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Review

Malignant cutaneous adnexal tumours of the head and neck: an update on management

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Abstract

Adnexal tumours form a heterogeneous group of relatively rare neoplasms. Many of them have a poor prognosis and treatment can sometimes be difficult and controversial. We summarise the latest publications relating to malignant cutaneous adnexal tumours of the head and neck, and give an update on their management. We discuss Merkel cell carcinoma and other rare malignant adnexal tumours including dermatofibrosarcoma protuberans and atypical fibroxanthoma.

Keywords: Review; Adnexal tumours; Head and neck

Introduction

In the United Kingdom, skin cancer is managed by specialist multidisciplinary teams of which maxillofacial surgeons are usually core members. Most skin cancers are basal cell carcinoma, squamous cell carcinoma, or melanoma, but it is important to understand the management of less common cutaneous malignancies (Table 1). We review the heterogeneous group of adnexal cancers of the skin, and outline their management.

Merkel cell carcinoma (MCC)

Merkel cells contain neurosecretory granules and are found on the basal layer of the epidermis.[1] Previously, MCC was described as trabecular carcinoma or cutaneous neuroendocrine carcinoma.[1] Its incidence is higher in men than in women (2:1)[2] and it is rare in young people with most cases occurring in those over 50 years of age.[3] White-skinned people have the highest risk. As with other non-melanoma skin cancers, its incidence is high in areas exposed to ultraviolet (UV) light and the sites most commonly affected are on the head and neck (48%).[4]

Although its cause is poorly understood, UV light is implicated and has been identified in cases in immunocompromised patients with autoimmune and iatrogenic diseases; partial regression of metastases has been reported after immunosuppression has been stopped.[5–8] In patients having psoralen-UV treatment for psoriasis its incidence is about 100 times higher than that seen in the general population,

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Table 1
Number of cases of cutaneous adnexal tumours reported in England in 2012.

Adnexal tumour	No. of cases
Merkel cell carcinoma	227
Microcystic adnexal carcinoma	23
Sebaceous carcinoma	115
Malignant fibrous histiocytoma/pleomorphic sarcoma	110

which indicates that both UVA and UVB radiation have a role.[9]

Viral infection could also be an important factor.[10] A new virus, Merkel cell polyomavirus (MCPyV) has been found in 80% of patients over 50,[11] and although little is known about transmission and latency, its prevalence in MCC lesions has been reported to range from 40% to 100%.[12,13] As it is present in both primary and metastatic disease,[14] it can be argued that MCPyV is a carcinogen.[15]

Commonly, MCC develops as an asymptomatic, rapidly growing, flesh-coloured, firm, and non-tender nodule but plaque-like variants also occur (Fig. 1).[16] The vascular pattern consists of polymorphic vessels. To act as an aide-mémoire for its clinical characteristics, the acronym "AEIOU" has been suggested: asymptomatic, rapidly expanding, immunosuppressed, older patient, UV exposure.[16] Differential diagnoses can include atypical fibroxanthoma, lymphoma, and amelanotic melanoma.

Since its description, several staging systems have been suggested and one by the American Joint Committee on Cancer in 2010 has been generally accepted (Tables 2 and 3). Recurrence has been reported to be between 25% and 30%. Regional lymph nodes are reported to be involved in 52% to 59%, and distal metastases in 34% to 36% of all cases, and as MCC has higher metastatic and mortality rates than melanoma, treatment needs to be aggressive.[17–20]

Operation is the primary treatment.[9] Some surgeons support the use of wide excision margins (1–2 cm), and Mohs micrographic surgery (MMS) has been suggested as a viable alternative as local recurrence rates were comparable to those after wide excision.[21] Further treatment depends on the route

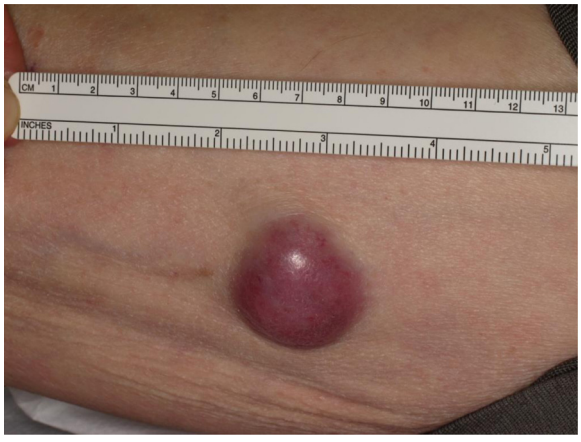

Fig. 1. Merkel cell carcinoma.

Table 2
TNM criteria for Merkel cell carcinoma (based on Lemos et al.[20]).

T:	
Tx	Primary tumour cannot be assessed
T0	No primary tumour
Tis	In situ primary tumour
T1	Primary tumour less than 2 cm
T2	Primary tumour more than 2 cm but less than 5 cm
T3	Primary tumour more than 5 cm
T4	Primary tumour invades bone, muscle, fascia, or cartilage
N:	
Nx	Regional nodes cannot be assessed
N0	No regional nodal metastasis
cN0	Nodes not clinically detectable
cN1	Nodes detected clinically
pN0	No nodal disease on pathological evaluation
pNx	Nodes not evaluated pathologically
N1a	Micrometastases
N1b	Macrometastases
N2	In-transit metastases
M:	
Mx	Distant metastases cannot be assessed
M0	No distant metastases
M1	Distant metastases
M1a	Distant skin, distant subcutaneous tissue or distant lymph node metastases
M1b	Metastases to lung
M1c	Metastasis to other visceral sites

of drainage of the primary lymph nodes.[22] Overall 5-year survival is around 62%.[23] Five-year survival for patients with local disease is 64%, but for those with nodal disease and distant metastases it is only 39% and 18%, respectively. The low rate for local disease is the result of the discrepancy between T1 lesions (79% 5-year survival) and T4 lesions (47% 5-year survival), which both fall into this category.[20]

There is also a mucosal variant but this has a much lower incidence (4.5%).[21] It most commonly affects the larynx followed by the nasal cavity, pharynx, mouth, and tongue, and has a poorer prognosis than the cutaneous type. Overall survival is 49% at 2 years.[23]

As metastases are common in MCC, and microscopic disease has been found in up to 50% of patients who have elective neck dissection,[24] there is much discussion about appropriate management when there are no sign of metastases in the neck. Gillenwater et al.[19] reported that recurrence developed in 23/24 patients who had presented with a clinically N0 neck

Table 3
Merkel cell staging (based on Lemos et al.[20]).

Stage	Stage grouping (TNM)		
0	Tis	N0	M0
IA	T1	pN0	M0
IB	T1	cN0	M0
IIA	T2/T3	pN0	M0
IIB	T2/T3	cN0	M0
IIC	T4	N0	M0
IIIA	Any T	N1a	M0
IIIB	Any T	N1b/N2	M0
IV	Any T	Any N	M1