Cutaneous neoplasia and hamartomas (BCC, SCC, melanoma, vascular malformations, sarcomas)

Overview of aetiology of neoplasms

Benign epithelial tumours
Hair follicle tumours
Tumours of sebaceous glands
Sweat gland tumours
Basal cell carcinoma (basal cell epithelioma, rodent ulcer)
Premalignant conditions
Squamous cell carcinoma (squamous epithelioma)
Epidermal naevi
Vascular birthmarks
Vascular malformations
Melanocytic naevi
Dermal melanocytosis
Malignant melanoma
Axillary lymphadenectomy
Inguinal lymphadenectomy
Soft tissue sarcoma
Cutaneous metastatic malignant tumours
Miscellaneous conditions

Overview of aetiology of neoplasms

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A neoplasm is an abnormal mass of tissue, the growth of which exceeds, and is uncoordinated with that of the normal tissues and which persists in the same excessive manner after cessation of the stimulus which evoked the change.

A malignant neoplasm is one that invades surrounding tissues and has a propensity to metastasize.

Initiation, promotion and progression lead to unrestrained growth and proliferation:

**Initiation:** a change in the genome of a cell

**Promotion:** the change made permanent by cellular division (initiators and promoters include UV and ionizing radiation, chemical carcinogens, viruses)
Progression: further cell division to form an invasive tumour

Inappropriate activation of normal cellular proto-oncogenes to become oncogenes – these proto-oncogenes encode growth factors, growth factor receptors or transcription factors
Inactivation of other cellular genes called tumour suppresser genes
p53 tumour suppresser gene is mutated in the majority of human cancers
UV radiation is the most important factor
First, mutations in cellular DNA and a failure of DNA repair
Second, production of immunosuppressive cytokines, depletion and alteration of antigen-presenting LCs and systemic induction of Tc-cells by altered LCs, inflammatory macrophages and cytokines
Sunburn, suntanning, local and systemic immunosuppression, skin cancer and precancer are attributed to UVB radiation (290–320 nm)
UVA radiation (320–400 nm) generates oxygen-free radicals that damage cell membranes and nuclear DNA, contributing to erythema, photo-ageing and carcinogenesis
 Interruption of intercellular and intracellular signalling cascades regulating transcription and function of viral oncoproteins in human keratinocytes
 Interaction between a physical carcinogen (UV part of the sunlight) and a ‘low risk’ (non-mutagenic) papillomavirus infection

Benign epithelial tumours

Seborrhoeic keratosis (seborrheic wart, senile wart, basal cell papilloma)

Incidence
Males = females, fifth decade onwards
White races
Stucco keratosis – non-pigmented seborrhoeic keratosis on the limbs
Dermatosis papulosa nigra – multiple facial lesions, dark skinned races, early onset

Aetiology
Familial – autosomal dominant
Inflammatory dermatosis
Manifestation of visceral malignancy
Oestrogen administration

Pathology
Accumulation of immature keratinocytes between basal and keratinizing layers
Acanthosis – thickening of the epidermis
Elongation of dermal papillae
Malignant transformation reported but rare (squamous type)

Clinical
Verrucous plaque
Face, hands and upper trunk
May be heavily pigmented
Multiple lesions aligned in direction of skin folds
May bleed, become inflamed and itch
May shed but then reform

**Treatment**
Curettage or excision
Cryotherapy
Topical trichloroacetic acid

**Digital fibrokeratoma**
Papillary or keratotic outgrowth in the region of a finger joint
Adults, males > females
Follows trauma
Hyperkeratosis and acanthosis (thickening of the epidermis, specifically the stratum spinosum)
Distinguish from supernumerary digit
Treat by excision

**Keratoacanthoma** *(Molluscum sebaceum)*
Rapidly evolving tumour which resolves untreated
Keratinizing squamous cells originating in pilosebaceous follicles

**Incidence**
White races
Males > females (×3)
One-third frequency of SCC
Middle age onwards

**Aetiology**
Sun exposure
Coal, tar and carcinogenic hydrocarbons (multiple lesions)
Injury and infection including skin graft donor sites
Association with carcinoma of the larynx, internal malignancies and leukaemia
Association with deficient cell-mediated immunity (multiple lesions)

**Pathology**
Keratin-filled crypt
Rapidly dividing squamous cells deriving from skin appendages
Atypical mitoses and loss of polarity

**Clinical**
Globular tumour
Keratin plug or horn
Radial symmetry
Resolution begins after 6 weeks, takes 3 months
Face, dorsum of hand
Torre’s syndrome: multiple internal malignancies, KAS and sebaceous adenomas
Distinguish from SCC

**Treatment**

Excise to provide good histological specimen
Spontaneous resolution leaves unsightly scar
5-FU and radiotherapy shorten time to resolution

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### Hair follicle tumours

**Trichilemmal cyst**

Sebaceous cyst of hairy skin
Wall resembles hair follicle
Situated on the scalp
Women > men
Middle age
Familial (AD)
Rupture causes cell proliferation and occasionally malignant change
Ruptured cell wall may fuse with surrounding skin (marsupialization)
Treat by excision

**Trichofolliculoma**

Multiple malformed hair roots arising from an enlarged follicle canal
Keratin-filled crater
Hairs may emerge from a central punctum
Mainly on the face

**Tricho-epithelioma**

Epithelial tumour differentiating towards hair follicle cells

**Incidence**

Rare, onset at puberty

**Aetiology**

Familial (AD)
Pathology
Resembles BCC
Rounded masses of fusiform cells
Lacunae filled with keratin
Tumour islands may connect with hair follicles
Malignant change (BCC)

Clinical
Pinkish nodules
Cheeks, eyelids and nasolabial folds
Often diagnosed as BCC, pigmented lesions mistaken for MM

Treatment
Excision

Pilomatrixoma (benign calcifying epithelioma of Malherbe)
Hamartoma composed of dead, calcified cells, which resemble those of hair matrix

Incidence
Uncommon
Mainly <20 years
Females > males
Association with myotonia atrophica

Pathology
Well-circumscribed dermal tumour
Cells mature from outer to inner layers
Similar to hair matrix cells maturing from cortex to root sheath
Central calcification and ghost cells, lacking basophilic granules

Clinical
Dermal/subcutaneous tumour
Head, neck, upper extremity
Stony hard consistency

Treatment
Excision

Tumours of sebaceous glands

Sebaceous adenoma
Benign tumour composed of incompletely differentiated sebaceous cells

Incidence
Rare tumour
Either sex
Mainly in the elderly

Pathology
Multilobular tumour of the upper dermis
Small basophilic sebaceous matrix cells
Larger cells containing fat globules

Clinical
Ulcers/plaques/sessile or pedunculated lesions
Yellowish hue
Face (including eyelid) or scalp
Sudden increase in growth rate

Treatment
Excision
Recur if incompletely excised
Radiosensitive

Sebaceous carcinoma
Malignant growth of cells differentiating towards sebaceous epithelium

Incidence and aetiology
Rare – 0.2% of skin cancers
May follow radiodermatitis

Pathology
Deeply set in dermis, epidermis usually uninvolved
Outer basophilic undifferentiated cells
Central differentiating cells with cytological atypia
Cytoplasmic vacuolation and fat droplets
Invasion

Clinical
Yellowish nodules on face and scalp
Slow or rapid growth
Metastasis uncommon

Treatment
Excision

Epidermoid cyst (sebaceous cyst)

Incidence and aetiology
Young and middle-aged adults
Inflammation and obstruction of a pilosebaceous follicle
Pathology
Epidermal lining
Birefringent keratin and breakdown products
Cholesterol clefts

Clinical
Spherical cyst in the dermis
Tethered to epidermis
Enlarge and suppurate through punctum

Treatment
Excision

Sweat gland tumours

Apocrine glands release lipid secretions in membrane-bound vesicles (e.g. breasts)
Eccrine glands release secretions by exostosis into ducts
Holocrine glands discharge whole cells which then disintegrate to release secretions
Sweat glands: mainly eccrine secretion, some apocrine
Sebaceous glands: holocrine secretion of sebum
Three benign eccrine sweat gland tumours: syringoma, acrospiroma, cylindroma

Syringoma (papillary eccrine adenoma)
Benign tumour of eccrine sweat gland origin

Incidence
Uncommon
Females > males
Onset during adolescence
Multiple tumours associated with Down’s syndrome

Pathology
Collections of convoluted sweat ducts in the upper dermis
Tail-like projection of cells – characteristic tadpole appearance

Clinical
Small, yellowish dermal papules, usually <3 mm
May appear cystic – injury may cause release of a small amount of clear fluid
Chest, face, neck
May resemble tricho-epithelioma or xanthelasma

Treatment
Excision
Intralesional electrodesiccation
Laser
Eccrine acrospiroma

Tumour derived from eccrine sweat duct epithelium
Epidermal, juxta-epidermal or dermal
Eccrine poroma commonest subtype: juxta-epidermal

Incidence
Male = female
Middle age
Usually acral – palms and soles

Pathology
Sweat gland duct cell proliferation
Cells contain glycogen and glycolytic enzymes
Overlying hyperkeratosis
Malignant change (malignant eccrine poroma) reported

Clinical
Hyperkeratotic plaque on sole or palm
May ulcerate

Treatment
Excision

Dermal cylindroma (dermal eccrine cylindroma, turban tumour, Spiegler’s tumour)

Derived from the coiled part of sweat glands (part secretory, part duct)

Incidence
Uncommon
Females > males (×2)
Often familial (AD)
Early adult life

Pathology
Columns of cells interspersed with hyaline material
Large and small (peripheral) cell types

Clinical
Scalp and adjacent skin
Pinkish fleshy tumours
Usually multiple and hairless
May be painful
Malignant change very rare
Distinguish from trichilemmal cyst

Treatment
Excision
**Adenoid cystic carcinoma of the scalp**

Malignant tumour of eccrine glands

Usually arises in salivary glands

Lacrimal glands and mucous glands of upper respiratory tract

Rarely arises primarily in the skin, mainly eccrine sweat glands of the scalp

Slow growing

Invades fascial planes, nerves and bone

Characteristic lattice-type appearance microscopically

Rarely completely excised

Not radiosensitive

Treat by excision with histological control of margins

**Sweat gland carcinoma**

Malignant epithelial tumour of the sweat glands

**Incidence**

Rare

Males = females

Middle age onwards

**Pathology**

Adenocarcinoma within the dermis

Eccrine or apocrine varieties

Eccrine commonest, frequently metastasizes

**Clinical**

Painful, reddish nodules within the dermis

Firm/hard

Irregular border

Occur anywhere, mainly scalp and face

Slow growth but may metastasize

**Treatment**

Wide excision

Monitor lymph nodes

**Basal cell carcinoma (basal cell epithelioma, rodent ulcer)**

Malignant tumour composed of cells derived from the basal layer of the skin

**Incidence**

Commonest malignant tumour of the skin in white races

Increased prevalence in locations of high sunlight exposure

Males > females except lower extremity lesions (female:male ratio = 3:1)
Uncommon in non-Caucasians
75% of patients > 40 years old

Aetiology
UV and ionizing irradiation
Burn and vaccination scars
Arsenicals
Immunosuppression
Occasionally has a familial inheritance
Malignant change in sebaceous naevi and other adnexal hamartomas
Face at much greater risk than other sun-exposed areas (may be related to density of pilosebaceous follicles)

Pathology
Tumour cells arranged in palisades
Cell–cell desmosomes preserved
Well-organized surrounding stroma
Varying degrees of atypia
Small buds of tumour off the main mass
Occasionally harbour a melanocytic proliferation
Mucin accumulation and central necrosis characteristic of cystic lesions

Clinical
Pinkish, pearly nodules
Telangiectasia
May be ulcerated, encrusted or pigmented
Nodular, ulcerated, superficial, sclerosing, cystic, morphoeic, desmoplastic
Usually slow growing
Very rarely metastasize via lymphatics
Long-standing tumours may invade deep into subcutaneous tissues
Distinguish from sebaceous hyperplasia

Gorlin's syndrome (basal cell naevus syndrome): AD inheritance, multiple BCCs, palmar pits, jaw cysts, sebaceous cysts, abnormalities of ribs and vertebrae, dural calcification
Bazek's syndrome: association of multiple BCCs with follicular atrophoderma

Treatment
Excisional biopsy: 2–5 mm margins, antibiotic cover if ulcerated
35% of incompletely excised tumours recur; re-excision of incompletely excised tumours shows residual tumour in only 30% of cases
5-FU or photodynamic therapy for superficial lesions
Radiotherapy

Guidelines for the management of basal cell carcinoma
• Review of surgical and non-surgical modalities used in the treatment of BCCs including
  • Mohs’ micrographic surgery
  • Radiotherapy