RARE SKIN TUMOURS

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Rare skin tumours – scope of the topic

Malignant tumours arising from skin elements excluding melanoma and common non-melanoma skin cancers (basal and squamous cell carcinomata).

Incidence

- Standardized incidence : 0.82/100,000 per year

  Rare skin cancers – population based cancer registry descriptive study of 151 consecutive cases diagnosed between 1980 – 2004

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Tumour sub-types

Breakdown of tumours		Standardized Incidence
Sarcoma	39%	0.37/100, 000 per year
Adnexal Ca	23%	0.16/100,000 per year
Merkel cell	18%	0.13/100,000 per year
Pagets	18%	0.15/100, 000 per year

Classification

- Epidermal tumours
  - Merkel cell carcinoma
- Dermal tumours
  - Dermatofibrosarcoma protruberans
  - Atypical fibroxanthoma
  - Dermal vascular tumours
- Adnexal tumours
  - Microcystic Adnexal Carcinoma (MAC)
  - Sebaceous carcinoma
  - Apocrine carcinoma
- Tumours arising from immigrants to the skin
  - Mycosis Fungoides

Principles of management of rare skin tumours

- History
- Examination
- Tissue diagnosis and confirmation of histology
- Staging the disease
- Operative management
- Adjuvant treatment and multidisciplinary care
- Surveillance

Epidermal tumours – Merkel cell carcinoma

- Merkel cell – neural crest derived cell which is functionally insignificant
- Incidence
- History – rapid growth and ulceration
- Examination – nodular lesion which may have evidence of in-transit or LN metastases
- Tissue diagnosis – small, round cells (DDx – small cell carcinoma), cytoplasmic granules
- Staging disease - CT PET scan

Principles of management – merkel cell tumour

- Surgical excision : wide local excision 2-3 cm, +/- SLNBx, elective LN dissection
- Management within the context of a multidisciplinary team
- Adjuvant radiotherapy – primary site and draining lymph node basin, exquisitely radiosensitive.
- Limb infusion for palliation
- Surveillance

Dermatofibrosarcoma protruberans (DFSP)

- Primary fibrosarcoma of skin
- History – slow growing nodule which may ulcerate
- Examination – aggregated protruberant nodules within a plaque
- Tissue diagnosis – fibroblasts in a "storiform" pattern with honeycomb extension into fat.
- Staging – locally aggressive, metastases rare

Principles of management - DFSP

- Wide local excision with reconstruction with a 3 cm margin including deep fascia.
- Multidisciplinary team setting with referral for adjuvant radiotherapy to the primary site.
- Surveillance - for local recurrence, metastases rare.

Atypical fibrous xanthoma (AFX)

- Spindle cell tumour of actinic damaged skin
- Within spectrum of disease with malignant fibrous histiocytoma (MFH)
- History – slow growing nodule
- Examination – series of skin nodules
- Tissue diagnosis – spindle cell sarcoma
- Staging – metastatic disease rare

Principles of management - AFX

- Wide local excision of 1-2 cm excluding deep fascia and reconstruction
- Management of the lymph nodal basin on clinical grounds.
- Adjuvant radiotherapy to limit local recurrence
- Surveillance – tendency to locally recur

Dermal vascular tumours

- Spectrum of diseases
  - Kaposi sarcoma
  - Haemangioendothelioma
  - Haemagiopericytoma
  - Angiosarcoma

Kaposi Sarcoma

- History – four clinical scenarios : European/African/Transplant/HIV-AIDS
- Examination – spectrum ranging from violaceous patches, plaques and nodules
- Tissue diagnosis – dilated jagged vascular channels lined by and with perivascular spindle cells; HHV – 8
- Disease staging – likelihood of dissemination related to underlying clinical setting.

Principles of management of KS

- Surgical excision and reconstruction with a modest margin : 5 - 10 mm
- Treatment within a multidisciplinary team
- Adjuvant radiotherapy – for treatment of multiple lesions
- Adjuvant chemotherapy – extensive or disseminated disease, Interferon therapy, limb infusion
- Medical management – anti-retroviral agents, immune modulation

Haemangioendothelioma

- Vascular tumour arising from tissue around medium and large calibre veins.
- Malignant potential - intermediate
- Variable clinical behaviour –

  Recurrence 40%

  Metastases 30%

  Mortality 15%

Haemangiopericytoma

- Considered to be derived from pericytes
- Variable clinical course
  - Benign – 2/3
  - Malignant – 1/3
- Markers of aggression
  - Tumour size
  - Histological
  - Site

Angiosarcoma

- Malignant endothelial sarcomas
- History – may be associated with lymphoedema post mastectomy (Stewart Treves syndrome) or post radiotherapy
- Examination – fleshy, ulcerated tumours with poor demarcation and tendency to bleed with minimal trauma
- Tissue diagnosis – degrees of differentiation, CD 31 stain.

Principles of management - angiosarcoma

- Wide local excision with 2 cm margin excluding the deep fascia and reconstruction
- Surgical excision recognizing functional and aesthetic considerations in addition to disease stage which determines curative potential versus palliation
- Interventional radiology as a surgical aid.
- Management of the nodal basin on clinical grounds
- Adjuvant radiotherapy

Adnexal tumours

- Tumours including sebaceous carcinoma, microcystic adnexal carcinoma (MAC), Apocrine carcinoma
- History : typically delay in diagnosis with relatively indolent course in early stages
- Tissue diagnosis
- Staging – advanced local disease or metastases at diagnosis

Principles of management of adnexal tumours

- Wide local excision of 2 cm excluding deep fascia and reconstruction
- Moh's controversy : "Will MAC be back?"
- Adjuvant radiotherapy to limit local recurrence and for perineural invasion (20%)
- Management of the lymph nodal basin – dissection of clinically and FNA positive nodes and elective radiotherapy to uninvolved nodal basin.

Tumours of dermal immigrant cells

- Mycosis fungoides – cutaneous T cell lymphoma
- Lymphoma may run an aggressive or indolent course
- Management
  - Obtaining a tissue diagnosis
  - Treatment of the underlying condition

Conclusion

Rare skin tumours characterized by :

- Delay in presentation
- Advanced disease at presentation
- Histopathological challenges in establishing tissue diagnosis
- Require aggressive local resection and reconstructive challenges
- Local recurrence due to advanced disease
- Often considered diagnoses of exclusion