65:1-11.

Aortitis

- Pathological term for inflammation of the aortic wall
- The classification of aortitis broadly includes underlying rheumatologic and infectious diseases, along with isolated aortitis

Circulation 2008;117:3039-51.

Clinical presentation

- Asymptomatic
- General syndrome: fever, malaise, weight loss, high ESR
- Pain (chest, back, abdominal): acute typical pain of aortic dissection, vague or nonspecific recurrent pain
- Aortic valve incompetence: aortic root dilatation, direct involvement
- Ischemic symptoms: coronary ischemia, abdominal ischemia, limb claudication
- Embolic phenomena

Clin Exp Rheumatol 2006;24:S41-7.

Causes of aortitis

- **Inflammatory:**
  - Large vessel vasculitis: TAK, GCA
  - Rheumatoid arthritis
  - Systemic lupus erythematosus
  - HLA-B27 associated spondyloarthropathies
  - Sarcoidosis
  - Other vasculitides: ANCA-associated vasculitis, Beçhet's disease, Cogan's syndrome, Relapsing polychondritis

Circulation 2008;117:3039-51.

Causes of aortitis

- **Isolated aortitis:**
  - Isolated idiopathic (thoracic aortitis)
  - Chronic periaortitis: Idiopathic retroperitoneal fibrosis, Inflammatory abdominal aortic aneurysm, Perianeurysmal aortitis, Idiopathic abdominal periaortitis

Circulation 2008;117:3039-51.

Causes of aortitis

- **Infectious:**
  - Bacteria: Salmonella spp., Staphylococcus spp., Streptococcus pneumoniae, other
  - Syphilis
  - Mycobacterium
  - Other

Circulation 2008;117:3039-51.

Laboratory testing

- Markers of inflammation: ESR, CRP
- Complete blood count
- Kidney and liver function
- Additional laboratory testing based on differential diagnosis: ANA, c-ANCA, p-ANCA, RF

Circulation 2008;117:3039-51.

Imaging modalities

Modalities	Advantages	Disadvantages
CT	Lesion extent Accurate cross-sectional imaging in demonstrating vessel wall change Evaluation of other organs which was helpful for differential diagnosis Excellent high spatial resolution Multiple planes and three-dimensional views	Exposure to ionizing radiation Contrast media
US	Lack of exposure to ionizing radiation	Limited field of view Operator dependence
MRI	Demonstration of early wall thickening even before luminal narrowing occurs	High cost Long scan time Difficult in detecting calcification in vessel wall
PET	Evaluation of metabolism (activity evaluation)	Poor spatial resolution Less information of anatomy

CT = Computed tomography, MRI = magnetic resonance imaging, PET = Positron emission tomography, US = Ultrasonography

Korean J Radiol 2017;18:786-98.

ACR classification criteria of TA

- ≥ 3 of 6 criteria (sens 90.5%, spec 97.8%)

1. Age at disease onset < 40 yr
2. Claudication of extremities
3. Decreased brachial artery p
4. Blood pressure difference >
5. Bruit over subclavian arteri
6. Arteriogram abnormality

- Arteriographic narrowing or occlusion of the entire aorta, its primary brances or
- Large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or silmilar causes; changes usually focal or segmental

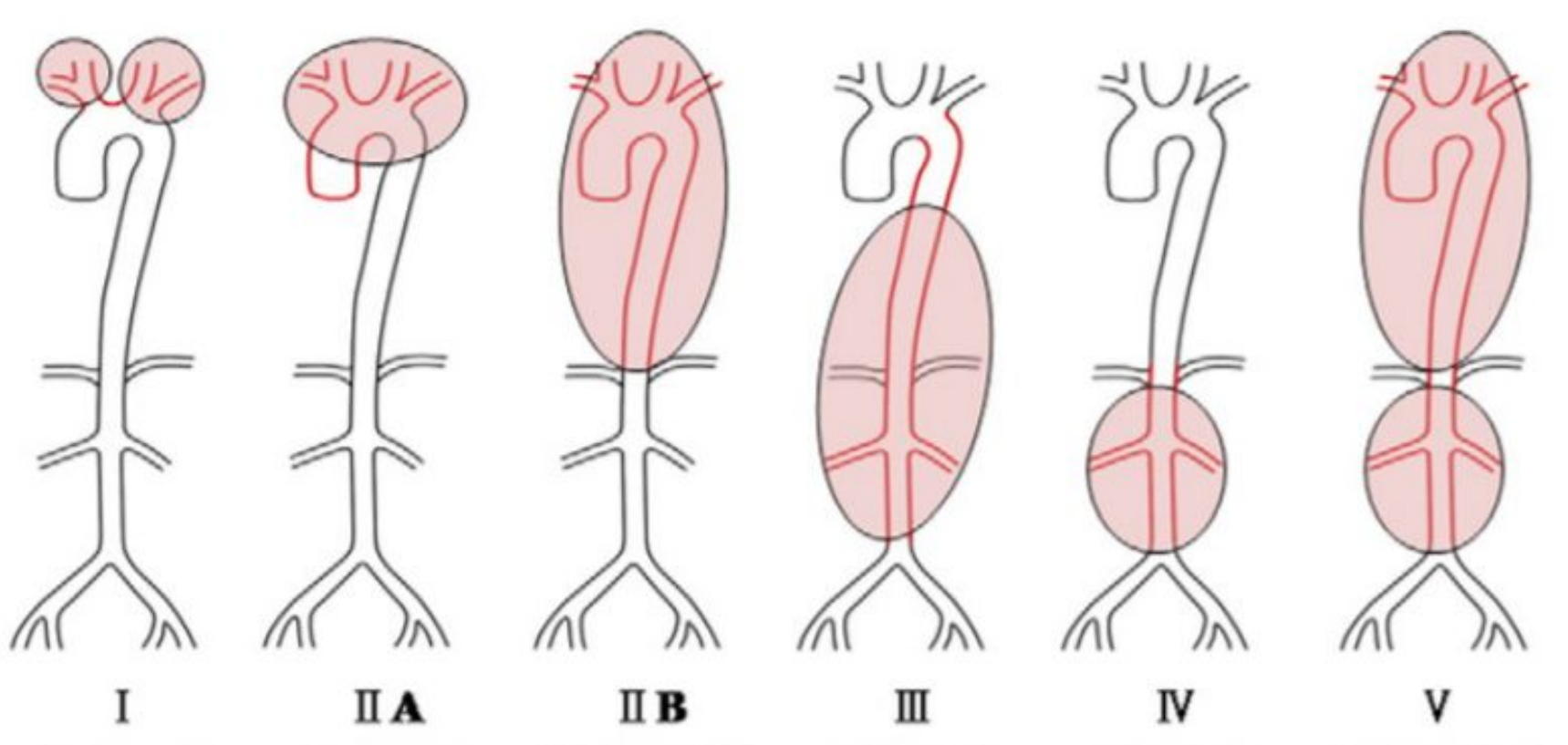
Arthritis Rheum 1990;33:1129-34.

Takayasu arteritis (TA)

- Incidence 0.4-1 case/1,000,000/year
- Onset usually occurs before the age of 50 years, which is a major distinction from giant cell arteritis, whose onset usually occurs after age 50
- Average age of diagnosis 25-30 years
- 75-97% of patients are female

Curr Rheumatol Rep 2005;7:270-5.
Circulation 2008;117:3039-51.
Arthritis Rheum 2013;65:1-11.

Angiographic classification of TA from the Takayasu conference 1994



Int J Cardiol 2012;159:14-20.

	Ocular causes	
	Anterior Segment of the eye	Posterior segment of the eye
Disease related	Neovascular glaucoma (6) Uveitis (7) Cataract Sclerokeratitis (8)	Amaurosis fugax (1) Central or branch retinal artery occlusion (2, 3) Central or branch retinal vein occlusion (4) Anterior ischaemic optic neuropathy (5) Vitreous haemorrhage Ocular ischaemia (1) Macular oedema (1) Exudative retinal detachment (5)
Treatment related	Steroid induced Cataract (1) Steroid induced Glaucoma (1)	Glaucomatous optic neuropathy (1) Steroid induced central serous retinopathy
	Extra-ocular causes	
	Anterior circulation	Posterior circulation
Disease related	Anterior circulation stroke*	Posterior circulation stroke†
Treatment related	Reperfusion injury	

J Clin Diagn Res 2014;8:MD06-7.