SMOOTH MUSCLE TUMOURS
NORMAL SMOOTH MUSCLE

• Cytology
• Immunohistochemistry
• Ultrastructure
Smooth Muscle Ultrastructure

- Many myofilaments running parallel to the long axis of the smooth muscle cell
- Myofilaments are of three types (thick myosin filaments, 12nm; these surrounded by thin actin filaments, 9nm)
- Actin and myosin filaments are contractile and aggregate into a myofibril.
- The intermediate filament desmin (10nm) is non contractile and is located in the dense body.
- There is a delicate basal lamina and many surface pinocytotic vesicles
LEIOMYOMAS

- Superficial: pilar (from arrector pilae muscle) or genital (vulva, nipple, scrotum)
- Deep
Pilar Leiomyoma

- Adolescents / young adults
- Solitary or multiple
- Extensor surfaces of limbs
- Confined to dermis
- Small (<20mm)
- Often painful
Pilar Leiomyoma Histology

- Confined to dermis
- Bundles / fascicles of smooth muscle
- Irregular border, non-encapsulated
- Occasionally mild degenerative atypia
- Very occasional mitosis (up to 1/10 hpf)
- Often epidermal hyperplasia
Pilar Leiomyoma Histology

- Confined to dermis
- Irregular border, not encapsulated
- Bundles of mature smooth muscle
- Occasional degenerative atypia
- Very occasional mitosis (up to 1 per 50 hpf)
- Often epidermal hyperplasia
Familial Pilar Leiomyomas

- Germline fumarate hydratase mutation (Krebs cycle enzyme)
- Autosomal dominant inheritance, variable penetrance
- Multiple cutaneous and uterine leiomyomas
- Renal cell carcinoma (non-clear cell)
Differential Diagnosis
Dermatomyofibroma

- Superficial dermal plaque
- Shoulder / trunk
- Spindle cells parallel to epidermis
- Adnexal structures preserved
- Myofibroblastic morphology
- Lacks desmin
Differential Diagnosis
Leiomyosarcoma

- Bigger (often > 20mm)
- Often infiltrates subcutis
- More atypia
- More mitoses
- Atypical mitoses
- Sometimes necrosis
Angioleiomyoma / Angiomyoma

- Middle aged adults, F>M
- Usually solitary
- Limbs, especially leg
- Painful
- Well demarcated
- Thick-walled blood vessels
Deep Leiomyomas

Deep leiomyomas will make more sense if we consider uterine smooth muscle tumours first
Uterine Leiomyomas

- Spindled
- Some epithelioid
- Some have fat (lipo-leiomyoma)
- Often hyalinised
- Occasional atypia (degenerative)
- Some mitoses
- ER and PR Positive
Intravenous leiomyomatosis

- In pre-menopausal women
- Benign neoplastic smooth muscle invades myometrial veins
- Some extend into extra-uterine veins (or IVC or heart)
- Usually good prognosis with hysterectomy
- Persistent disease in 30%
- Occasionally fatal
Benign Metastasising Leiomyoma

- Exclusively females
- Benign smooth muscle tumour in lungs of women with uterine (or previous uterine) leiomyoma.
- A few are associated with intravenous leiomyoma
- Generally good prognosis. A few die of respiratory failure.
Leiomyomatosis Peritonealis Disseminata

- Exclusively women
- Child-beariing age
- Black females > white females
- Associated with hormone production (pregnancy, contraceptive hormone therapy)
- Myriad of sub-serosal smooth muscle nodules
- Clinically indolent
Deep Leiomyomas

- Somatic soft tissue (limbs) (M = F) Virtually no mitotic activity
- Body cavity LM (pelvis, retroperitoneum, F >>> M) These often hyalinised and ER+ & PR+. Often many typical mitoses. These (particularly in pelvis) of hormonally driven uterine leiomyoma type.
Deep Leiomyoma Histology

- Subcutis / subfascial
- Bundles / fascicles of smooth muscle at right angles to each other
- No mitoses or very few (in non-uterine type)
- Little atypia (and if present degenerative without large nucleoli)
- No necrosis
Immunosuppression related soft tissue tumours?
Immunosuppression-related soft tissue tumours

- Kaposi sarcoma (HHV8)
- EBV-associated smooth muscle tumour
EBV-associated smooth muscle tumour

- HIV / AIDS
- Post-transplant immunosuppression
- Congenital immunosuppression

- Well-differentiated smooth muscle tumour
- 50% have foci of round myoid cells
- Little atypia, few mitoses
- SMA+ desmin +/-
EBV-associated SMT

- Multifocal
- Soft tissue
- Liver, lung, spleen, thyroid
- Dura
- Clinically indolent
Deep Soft Tissue Leiomyosarcoma

- Middle-aged adults. Rare in childhood
- Half arise in retroperitoneum (2/3 of these in women)
- Vena cava, a common site (3/4 of these in women)
- Limbs / trunk, M=F
INTERMISSION
Haemangiopericytoma
Solitary Fibrous Tumour

Not a smooth muscle or perivascular myocyte tumour. But the cellular subset of SFTs used to be considered a haemangiopericytoma. Actually myofibroblastic (CD34 pos, smooth muscle markers neg)
Perivascular Smooth Muscle (Myopericyte) Tumours

- Sino-nasal haemangioepericytoma
- Myopericytoma
- Angiomyoma (Angioleiomyoma)
- Myofibroma
- Glomus tumour
Sinonasal Haemangiopericytoma
Myopericyte tumours (myopericytoma, angiomyoma, myofibroma, glomus tumour)

- Superficial tumours
- May show some degenerative atypia
- Smooth muscle marker positive (SMA and H-caldesmon)
- Tend to be desmin negative
Myopericytoma

• Perivascular concentric growth
• Haemangiopericytic vasculature
• SMA pos, H-caldesmon pos
• Desmin neg
• Clinically indolent
Myofibroma
Glomus Tumours
normal glomus body
Glomangiomyoma
Classic Angiomyolipoma

- AML benign mesenchymal tumour comprising variable mixtures of fat spindled/epithelioid smooth muscle cells and thick-walled blood vessels
- Kidney is classic location
- Some related to tuberous sclerosis (M=F), in sporadic cases F>M (4:1)
- The muscle is of ‘perivascular epithelioid type’ i.e. HMB45+ and melan A+
Classic AMLs (interesting facts)

- Clinically benign (but may result in catastrophic retroperitoneal haemorrhage)
- May be multifocal (especially in tuberous sclerosis)
- Occasional degenerative nuclear atypia
- Can invade renal vein / IVC
- Can be present in LNs (multifocal growth)
Epithelioid AML

- >50% are associated with tuberous sclerosis
- Clinically more aggressive than classic AML
- Larger, sometimes necrotic, infiltrative margins, renal vein / IVC extension
- Approximately 1/3 metastasise (LN, liver, lungs, spine)
- Differential diagnosis (RCC, metastatic MM)
Perivascular epithelioid cell proliferations (HMB45 pos, melan A pos)

- PECOMA
- Angiomyolipoma
- Lymphangioleiomyomatosis