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# References for Dermal Tumours talk by Mr. Paul Millican

- Authors of Article, Aaron H.J. Withers, Nicholas D.L. Brougham, Rachel M. Barber and Swee T. Tan. 2011. *Atypical fibroxanthoma and malignant fibrous histiocytoma* Journal of Plastic, Reconstructive & Aesthetic Surgery. Volume:64, Pages e273ee278. DOI 10.1016/j.bjps.2011.05.004
- Authors of Article, M. Poulsen and J. Harvey. 2002. *Is there a Diminishing Role for Surgery for Merkel Cell Carcinoma of the Skin?* ANZ J Surg. Volume 72, Pages 142 - 146
- Authors of Article, Paul T. Martinelli, Philip R. Cohen, Keith E. Schulze, Jaime A. Tschen and Bruce R. Nelson. *Sebaceous Carcinoma*. Skin Cancer. Chapter 17. Pages 240 – 250.

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# Diagnosis & Management of Dermal tumours

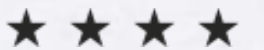
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## 5 - Fierce Tumours

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# benign dermal tumours

- \* cysts, dermoids,
- \* lipoma
- \* dermatofibroma & cellular dermatofibroma
- \* neurofibroma & neurofibromatosis NF-1, NF-5
- \* keloid
- \* haemangioma & variations
- \* sebaceous adenoma & Muir-Torre syndrome
- \* pilomatrixoma
- \* xanthelasma



# Skin Malignancies

- \* BCC
- \* SCC
- \* Melanoma
- \* 5 Fierce Tumours
- \* Lymphocyte Associated & Cutaneous Infiltrates
- \* Malignancies Metastatic to Skin
- \* Rare Epithelial Malignancies .. eg extra-mammary Paget's

# Viral associated tumours

Viral-Assoc'd Nonmelanoma Skin Cancers -- Amer J DermatoPathology August2009 Vol 31 - Issue 6 - pp 581-573

- \* HPV human papilloma virus
  - \* SCC
  - \* Actinic Keratoses - HPV subtypes : 21,5,8,16, & 18 -  
see [http://www.freak-search.com/en/thread/5012871/viral\\_origin\\_of\\_cancers\\_cont](http://www.freak-search.com/en/thread/5012871/viral_origin_of_cancers_cont).
  - \* Organ Transplantation ..... NEJM 2003, 348:1681-1691 Euvrard S. et al
  - \* Verruca vulgaris - wart papilloma virus
- \* Epstein-Barr Virus to cancer of the nose/pharynx, burkitt's lymphoma
- \* Kaposi's Sarcoma HHV8 human herpes virus-8
- \* Merkel Cell polyoma
- \* HTLV-1 human T-cell leukaemia virus-1
- \* Hepatitis B & C

# 5 Fierce Tumours

- Aggressive skin malignancies
  - Needing wide excision
    - Not melanoma / SCC / BCC
    - Not lymphatic system
    - Primary

# 5 Fierce Tumours

(1) **M**erkel Cell Carcinoma

(2) **A**ppendageal Tumours

- **S**ebaceous - **S**ebaceous Gland Carcinomas
- **E**ccrine - **M**icrocystic Adnexal Carcinoma
- **A**pocrine - **A**denoid Cystic Carcinoma

(3) **D**FSF - Dermato Fibro Sarcoma Protuberans

(4) **A**ngiosarcoma // Kaposi's Sarcoma

(5) **M**FH - Malignant Fibrous Histiocytoma, **A**FX Atypical Fibrous Xanthoma



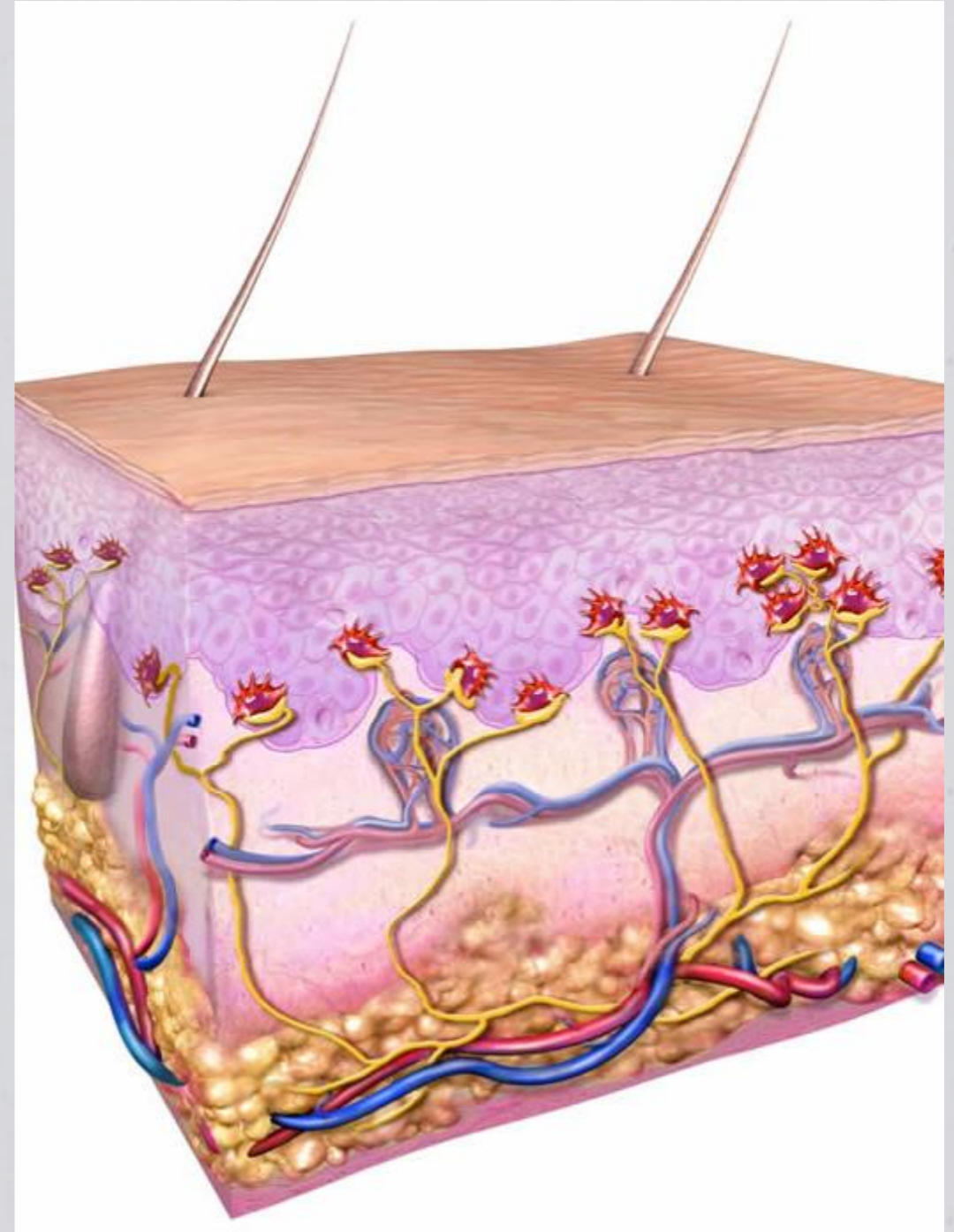


# Merkel Cell carcinoma

Neuro-Endocrine Ca of the Skin, Small Cell Ca of Skin

[www.merkelcell.org](http://www.merkelcell.org)

- \* cell of origin .... merkel cells = touch-sensitive cells in the basal layer of the epidermis
- \* forehead, brow, nasolabial, chin, elbows, anterior shin,
- \* pink/red nodular appearance -similar to BCC
- \* 33% - local recurrence
- \* 75% - lymph node involvement
- \* 33% - distant mets





# Neuro-Endocrine Tumours

Cell Carcinoma - **Skin**

## NET's

Cell **Lung** Carcinoma SCLC

- \* Medullary Carcinoma **Thyroid**
- \* **Adrenal** Pheochromocytoma
- \* Carcinoid - ileum, **appendix**, urinary tract
- \* GEPS - gasto-entero-pancreatic **Foregut** gastrinoma, **Pancreatic** insulinoma, glucagonoma, somatostatinomas, VIPomas, Zollinger-Ellison syndrome,
- \* Pituitary - ant. pituitary NET

\* MEN-1 MEN-2 von Hippel-Lindau Carney complex

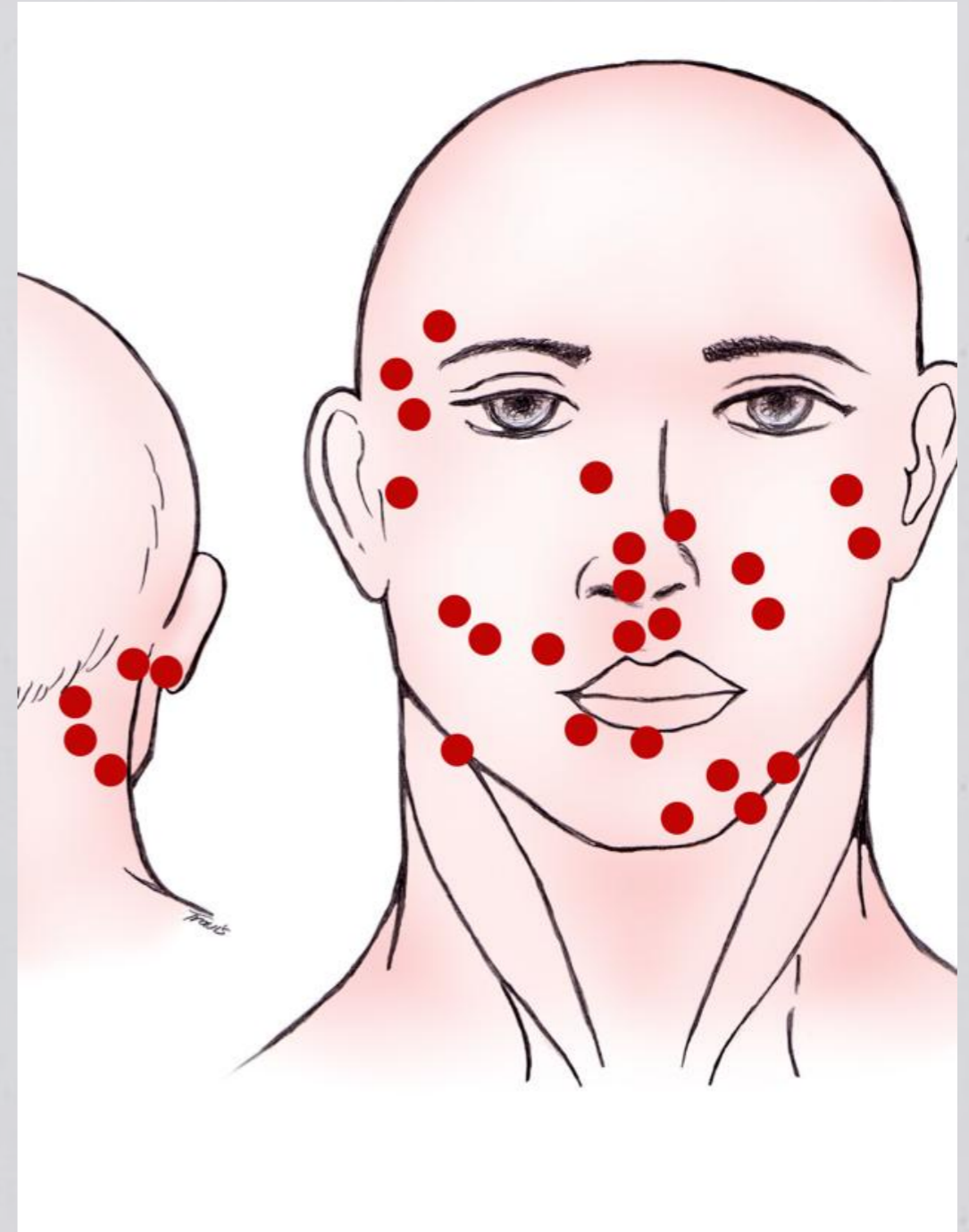


# Merkel Cell carcinoma

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[www.merkelcell.org](http://www.merkelcell.org)

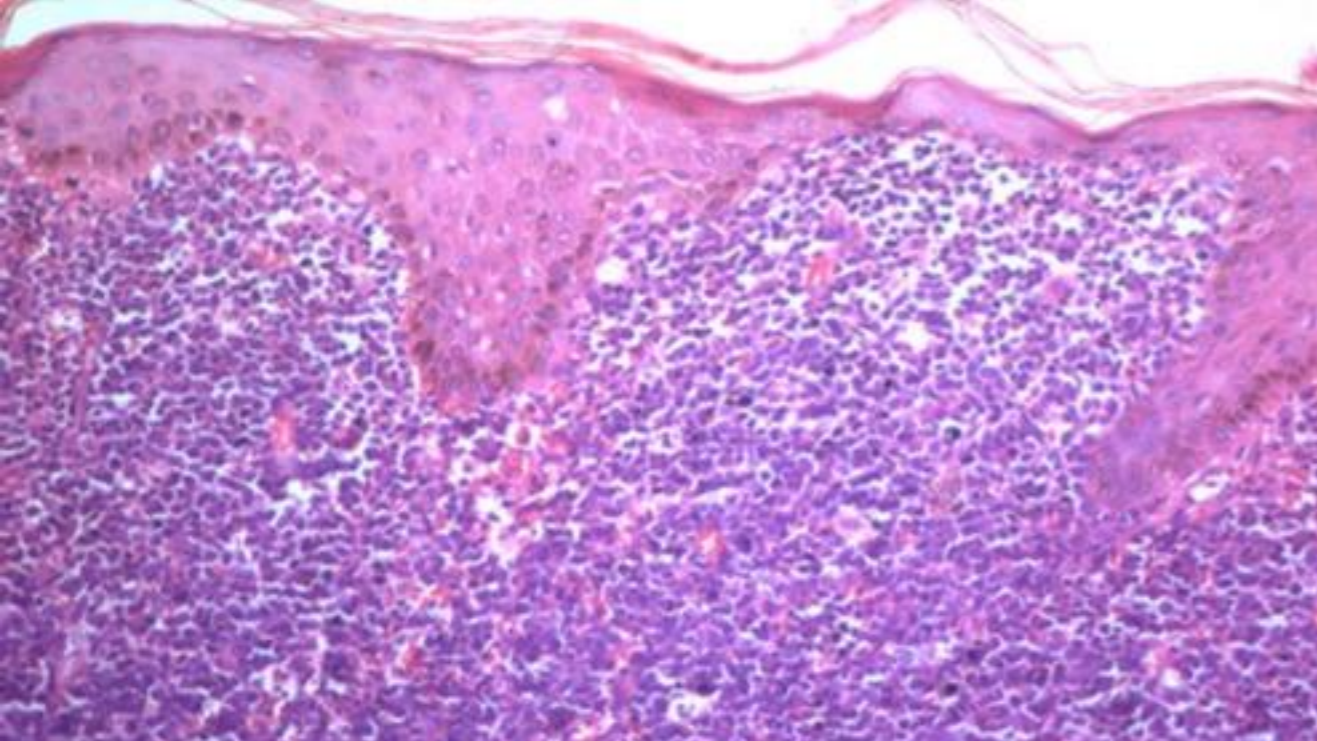
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# Merkel Cell carcinoma

Neuro-Endocrine Ca of the Skin, Small Cell Ca of Skin

[www.merkelcell.org](http://www.merkelcell.org)

## \* AETIOLOGY

\* Age > 65 years ..... average 75 years

\* Fair skin

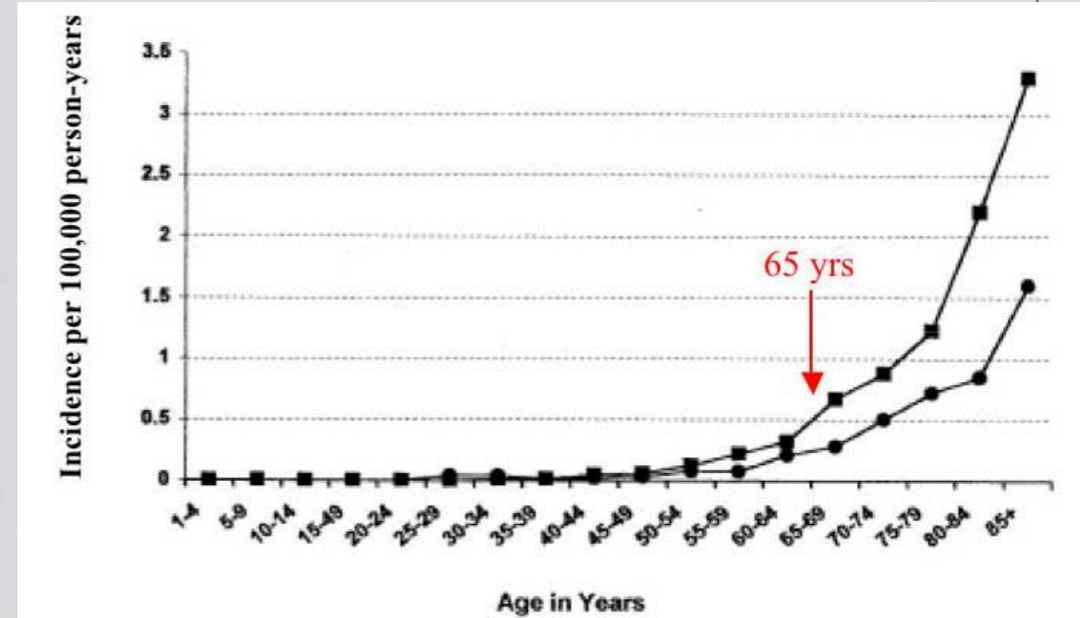
\* Excessive Sun Exposure

\* Chronic Immune Suppression ....x10 transplantation, x8 HIV, x40 CLL, (90% of Merkel patients have normal immunity)

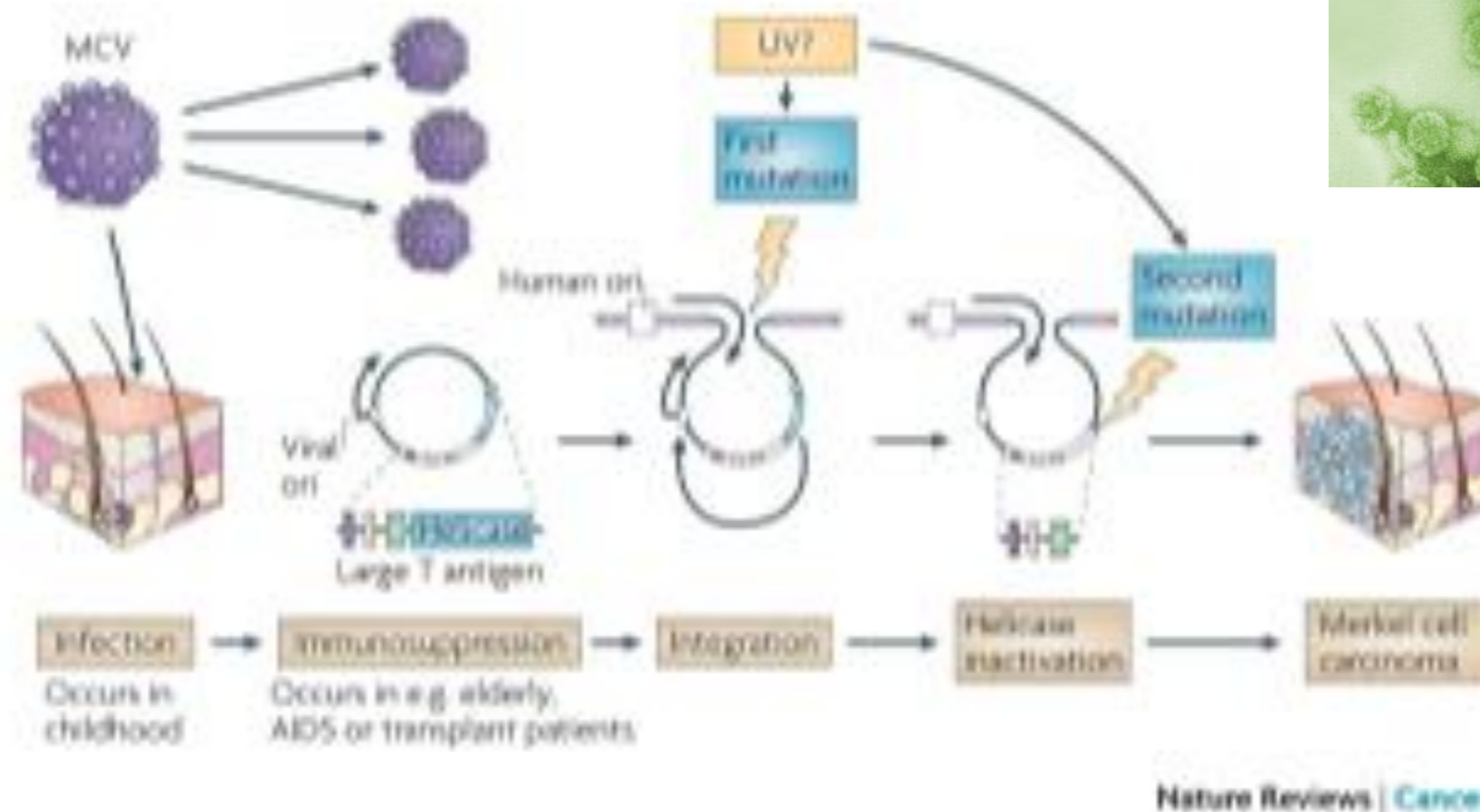
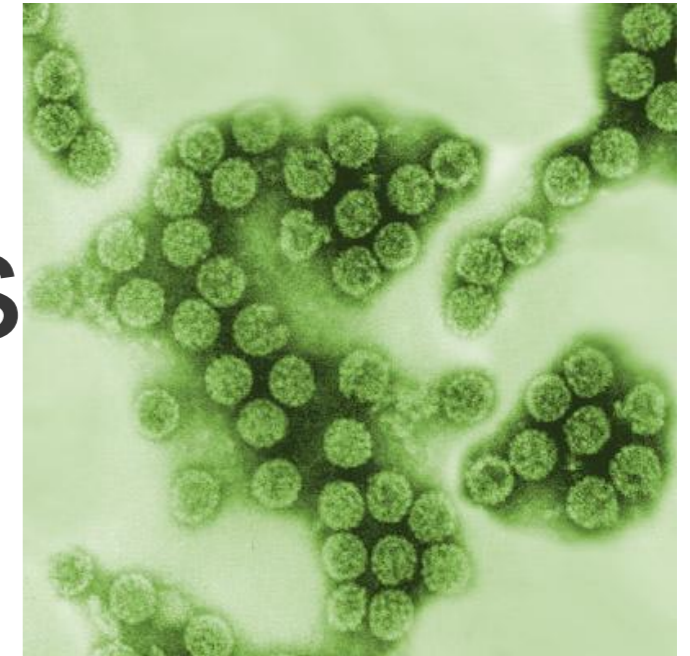
\* Viral ..... **merkel cell polyoma virus (MCPyV)** 80% tumours tested - Feng 2008, (24% of Australian tumours tested ..... Garneski 2008)

\* >50% tumours trisomy chromosome 6, multiple other chromosomal aberrations described

\* High incidence 2nd neoplasm



# Merkel cell polyomavirus



**Merkel cell polyomavirus (MCV)**, which has tumour-specific truncation mutations, illustrates common features among the human tumour viruses involving immunity, virus replication and tumour suppressor targeting. Although MCV is a common infection, **loss of immune surveillance through ageing, AIDS or transplantation and subsequent treatment with immunosuppressive drugs may lead to resurgent MCV replication in skin cells.**

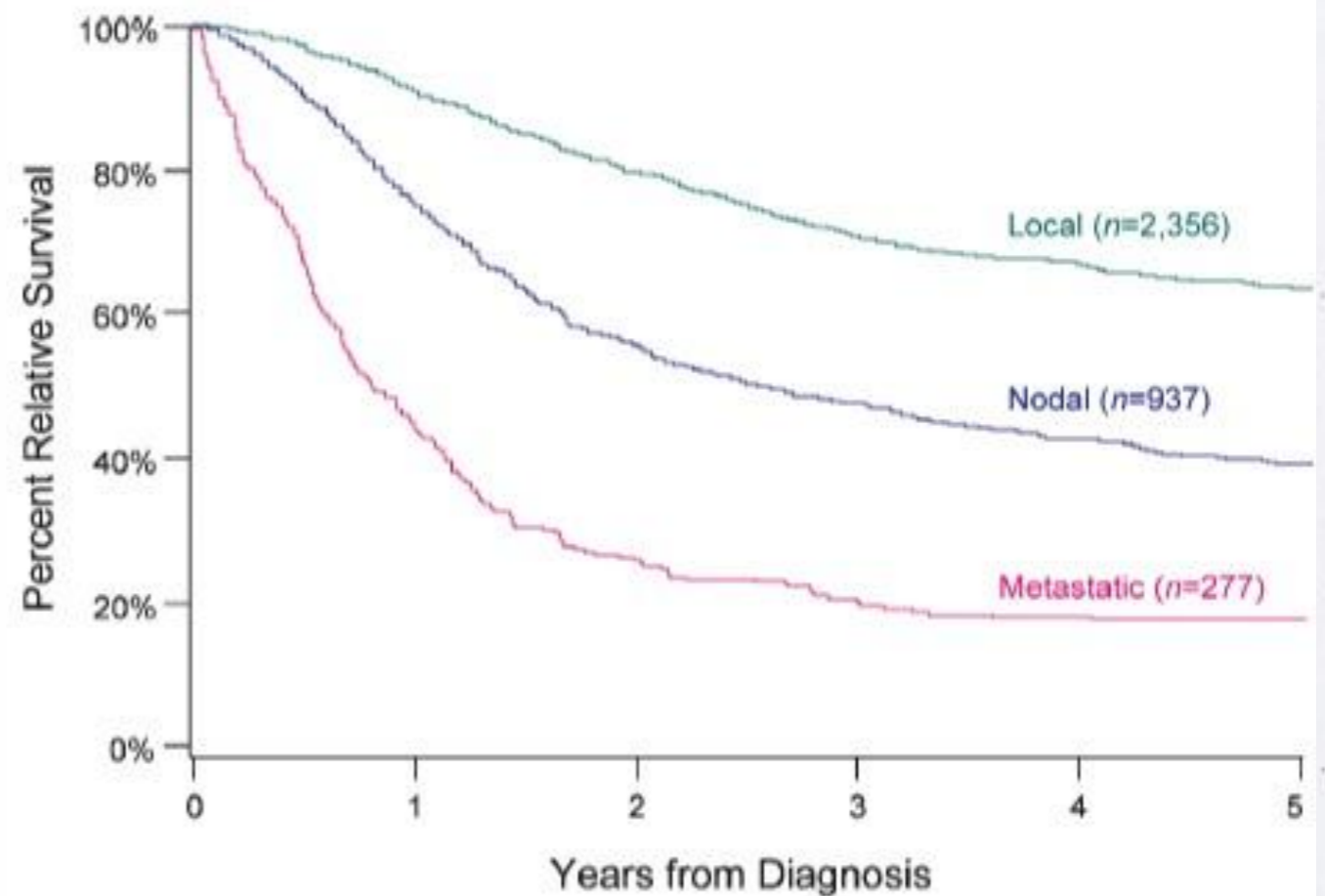
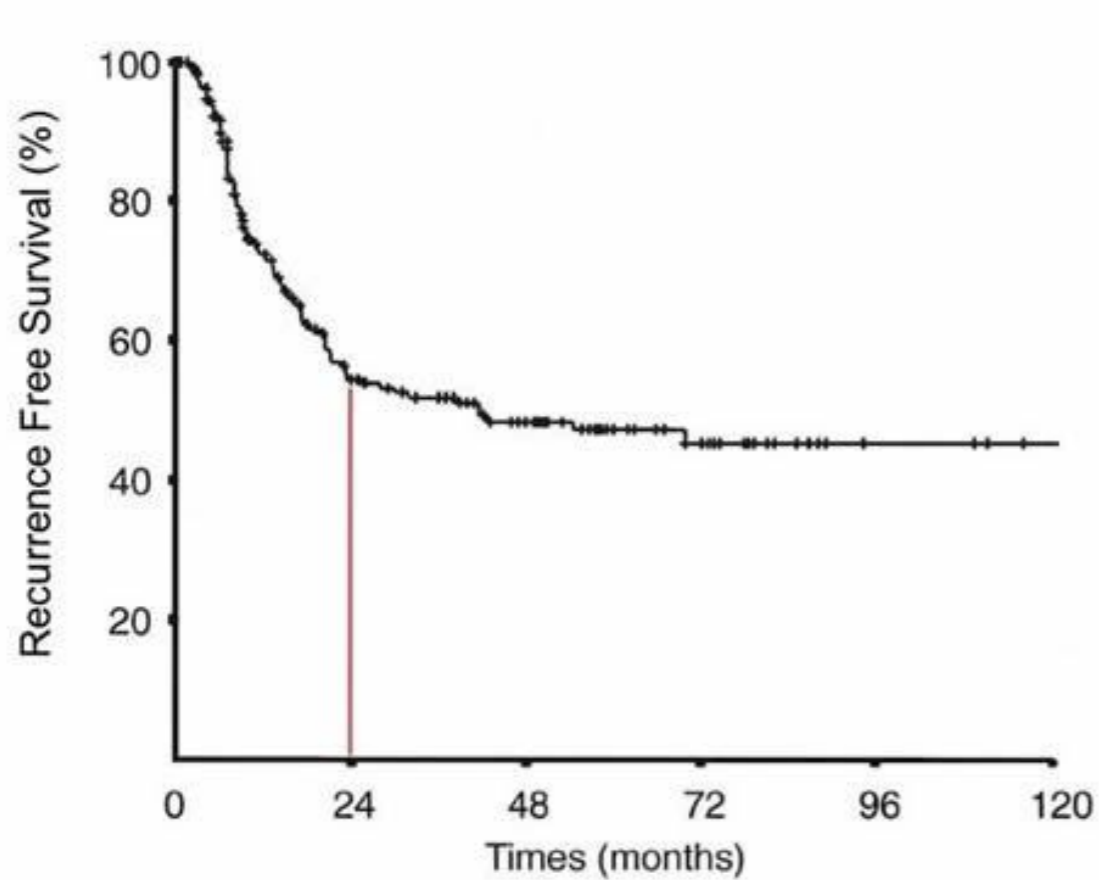
If a rare integration mutation into the host cell genome occurs, the MCV T antigen can activate independent DNA replication from the integrated viral origin that will cause DNA strand breaks in the proto-tumour cell. A second mutation is required for the survival of the nascent Merkel tumour cell.

**Exposure to sunlight** (possibly ultraviolet (UV) irradiation) and other environmental mutagens may enhance the sequential mutation events that turn this asymptomatic viral infection into a cancer virus.



# Merkel Cell carcinoma

## SURVIVAL



# Treatment of Merkel Cell Carcinoma

(aggressive but controversial)

- \* Aggressive
- \* Local
- \* Adjuvant local
- \* Nodes
- \* Wide local excision + nodal adjuvant therapy of nodes in Sentinel Node +ve patients (surgery or XRT)
- \* Narrow margins + adjuvant local/nodal XRT
- \* weedon p883 bottom of page - Rx of MCC

# Merkel Cell carcinoma

- TREATMENT OPTIONS

- \* Surgery to the PRIMARY

- \* Surgery - wide excision + adjuvant XRT (> surgery alone)

- \* +/- SLND [+ve to XRT, 25% -ve relapse]

- \* +/- Local field XRT

- \* +/- Nodal XRT

- \* RADIATION alone (Paulsen M - Bne + Westmead)

- \* Minor excision + XRT local field & nodes

- \* 60% out-of-field relapse, 25% in-field relapse

- \* 37% 5 year survival

# Merkel Cell carcinoma

TREATMENT OPTIONS - Cancer 2007, 110:1-12 - Bichakjian CK

- \* PRIMARY

- \* 1cm margin <2cm tumours

- \* 2cm margin >2cm tumours + XRT local field



# Merkel Cell carcinoma

Cancer 2011, Nov 9 - Fields RC, [Dan Coit](#) - Memorial Sloan-Kettering

- **Recurrence after complete resection and selective use of adjuvant therapy for stage I through III Merkel cell carcinoma.**

\*364 patients

\*Surgery + SLND

\*Adjuvant therapy in young patients, large tumours, lymphovascular invasion, incomplete exc'n, nodal involvement

\*30% developed loco-regional recurrence, (80% of these in clinical node pts or untreated)

# Merkel Cell carcinoma

Ann Surg 2012 Feb;255(2)404 - Fields RC, Dan Coit - St Louis, Missouri

- **Five hundred patients with Merkel cell carcinoma evaluated at a single institution**
  - \*500 patients ..... average age 71 years
  - \*56% 5 year survival,
  - \*Lymph Node +’ve significant in Disease Specific Death



# Merkel Cell carcinoma

J Am Acad Derm 2011 (Oct) - Howie JR et al - Westmead NSW

- **Merkel cell carcinoma: An Australian perspective and the importance of addressing the regional lymph nodes in clinically node-negative patients. 500 patients ..... average age 71 years**

\*136 patients since 1980

\*54% developed recurrence - mostly lymph nodes

\*one third of recurrences in LN where primary only treated

\*62% 5 year survival

\*Significant DFS with adjuvant XRT

# Merkel Cell carcinoma

Am J Otolaryngol 2012 :33 (1) 88-92 - Mendenhall WM - XRT, Gainesville Florida

- **Cutaneous Merkel cell carcinoma**

\*40 patients since 1984

\*37 pts surgery+XRT, one third had chemotherapy as well

\*one third of recurrences in LN where primary only treated

\*79% loco-regional control

5yr outcomes	I	II
loco-regional control	87%	67%
mets-free survival	71%	37%
cause-specific survival	58%	27%
overall survival	48%	18%



# Merkel Cell carcinoma

## ( references )

- \* Int J Radiat Oncol Biol Phys. 2010 Nov 1;78(3):703-9. Epub 2009 Nov 24.
- **The role of radiotherapy alone in patients with merkel cell carcinoma: reporting the Australian experience of 43 patients.** [Veness M](#), [Foote M](#), [Gebski V](#), [Poulsen M](#) Westmead Hospital, University of Sydney,
- \* ANZ J Surg. 2002 Feb;72(2):142-6.
- **Is there a diminishing role for surgery for Merkel cell carcinoma of the skin? a review of current management.** [Poulsen M](#), [Harvey J](#). Mater Queensland Radium Institute, Brisbane, Australia.
- Erratum in **ANZ J Surg.** 2003 Apr;73(4):249.

# Merkel

\* <http://emedicine.medscape.com/article/1295325-overview>

- \* The most important developments during 2010 have involved further explication of the link of a newly identified polyomavirus called Merkel cell polyomavirus (MCPyV), which is found in 60-80% of specimens of Merkel cell cancer, suggesting that virus can play a proximate and pivotal role in the development of skin cancers.[1, 2]
- 1. Mogha A, Fautrel A, Mouchet N, Guo N, Corre S, Adamski H, et al. Merkel cell polyomavirus small T antigen mRNA level is increased following in vivo UV-radiation. *PLoS One*. Jul 2 2010;5(7):e11423. [\[Medline\]](#). [\[Full Text\]](#).
- 2. Mangana J, Dziunycz P, Kerl K, Dummer R, Cozzio A. Prevalence of Merkel Cell Polyomavirus among Swiss Merkel Cell Carcinoma Patients. *Dermatology*. Jul 30 2010;[\[Medline\]](#).
- 3. WIKIPEDIA - Merkel cell polyomavirus - [http://en.wikipedia.org/wiki/Merkel\\_cell\\_polyomavirus](http://en.wikipedia.org/wiki/Merkel_cell_polyomavirus)





# 5 Fierce Tumours

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# Adnexal malignancies

<http://www.nature.com/modpathol/journal/v19/n2s/full/3800511a.htm>

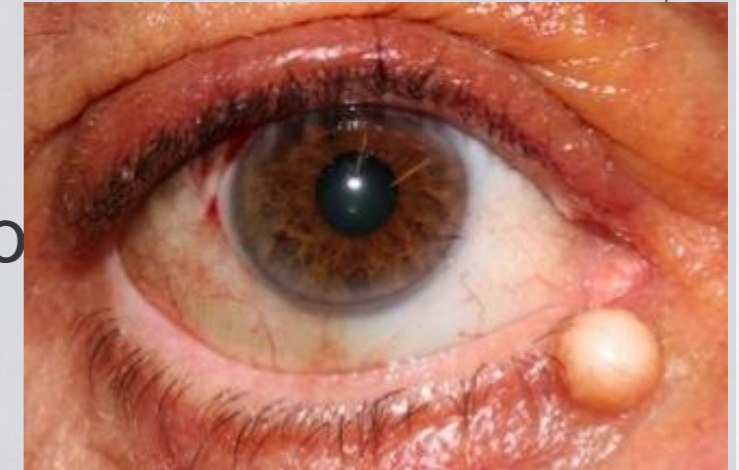
!

\* **Sebaceous** (pilosebaceous)

\* Eccrine

\* Apocrine

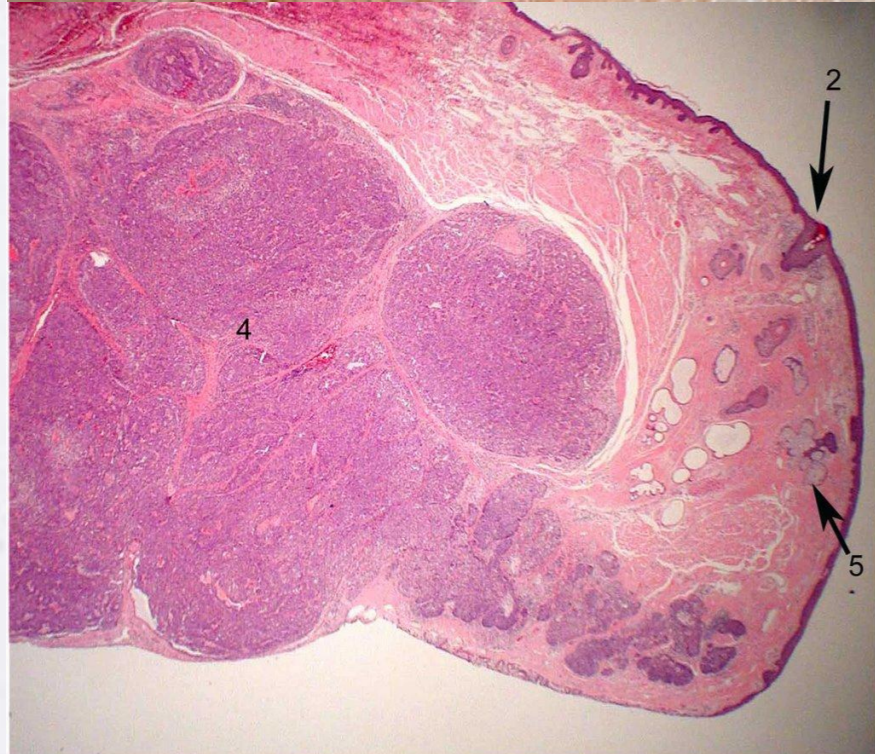
# Sebaceous Carcinoma (Sebaceous)



- \* ocular adnexa ---- meibomian glands, glands of Zeis
- \* masquerade as chalazion
- \* extra-ocular--- scalp, chest, (vulva - Bartholin's gland)
  - \* elderly patients - head & neck
  - \* yellow-tan nodule
  - \* 30% lymph node metastasis
  - \* 20% 5 year mortality
  - \* wide surgical excision, +/- SND / RND, +/- XRT



# Sebaceous Carcinoma (Sebaceous)





# sebaceous gland carcinoma

( reference )

[http://www.mhprofessional.com/downloads/products/0071472568/0071472568\\_chap17.pdf](http://www.mhprofessional.com/downloads/products/0071472568/0071472568_chap17.pdf)



# Adnexal malignancies

<http://www.nature.com/modpathol/journal/v19/n2s/full/3800511a.htm>

\* Sebaceous

\* **Eccrine** - malignant adnexal ca

\* Apocrine



# M.A.C. - Microcystic Adnexal Carcinoma (Eccrine)

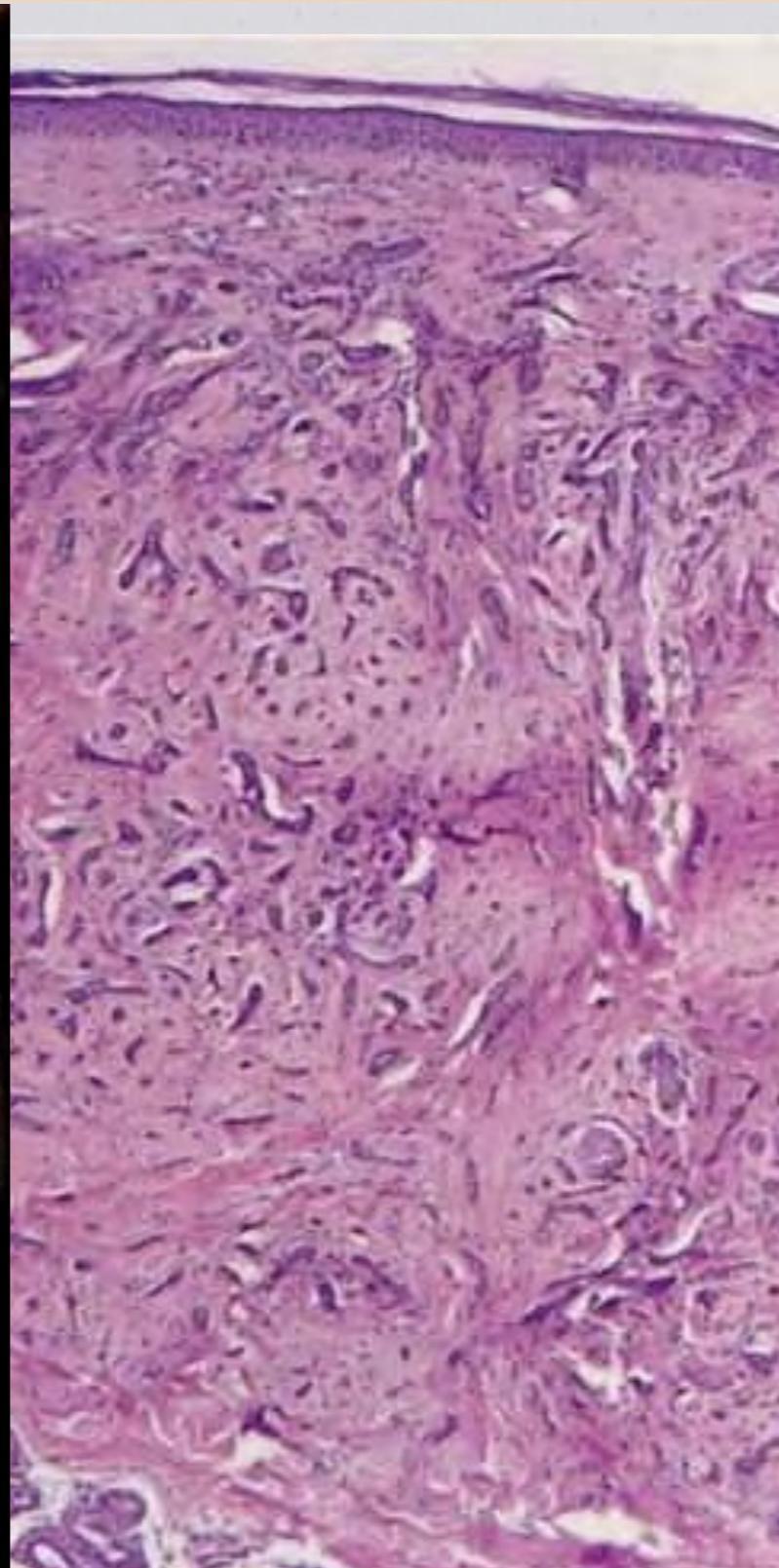
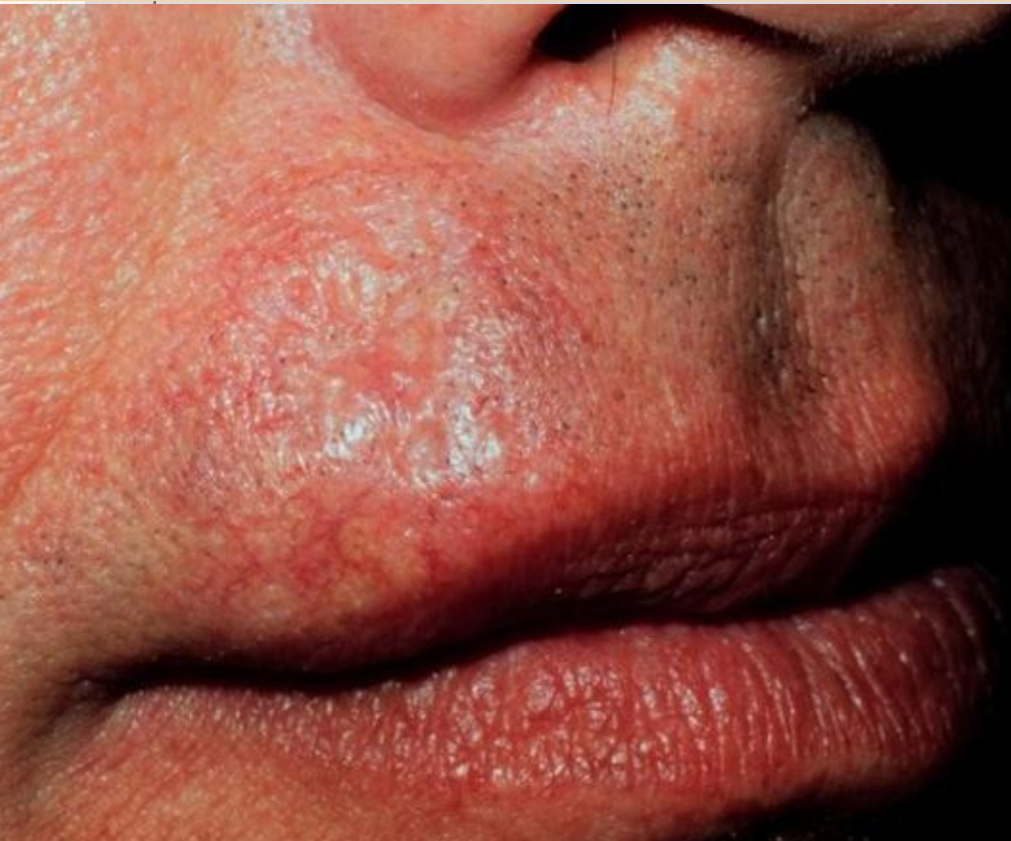
- \* malignant syringoma, sclerosing sweat duct ca
  - \* plaque / nodule on upper lip
    - \* face, extremities, trunk, scalp,
  - \* slow growing, locally aggressive
  - \* adults, all ages / races
  - \* ? prior XRT, immunodeficiency, epithelial naevus
- \* local recurrence >50% [2% / patient year ]
- \* wide exc'n + XRT (BNE 93% cure) v Mohs Dermatology

# M.A.C. - Microcystic Adnexal Carcinoma (Eccrine )





# Microcystic Adnexal ca of lip



Medscape® [www.medscape.com](http://www.medscape.com)





# M.A.C. - Microcystic Adnexal Carcinoma

## (references)

\* J Med Imaging Radiat Oncol. 2010 Oct;54(5):477-82.

- **Microcystic adnexal carcinoma of the skin: the role of adjuvant radiotherapy ----- BRISBANE**
- [Baxi S](#), [Deb S](#), [Weedon D](#), [Baumann K](#), [Poulsen M](#).

- **INTRODUCTION:**

- Microcystic adnexal carcinoma (MAC) is a rare cutaneous tumour where the role of radiotherapy remains undefined. We contrast our institutional experience with current literature, define the local control rate and suggest a role for adjuvant radiotherapy in the treatment of this cancer.

- **METHODS:**

- From 1992-2008, a retrospective review was undertaken for MAC treated with curative intent involving surgery and adjuvant radiotherapy at the Royal Brisbane Hospital and Mater Hospital Brisbane. Clinical, pathologic and treatment details as well as patterns of recurrence were analysed.

- **RESULTS:**

- 14 of MAC received local excision and adjuvant radiotherapy. The median age was 71 years old, with nine of the cases in men. All cases occurred in the head and neck region with mean tumour size of 20.5 mm and mean depth of invasion of 9.9 mm. Peri-neural invasion occurred in 56% of cases and 69% had positive surgical margins. Various adjuvant radiotherapy schedules were used to treat the primary site resulting in a crude local control rate of 93%. Primary and nodal relapses were salvaged with further treatment.

- **CONCLUSION:**

- While Mohs micrographic surgery may be considered the gold standard, wide local excision and adjuvant radiotherapy offers comparable control rates. Doses of 50 Gy or greater should be prescribed with generous margins (3-5 cm) owing to its tendency for peri-neural and deep invasion. There was no evidence that radiotherapy can cause aggressive transformation of the tumour. The role for definitive radiotherapy remains uncertain.



# Adnexal malignancies

<http://www.nature.com/modpathol/journal/v19/n2s/full/3800511a.htm>

\* Sebaceous

\* Eccrine

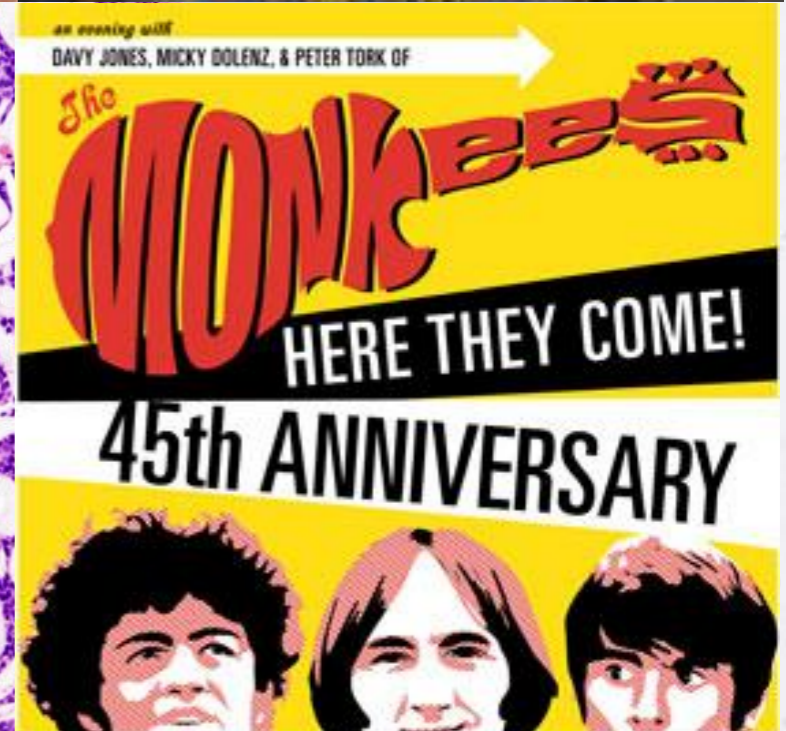
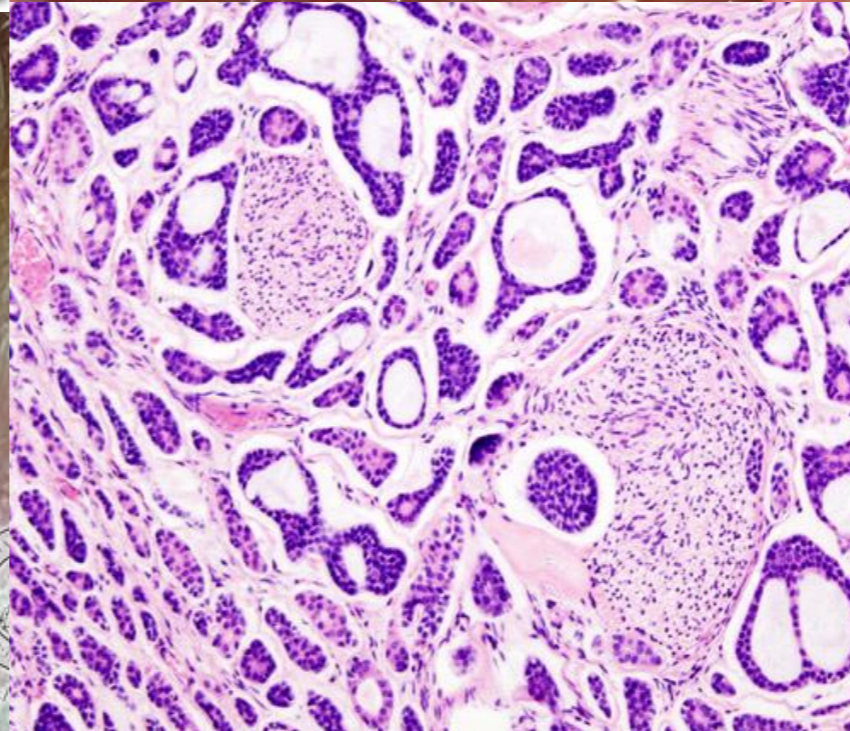
\* **Apocrine** - adenoid cystic carcinoma

# A.C.C. - Adenoid Cystic Carcinoma (Apocrine)

- \* glandular ---- salivary , lacrimal glands, breast, airways,
  - \* (30% of submandibular ca's)
- \* cutaneous --- scalp, chest, (vulva - Bartholin's gland)
  - \* only 50 reported cases
  - \* local recurrence - COMMON
  - \* perineural invasion - COMMON
  - \* metastasis rare
  - \* TREATMENT - wide surgical excision +/- XRT



# A.C.C. - Adenoid Cystic Carcinoma (Apocrine)





# A.C.C. - Adenoid Cystic Carcinoma (Apocrine)



© Waikato District Health Board 2010



© Waikato District Health Board 2010



# A.C.C. - Adenoid Cystic Carcinoma (references)

## \* glandular ----

\* Cancer. 2011 Dec 16. doi: 10.1002/cncr.26740. [Epub ahead of print]

- **A comparison of the demographics, clinical features, and survival of patients with adenoid cystic carcinoma of major and minor salivary glands versus less common sites within the Surveillance, Epidemiology, and End Results registry.**
- [Li N](#), [Xu L](#), [Zhao H](#), [El-Naggar AK](#), [Sturgis EM](#).

## \* cutaneous ---

\* Cutis. 2008 Mar;81(3):243-6.

- **Primary cutaneous adenoid cystic carcinoma: a case report and review of the literature.**
- [Barnes J](#), [Garcia C](#).
- University of Oklahoma College of Medicine, Oklahoma City, USA





# 5 Fierce Tumours

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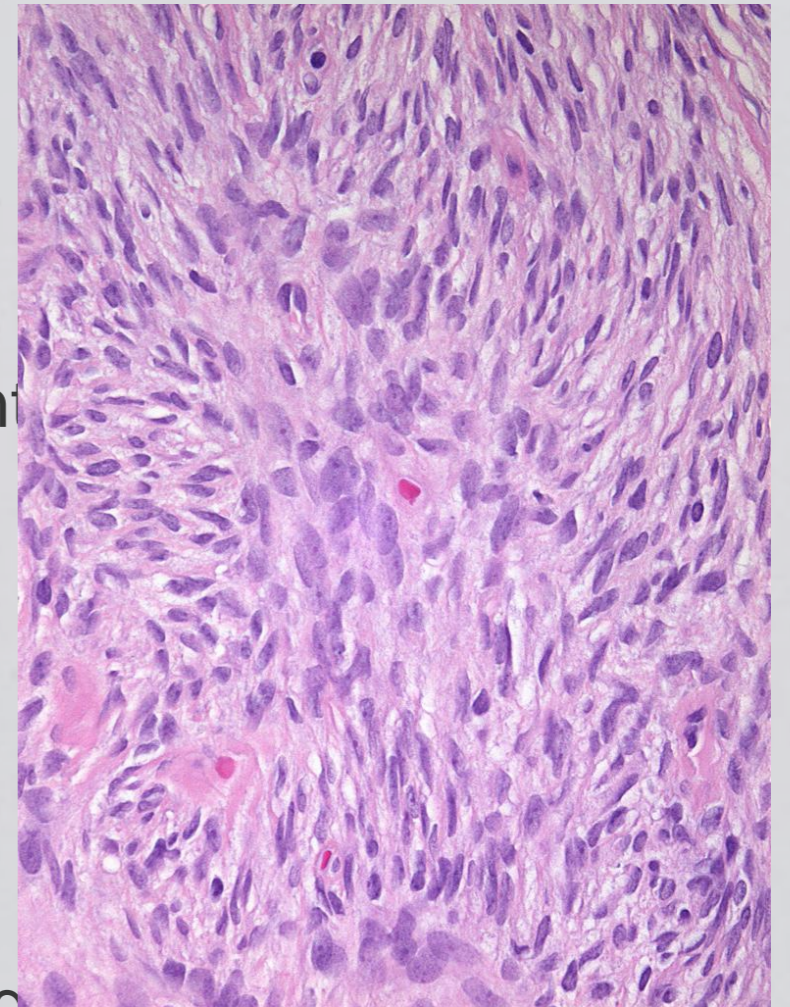
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(5) **M**FH - Malignant Fibrous Histiocytoma, AFX Atypical Fibrous Xanthoma

# D.F.S.P. Dermato Fibro - Sarcoma Protuberans

- \* slow growing, locally aggressive
- \* differentiate from **cellular dermatofibroma**
- \* solitary or multiple nodules - 40% non-protuberant
- \* high local recurrence, rarely metastasizes
- \* **trunk / limbs** of young /middle age adults
- \* may progress to fibrosarcoma or MFH
- \* **SURGERY** - aggressive 5cm margins in width & depth +/- fascia,
  - \* difficulty differentiating between recurrence and scar tissue
  - \* **imatinib** - tyrokinase inhibitor - role in patients with metastasis





# dermatofibroma = fibrous histiocytoma



- \* dermatofibroma

- \* [lower] extremities, shoulder girdle of young [female] adults

- \* **cellular dermatofibroma**

- \* recurrence of prior incomplete dermatofibroma exc'n

- \* young people - rapid growth,

- \* extend deeply and widely

- \* may resemble keloid

- \* SURGERY - wider margins than non



# epithelioid sarcoma

- \* most common sarcoma of distal limbs
- \* simulates a granuloma
- \* slow growing painless nodule on
- \* deep dermal, S/C in origin
- \* WIDE EXC'N - 40% local recurrence  
nodes



FIGURE 7: Epithelioid sarcoma. Solitary palmar nodule



# fibrous histiocyoma

- \* **term** for tumours sharing certain morphological features :
  - \* fibrous-like spindle cells
  - \* histiocytes - cells thought to function as, or transform into a fibroblast
  - \* heterogenous
  - \* histiocyte is a tissue macrophage or dendritic cell



DFSP





\* Cancer Treat Rev. 2008 Dec;34(8):728-36.

dfsp

( references )

- **Dermatofibrosarcoma protuberans: wide local excision vs. Mohs micrographic surgery.**
- [Paradisi A](#), et al. Dermatology Dept. Rome Italy

• **BACKGROUND:**

- Dermatofibrosarcoma protuberans (DFSP) is an uncommon tumor of the skin with high rates of local recurrence. It is debated whether Mohs micrographic surgery (MMS) involves lower recurrence rates than wide local excision (WLE).

• **METHODS:**

- The records of 79 patients with DFSP who underwent WLE (n=38) or MMS (n=41) in 1990-2005 were reviewed retrospectively. The primary endpoint was tumour recurrence rate.

• **RESULTS:**

- Five of the 38 WLE patients (follow-up=4.8 years) had recurrences (13.2%, 95% CI 4.4-28.1%) as opposed to none (95% CI 0-8.6%) of the 41 MMS patients (follow-up=5.4 years). Pooling of these data with those from the literature yielded 6/463 recurrences for MMS (1.3%, 95% CI 0.5-2.8%) and 288/1394 recurrences for WLE (20.7%, 95% CI 18.6-22.9%). The relative risk of recurrence for WLE vs. MMS patients was 15.9 (95% CI 7.2-35.5).

• **CONCLUSIONS:**

- Significantly lower recurrence rates were recorded in our patients subjected to MMS compared with those treated with WLE. There is inconclusive evidence for any advantage of MMS in non-primary cases, while MMS was most effective in treating head and neck tumours.

\* Dermatol Surg. 2012 Jan 30. doi: 10.1111/j.1524-4725.2011.

• **Dermatofibrosarcoma Protuberans: A Review of the Literature.**

- [Bogucki B](#), et al Division of Dermatology, Washington University, Saint Louis, Missouri.





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(5) **MFH** - Malignant Fibrous Histiocytoma, AFX Atypical Fibrous Xanthoma

# Malignant Fibrous Histiocytoma

## \* MFH variants :

- \* angiomatoid = angiomatoid fibrous sarcoma
- \* myxoid = myxofibrosarcoma
- \* giant cell = rare
- \* inflammatory = MFH, ? liposarcoma

## \* pleomorphic = MFH

- \* >5cm in deep soft tissues / proximal limb muscles
- \* adults 50 - 70 years
- \* no predisposing factors
- \* 15-30% 5 year survivals - local recurrence, nodal + lung mets



# MFH - malignant fibrous histiocyoma

J Plast Reconstr Aesthet Surgery 2011 (Nov)64:(11) - Withers AH, Wellington  
NZ

- **Atypical fibroxanthoma and malignant fibrous histiocyoma**

- \*1996-2007 decade - 100 AFX, 15 MFH among  
50,000 lesions

- \*AFX incorrectly mostly Bx only - recurrence  
significant

- \*Immuno-Hx doesn't differentiate between AFX &  
MFH

- \*MFH re-classified as undifferentiated pleomorphic  
sarcoma





# MFH - malignant fibrous histiocytoma

J Plast Reconstr Aesthet Surgery 2011 (Nov)64:(11) - Withers AH, Wellington NZ

	<b>AFX</b>	<b>MFH</b>
site - head & neck	95%	71%
age	77	74
size	16mm	21mm
sun aetiology	+++	+
nodal & distant mets	- (?+)	+++
clearance	1cm	3-5cm
adjuvant XRT	-	+
local recurrence (+/- mets)	7-12%	40-50%

# AFX

- \* rapidly growing red nodule - ulcer
- \* scalp, forehead, ear, neck
- \* BCC / SCC exc'n margin





# AFX

[http://www.globalskinatlas.com/search\\_resultsimg.cfm](http://www.globalskinatlas.com/search_resultsimg.cfm)





# AFX

[http://www.globalskinatlas.com/search\\_resultsimg.cfm](http://www.globalskinatlas.com/search_resultsimg.cfm)





\* J Plast Reconstr Aesthet Surg. 2011 Nov;64(11):

**Atypical fibroxanthoma and malignant fibrous histiocytoma.**

[Withers AH](#), [Brougham ND](#), [Barber RM](#), [Tan ST](#).

**Source**

[Wellington Regional Plastic, Maxillofacial & Burns Unit, Hutt Hospital, Wellington, New Zealand.](#)

**Abstract**

**BACKGROUND:**

Atypical fibroxanthoma (AFX) and malignant fibrous histiocytoma (MFH) are soft-tissue tumours with variable aggressiveness. There is considerable debate about the relationship between these lesions, as histological and immunochemical differentiation is difficult.

**METHODS:**

Current opinions and evidence for diagnostic differences between AFX and MFH were reviewed. Consecutive cases of AFX and MFH were identified from our non-melanoma skin cancer (NMSC) database 1996-2007 for the Central Region of New Zealand.

**RESULTS:**

Of the 50,411 NMSC lesions excised surgically from 26,138 patients, there were 101 AFX and 15 MFH cases. Three MFH cases were originally diagnosed as AFX. AFX and MFH share similar patient demographics, size and location and histological and immunohistochemical features. Most diagnostic biopsies of AFX were not followed by formal excision. Incomplete excision occurred in a large proportion of patients with AFX, which often did not proceed to re-excision, resulting in local recurrence. Cases of MFH generally underwent definitive treatment including re-excision if incompletely excised, and postoperative adjuvant radiotherapy.

**CONCLUSIONS:**

The failure to treat AFX adequately may have resulted from the lack of appreciation of its aggressiveness. Contrary to the literature, we found few clinical differences between AFX and MFH. AFX and MFH also share similar histologic features and there are no immunohistochemical markers that reliably distinguish them. AFX is best considered a distinct entity with MFH, now reclassified as an undifferentiated pleomorphic sarcoma.





# 5 Fierce Tumours

(1) **M**erkel Cell Carcinoma

(2) **A**ppendageal Tumours

- Sebaceous - Sebaceous Gland Carcinomas
- Eccrine - Microcystic Adnexal Carcinoma
- Apocrine - Adenoid Cystic Carcinoma

(3) **D**FSP - Dermato Fibro Sarcoma Protuberans

(4) **A**ngiosarcoma // Kaposi's Sarcoma

(5) **M**FH - Malignant Fibrous Histiocytoma, AFX Atypical Fibrous Xanthoma

# angiosarcoma

- \* involves upper part of face or scalp of elderly people
- \* may occur >10 years post-irradiation
- \* Men >> women
- \* multifocal, bluish violaceous nodules
- \* thrombocytopenia with metastasis
- \* extensive local growth --- advanced presentation
- \* metastasis to regional nodes & lungs
- \* poor prognosis <15% 5year survival



# angiosarcoma



FIGURE 2:  
Cutaneous  
angiosarcoma.  
Ulcerated  
erythema-  
tous-viola-  
ceous  
plaque on  
scalp



FIGURE 1: Angiosarcoma on chronic lymphedema (Stewart-Treves syndrome). Violaceous nodule on upper limb



# kaposi's sarcoma

- \* 4 types: classic, african, HIV, immunosuppression related,
- \* HHV-8 human herpesvirus type 8 aetiology in all types
- \* 10% of AIDS patients now develop KS
- \* distribution in HIV = multiple lesions on trunk, limbs, H&N
- \* visceral lesions also, mucosal surfaces & internal organs
- \* most patients with KS die from opportunistic infections

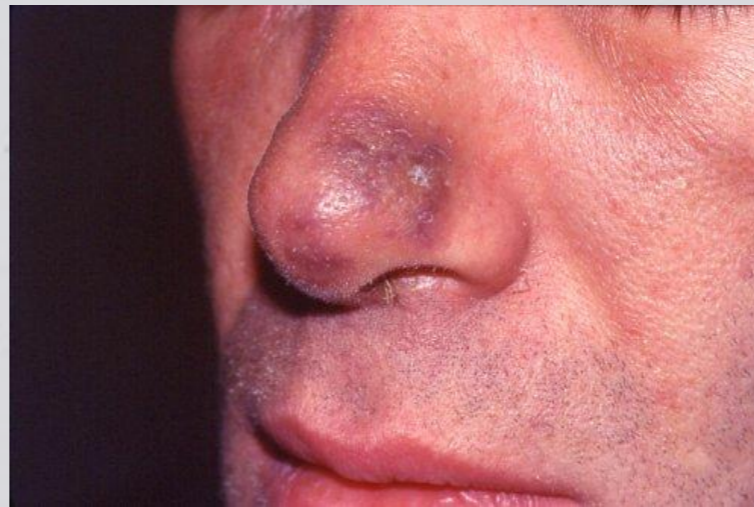
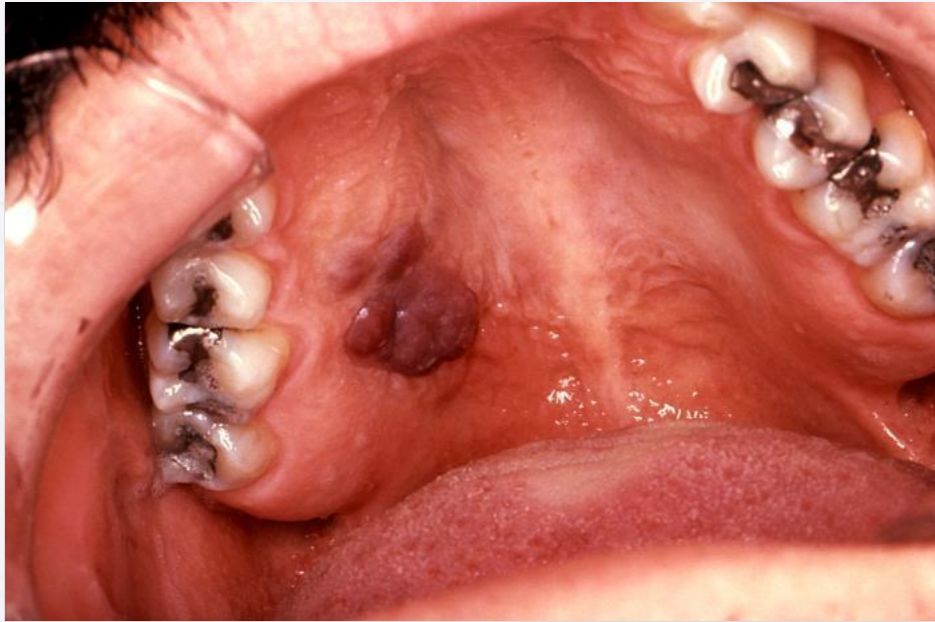


# kaposi's sarcoma rx

- \* Treatment :
  - \* symptom palliation
  - \* shrinkage of tumour
    - \* surgery for solitary lesions
    - \* XRT
    - \* Aldara 50% response rate
  - \* prevention of disease progression
  - \* anti-retroviral therapy
    - \* Doxorubicin
    - \* Paclitaxel



# Kaposi's





# Kaposi's



\* Malignant Glomus Tumour



# 5 Fierce Tumours



**M**erkel Cell Carcinoma



**A**ppendageal Tumours

- **S**ebaceous - **S**ebaceous Gland Carcinomas
- **E**ccrine - **M**icrocystic Adnexal Carcinoma
- **A**pocrine - **A**denoid Cystic Carcinoma



**D**FSP - Dermato-Fibro-Sarcoma Protuberans



**A**ngiosarcoma // Kaposi's Sarcoma



**M**FH - Malignant Fibrous Histiocytoma, **A**FX Atypical Fibrous Xanthoma



