Myogenin
Case 23

- Alveolar rhabdomyosarcoma
Alveolar rhabdomyosarcoma

- Generally older children than ERMS
- Less frequent than ERMS (20% of RMS)
- Most commonly arises in **extremities**
- Often high stage at presentation

**Histopathology:**
- Monomorphous round nuclei
- Alveolar / discohesive growth pattern
- Wreath-like multinucleate cells
- Rhabdomyoblasts
- NB SOLID VARIANT (no alveolar pattern)
- **IHC: myogenin diffuse/strong**
Alveolar rhabdomyosarcoma

- **Genetics:**
  - $t(2;13)(q35;q14) - PAX3$
  - $t(1;13)(q36;q14) - PAX7$  \[\text{Fusion with FOXO1}\]

- **Behaviour**
  - Higher grade, more aggressive than ERMS
Case 24

- Newborn male, mass over thenar eminence
Case 24

- Infantile (congenital) fibrosarcoma
Infantile fibrosarcoma

- Histology:
  - Looks the same as adult fibrosarcoma
  - Primitive spindle/round cells
  - Lymphocytes/EMH
  - Herringbone pattern, cords, sheets
  - Necrosis/haemorrhage/calcification
  - Infiltrative growth
Infantile fibrosarcoma

- Nearly all in first year of life
- Congenital in many cases

- Characterised by ETV6-NTRK3 fusion, t(12;15)(p13;q25)
- Same as in renal congenital mesoblastic nephroma

- Rapidly growing, may be huge
- Distal extremities, trunk, head/neck

- Good prognosis (<5% fatality)
Case 6

- 2 year old girl, right second toe nodule
Case 6

- Inclusion body fibromatosis
Inclusion body fibromatosis

- Syn Infantile digital fibroma/fibromatosis
- Benign but high recurrence rate (up to 75%)
- Almost always on digit (2\textsuperscript{nd}/3\textsuperscript{rd}/4\textsuperscript{th})
- May see synchronous / metachronous lesions
- Usually manifests in infancy
- Suspect whenever history of soft tissue lesion from digit of a young child/infant
- Look hard for eosinophilic inclusions (don’t confuse with RBCs)
- Confirm with trichrome stain
Case 25

- 1 year old boy, axillary lump
Case 25

- Fibrous hamartoma of infancy
Fibrous hamartoma of infancy

- < 2yo, 20% congenital
- Poorly circumscribed
- Axillary/inguinal, trunk, limbs
- Organoid
  1. Fibrous tissue
  2. Loose primitive mesenchymal cells
  3. Mature fat
- HARD TO EXCISE COMPLETELY
- 15% recurrence rate
Case 12

- 7 year old girl, lesion from thigh
Case 12

- Juvenile xanthogranuloma
Juvenile xanthogranuloma

- May be multiple
- Head and neck/trunk/extremities
- Tend to involute over time

Histology:
- Papules/nodules
- May be deep
- Depends on stage
  - Early – histiocytes, minimal lipid
  - Fully developed – foamy histiocytes, eosinophils, Touton giant cells
  - Late – fibrosis, eosinophils, few lipidized/giant cells
Case 21

- 7 mth old girl, scaly purpuric rash torso, axilla, scalp.
Case 21

- Langerhans cell histiocytosis
Suspect when see an odd mononuclear cell infiltrate, esp when accompanied by eosinophils

Don’t always see the classic “coffee bean”/grooved nuclei

Infiltrate may be obscured by other cell types depending on the site (eg osteoclastic giant cells in bone)

BRAF-V600E molecular testing in all cases (~50% paediatric cases +ve; if present, worse prognosis)
Lymphomas in children

1. Hodgkin lymphoma
2. Burkitt lymphoma
3. Acute lymphoblastic lymphoma
4. Anaplastic large cell lymphoma
5. Diffuse large B cell lymphoma