Adnexal tumours of skin can be classified based on putative histogenesis, or what structure the tumour recapitulates.

This may or may not be the true origin of the tumour.

Remains a useful framework for thinking about them.

Generally uncommon and variable histological features.

Sometimes precise categorisation is difficult, as morphological features of different tumours can overlap.
• Benign tumours are uncommon and malignant examples usually even rarer

• Features of malignancy may be difficult to appreciate or not present in a small biopsy

• Some adnexal tumours are seen in the context of syndromes
CATEGORISATION

• Hair follicle

• Sweat glands – eccrine, apocrine

• Sebaceous gland
HAIR FOLLICLE TUMOURS
TRICHOEPI THELIOMA & TRICHOBLASTOMA

• I think there is a lot of overlap between TE & TB and they’re basically the same tumour, with TB being larger and extending more deeply

• Lobules of basaloid cells

• No epidermal connection

• Lobules surrounded by a cellular stroma – recapitulates hair follicle
TRICHOEPITHELIOMA & TRICHOBLASTOMA

- What is the arrowed structure?
- Papillary mesenchymal body
- Immunohistochemistry reported to be useful in distinguishing between TB/TE and BCC
- In reality, not straightforward
- CK20 is the marker I’ve found to be the most useful
TRICHOEPITHELIOMA & TRICHOBLASTOMA

- Pigmented TB

- Often has a small amount of melanin focally
• BCC may have an epidermal connection (TB usually not, TE can do)

• Cells of BCC are larger, more apoptosis

• Both may have mitotic activity, particularly TB

• BCC usually has peritumoural clefting with mucin, at least focally

• BCC usually lacks cellular stroma

• Generally speaking, safer managed as a BCC if unsure, MDT discussion can be helpful. Often recommend conservative complete excision anyway.
DESMOPLASTIC TRICHOEPITHELIOMA (DTE)

- Totally different morphology to TE
- No epidermal connection
- Narrow basaloid epithelial strands
- Keratocysts
- Focal calcification
DESMOPLASTIC TRICHOEPITHELIOMA (DTE)

- Often has a foreign body giant cell reaction
- No true ductal differentiation
- Difficult to distinguish from microcystic adnexal carcinoma on a small biopsy
- I recommend conservative complete excision
TRICHILEMMOMA

- Lobules of clear cells, focal connection to the epidermis
- Can have papillomatosis in overlying epidermis – clinical can be "wart"
- Peripheral palisade
- Classically has a hyaline basement membrane mantle
TRICHILEMMOMA

- Lobules of clear cells, focal connection to the epidermis
- Can have papillomatosis in overlying epidermis – clinical can be ‘wart’
- Peripheral palisade
- Classically has a hyaline basement membrane mantle
TRICHILEMMOMA

- Desmoplastic trichilemmoma
- Infiltrative appearance, hyalinization
- Can be mistaken for SCC
TRICHILEMMOMA

• Trichilemmomas can be associated with what syndrome?
  
  • Cowden’s

• PTEN mutation
Trichilemmomas can be associated with Cowden's syndrome and PTEN mutation. Multiple skin and mucosal lesions, visceral malignancies especially breast, thyroid.
SWEAT GLAND TUMOURS
SWEAT GLAND TUMOURS

• Benign: poroma, hidradenoma, cylindroma, syringoma, spiradenoma

• Malignant: microcystic adnexal carcinoma (MAC), porocarcinoma

• Others (e.g. syringoid eccrine ductal carcinoma, malignant transformation in pre-existing benign neoplasm)
BENIGN SWEAT GLAND TUMOURS: CLINICAL FEATURES

• Generally located on the head and neck

• Nonspecific clinically and often thought to be a cyst

• May result in these tumours being ‘shelled out’ or curetted
POROMA

• Clinical – most commonly sole or side of the foot

• But can occur at other sites

• Reddish, slightly scaly nodule
• Lobules of small, basaloid, regular cells that are connected to the epidermis
Ductal differentiation
What ancillary tests can be used to highlight ductal differentiation?

- Special stains: PAS – highlights duct lining ‘cuticle’
- Immunohistochemistry – EMA, CEA
- In reality – can be difficult to interpret
- Levels may be more helpful
HIDRADENOMA

- Well-circumscribed tumour
- Lobulated, sometimes cystic masses
- Eosinophilic or clear cells
- Ductal diff usually evident
HIDRADENOMA

- Well-circumscribed tumour
- Lobulated, sometimes cystic masses
- Eosinophilic or clear cells
- Ductal diff usually evident
HIDRADENOMA

• Look for atypical features: cytological atypia, mitoses (≥2/10 HPF) ‘atypical hidradenoma’

• Deposits in local LNs have been reported in otherwise benign-appearing hidradenomas
CYLINDROMA

• No connection to epidermis

• Multiple basaloid tumour lobules in jigsaw or mosaic pattern

• Smaller outer cells and larger inner cells

• Hyaline mantle and deposits of hyaline material ‘cylinders’

• Morphological overlap with spiradenoma (‘spiradenocylindroma’)

[Image of CYLINDROMA]
Multiple cylindromas may be associated with other tumours/lesions:

- Brooke-Spiegler: trichoepitheliomas, spiradenomas, milia

Can see difficult to classify tumours e.g. spiradenocylindroma

Germline mutation in CYLD reported in Brooke-Spiegler
SYRINGOMA

• Usually presents clinically as multiple small papules, female predominance

• Ducts described as having typical ‘tadpole’ morphology

• May see clear cell change

• Distinguish from DTE: DTE has keratocysts and no true ductal diff

• May be impossible to exclude MAC on small biopsy – CPC essential
SYRINGOMA
SPIRADENOMA

• Clinical: nodule, may be painful, may look blue-ish or vascular

• Basaloid tumour lobules present in dermis, may extend into subcutaneous fat
SPIRADENOMA

- 2 different cell types – smaller outer cells and larger inner ones
- Can be richly vascular
- Lymphocytes often present
MICROCYSTIC ADNEXAL CARCINOMA (MAC)

- Clinical:
  - Plaque
  - Usually on head and neck, especially upper lip
  - Locally aggressive
  - Does not usually metastasise
MICROCYSTIC ADNEXAL CARCINOMA (MAC)

- Ductal structures
- Resembles syringoma or DTE
MICROCYSTIC ADNEXAL CARCINOMA (MAC)

- Deeply infiltrative
- Perineural invasion
SEBACEOUS TUMOURS
SEBACEOUS TUMOURS

- Sebaceous hyperplasia – probably a true neoplasm, as mutations in oncogenes has been reported
- Sebaceous adenoma
- Sebaceoma
- Sebaceous carcinoma

- Interobserver variability in the diagnosis of these lesions is reported to be high
What syndromic associations are there with sebaceous neoplasms (SN)?
SEBACEOUS ADENOMA

• Clinical: papule or nodule on the face – often thought to be BCC
SEBACEOUS ADENOMA

• Orderly recapitulation of the lobular architecture of the normal sebaceous gland
SEBACEOUS ADENOMA
SEBACEOUS ADENOMA

• Less than 50% basaloid cells

• Can show prominent connection to the epidermis or appear to replace it

• The classical neoplasm associated with Muir-Torre syndrome (MTS)
SEBACEOMA

• No orderly lobular architecture

• More than 50% basaloid cells

• ΔΔ sebaceous carcinoma
SEBACEOMA
SEBACEOMA

- No orderly lobular architecture
- More than 50% basaloid cells
- ∆∆ sebaceous carcinoma
SEBACEOUS CARCINOMA

• Periocular and extraocular

• Initially periocular was reported to behave more aggressively, but there was no difference in overall survival in a large study
SEBACEOUS CARCINOMA
PERIOCULAR

• 2nd or 3rd most common malignant tumour of the eyelid after BCC and maybe SCC

• Clinical diagnosis is difficult as the presentation is varied e.g. like chalazion
SEBACEOUS CARCINOMA
PERIOCULAR
SEBACEOUS CARCINOMA PERIOcular

- Pathological diagnosis may also be challenging

- Can show pagetoid pattern and mimic Bowen’s
SEBACEOUS CARCINOMA

• Poorly differentiated examples can have very focal sebaceous differentiation only
SEBACEOUS CARCINOMA
SEBACEOUS CARCINOMA
SEBACEOUS CARCINOMA

• Beware of a poorly differentiated carcinoma on the eyelid

• Immuno not particularly helpful – e.g. adipophilin is good when it works well but these are usually cases where there is obvious sebaceous diff morphologically

• EMA, CK7 can be useful.
MUIR TORRE SYNDROME (MTS)

• Considered to be a phenotypic variant of Lynch syndrome (HNPCC)

• Autosomal dominant with variable penetrance

• Molecular basis of Lynch: germline mutations in MMR proteins: MSH2, MSH6, MLH1, PMS2.

• Leads to microsatellite instability

• MSH2 mutation particularly associated with MTS

• Amsterdam criteria for diagnosis

• Cutaneous SNs may antedate the visceral malignancies
• What are the commonest visceral malignancies in MTS?

• GI, bladder, endometrial
• What factors make a cutaneous SN more likely to be associated with MTS?

• Younger patient
• Multiple
• Outside head and neck
• Loss of MMR proteins doesn’t equal MTS

• Intact MMR proteins on IHC does not exclude MTS

• Increasingly moving towards clinical criteria/ clinical correlation

• However, sebaceous tumours apart from seb H are uncommon and it is always worth highlighting the potential association with MTS
Images in this presentation used for teaching purposes only.