VASCULITIS

TAKAYUSU'S ARTERITIS

Inflammation of **aorta, branches, pulmonary arteries**
90% female; age 10-30
(a)**Acute systemic phase** (pre-pulseless): non-specific inflammation (constitutional symptoms)
(b)**Chronic obliterative phase** (pulseless): claudication/pulse inequality/bruits/BP inequality
Investigations: ESR/CE-MRA/biopsy: **patchy granulomatous inflamm**, intimal profil, fibrosis of adventitia/media
Management: **methotrexate**/myco mof/AZA steroids; **angio** (short lesions); **vein bypass** (long lesions)

BUERGER's DISEASE (THROMBANGITIS OBLITERANS)

Segmental thrombotic occlusion of *medium/small-sized arteries*
Young men (age increasing, women increasing due to smoking)
Smoking is main risk factor
Acute hypercellular thrombosis: lymphocytes/fibroblasts/giant cells + thrombosed lumen
(IEL intact, no wall necrosis, aneurysm, calcification or atherosclerosis)
Symptoms: **lower limb rest pain** (claudication rare, limited to foot)
Signs: (a)**tissue loss** (ulcer/gangrene) (b)**absent pedal pulses** (fem/pop++)
Angio: normal to pop; sudden **occlusion at tibial/peroneal** level
Management: **smoking cessation**

GIANT CELL ARTERITIS/TEMPORAL ARTERITIS

Systemic granulomatous vasculitis affecting branches of aorta *(large/medium-sized arteries)*
Age >50; 17/million/yr of over 50s; **Female** 5x
Risks: **smoking** and **atherosclerosis**

Symptoms:
Headache (generalised or localised)
Sudden blindness (optic and ciliary artery)
Jaw claudication (facial and maxillary artery)
Tongue claudication (lingual artery?)

Signs:
Visual defect
Scalp tenderness (sup. temp/occ art.)
Temporal artery thick/tender/absent pulse

Diagnosis: **ACR Criteria** (any **3 of 5**) have sensitivity 93.5% and specificity 91.2%
Age>50, new localised headache, tender/pulseness temporal art, ESR>50, abnormal bx (granulomatous inflamm/mononuclear infiltration)

Biopsy: **granulomatous** inflammation/**mononuclear** infiltration (50% negative due to skip lesions)
Management: **methotrexate/steroids**

WEGENER's GRANULOMATOSIS

Systemic necrotizing vasculitis of *small/medium-sized arteries*
Triad: URT + lung + kidneys
Cutaneous features: palpable purpura, ulcers, subcutaneous nodules
Bloods: **ESR/c-ANCA**
Imaging: CT shows **mucosal thickening, sinus opacification**, air-fluid levels
CXR: **mass lesions** in chest
Biopsy:
Management: **immunosuppressants**

POLYARTERITIS NODOSA (PAN)

Systemic necrotizing vasculitis of *small/medium-sized arteries*
Age **40-60**; **men** 2x; **HepB** endemic areas
Renal failure in 70%; **HTN**; **aneurysm**; **mononeuritis multiplex**
Symptoms: constitutionals/GI upset/symptoms related to features
Signs: nail fold infarct, digital infarct, palpable purpura, livedo reticularis
Labs: anaemia, ESR, **c-ANCA**, HepB and **proteinuria**
Angio: saccular/fusiform **aneurysms**
Biopsy:
Management: **steroids** +/- **cyclophosphamide**

CUTANEOUS VASCULITIS

Mainly venules (can be capillaries/arterioles)

IDIOPATHIC CUTANEOUS VASCULITIS:
Palpable purura below know; symmetrical and worse when sitting
Diff dx: urticaria (ICV lasts >24hrs)
Bx: leucoclastic vasculitis, VEC swelling, necrosis and haemorrhage, fibrin deposition, PMNs

NECROTIZIGING VASCILITIS
Hypersensitivity to – infection (URTI → HSP)
-drugs (antibiotics, diuretics, NSAIDs)
-systemic disease (associated with immunological disorders)

CUTANEOUS VASCULITIS IN SYSTEMIC DISEASE
Small vessel vasculitis affecting skin (esp in CTDs/cyroglobulinaemia)
Churg-Strauss/Behcet's
Biopsy and investigations targeted at aetiology
Management: NSAIDs/steroids/antihistamines