VASCULAR TUMOURS

- benign
  - infantile haemangiomas
  - congenital haemangiomas (RICH, NICH and PICH)
  - tufted angioma (with or without Kasabach Merritt syndrome)
  - spindle cell haemangioma (heamangioendothelioma)
  - epithelioid haemangioma
  - pyogenic granuloma

- locally aggressive or borderline
  - Kaposiform haemangioendothelioma (with or without Kasabach Merritt syndrome)
  - retiform haemangioendothelioma
  - composite haemangioendothelioma
  - papillary intralymphatic angioendothelioma (Dabska tumour)
  - Kaposi sarcoma

- malignant
  - angiosarcoma
  - epithelioid haemangioendothelioma

- others
  - polymorphous haemangioendothelioma
  - targetoid haemangioma
  - glomeruloid haemangioma
  - microvenular haemangioma

1. INFANTILE HAEMANGIOMA

Commonest soft tissue tumour of childhood;
Not present at birth, clinically apparent later; involute
Most=focal nodules; Others=plaques if segmental

NODULAR HAEMANGIOMATA:
Stage 1: Rapid proliferation (first 6 months)
-Superficial dermis: strawberry naevus (bright red, lobulated)
-Deep dermis: swelling with blueness or no discolouration
Stage 2: Slow growth until 12 months
Stage 3: Involution (50% by 5 yrs/70% by 7 yrs/90% by 9 yrs) leaves residual skin changes

SEGMENTAL HAEMANGIOMATA: Plaque-like involving larger area of skin
SUBGLOTTIC: neck and beard area; follow for 12-16 weeks as 60% risk of airway haemangioma
PAROTID: pre-auricular plus parotid gland haemangiomata
PHACE: *posterior fossa, haemangiomata, arterial anomalies, coarctation of aorta, eye abnormalities*
Segmental haemangiomata on face (upper/forehead) in almost exclusively females

Diagnosis:
Clinical history and examination
Imaging for underlying abnormalities (MRI for neuro, ECHO or angio for cardiac)
US: well defined, echogenic, highly vascular, larger central feeding arteries and draining veins
MRI: well lobulated tumour isotense/hypotense vs muscle on T1, hyperintense on T2
Biopsy rarely required; express GLUT-1

Management:
Indications: disfigurement or mass effect (visual axis/airway)
B-blockers first line; particle embolization to reduce vascularity until involutes (if large w/HOCF)
Surgery: early in proliferative phase if mass effect; later if involuted for cosmesis

2. CONGENITAL HAEMANGIOMA

Fully formed at birth and involute rapidly (RICH), never (NICH) or partially (PICH)
Difference to IH: natural history as above, immunohistochemically, histologically (no GLUT-1)
Coagulopathy when large

3. PYOGENIC GRANULOMA ie lobular capillary haemangioma

Second commonest vascular tumour
Head and neck; red papule; grows rapidly, ulcerates and bleeds

4. TUFTED ANGIOMA

Neck or upper trunk; red or purple patch; slow-growing and may resolve
GLUT-1

5. KAPOSIFORM HAEMANGIOENDOTHELIOMA (KSE)

Trunk, shoulder, thigh; reddish discolouration
GLUT-1

**TA and KSE associated with Kasabach-Merritt phenomenon (severe coagulopathy, thrombocytopaenia, variety of soft tissue lesions)