CASE REPORT

Primary cutaneous mucoepidermal carcinoma

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SUMMARY

Mucoepidermal carcinoma (MEC) is a tumour having mixed components of mucus secreting and epidermoid cells. Salivary glands are the most common site of origin. Primary cutaneous MEC is a rare presentation. We report a primary cutaneous MEC in a 98-year-old woman presenting a noduloulcerative lesion over the dorsum of the nose. Histopathology of the tumour showed nests of epidermoid cells with glandular differentiation and mucin production. The diagnosis was confirmed by immunohistochemistry.

BACKGROUND

Mucoepidermal carcinoma (MEC), as the name suggests, is a carcinoma which has mucus-secreting and epidermoid cells. MEC arises mostly from salivary gland with parotid gland being the usual site of involvement. Other sites which may develop primary MEC are lacrimal passages, sinonasal tract, oesophagus, bronchi, thymus, thyroid, breast, pancreas and prostate.¹ Cutaneous involvement may be metastatic or primary. Primary cutaneous mucoepidermal carcinoma (PCMEC) is a rare presentation of MEC. We present a 98-year-old woman presenting with a slow-growing cutaneous

Figure 2 Histopathology showing nests of epithelioid cells with microglandular lumina (H&E, 10x).

lesion on the face which was histopathologically diagnosed and confirmed by immunohistochemistry to be PCMEC.

CASE PRESENTATION

A 98-year-old woman presented in the dermatology outpatient department with a lesion at the dorsum of the nose which was present for 2 years and was gradually progressing since then. One year later, the patient developed ulceration over the lesions which was asymptomatic but later developed mild pain, for which she presented to us. There was no history of any pre-existing lesion or history of manipulation with thorn. On examination, over the dorsum of the nose, there was a single well-defined erythematous nodule of approximately 2 cm x 1.5 cm with central ulceration with yellowish crusting and irregular surface ([figure 1](#)). There was no lymphadenopathy. Systemic examination was unremarkable.

INVESTIGATIONS

Routine haematological and biochemical parameters were normal. Histopathology showed mildly acanthotic epidermis; the dermis revealed tumour epithelial cells forming irregular nests with microglandular lumina and surrounding wispy mucus ([figure 2](#)).

Nest of tumour cells had mildly pleomorphic round, vesicular nuclei and moderate eosinophilic cytoplasm forming microglandular lumina at the periphery; in the centre, keratinised squamous cells were seen (H&E, 40x; [figure 3](#)). Periodic acid schiff (PAS) positive intracytoplasmic granules and extracellular mucin (100x) were seen ([figure 4](#)). These findings were suggestive of

Figure 1 Noduloulcerative lesion on the dorsum of the nose.

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Figure 3 Mildly pleomorphic round, vesicular nuclei and moderate eosinophilic cytoplasm forming microglandular lumina at the periphery (H&E, 40x).

MEC. Immunohistochemistry for p63 was positive. Radiological examination of chest, abdomen, neck and sinonasal tract was within normal limits.

DIFFERENTIAL DIAGNOSIS

Adenosquamous carcinoma (ASC) though occasionally used synonymously is reported to have a more aggressive course and histopathologically more keratinocyte atypia, intraepidermal origin and less of mucin production. Metastatic MEC normally

Figure 4 (H&E, 40x) Periodic acid schiff (PAS)-positive intracytoplasmic granules and extracellular mucin (100x).

will have presenting complaints of the primary site and is more aggressive. Histopathology of primary and metastatic cutaneous MEC may be identical though the latter shows more keratinocyte atypia. p63 positivity is seen in PCMEC which differentiates it from metastatic variant. A final diagnosis of primary cutaneous MEC was made.

TREATMENT

Patient was advised excision but she refused.

OUTCOME AND FOLLOW-UP

The patient did not return for follow up because of her age and indolent course of disease.

DISCUSSION

MEC accounts for 10%–30% of salivary gland malignancies.² Stewart first described MEC of parotid in 1945.³ There are case reports of origin from other organs like sinonasal tract, bronchus, oesophagus, thyroid, thymus, etc.¹ Skin as the primary site of origin is a rare entity, the etiopathogenesis of which is not very clear. It may arise de novo or on a pre-existing nevus sebaceous.⁴ It is speculated that PCMEC arise from ectopic salivary glands which are derived from sweat glands.⁵ Landman *et al* proposed a sweat gland origin hypothesis for PCMEC.⁶ Galleger first described PCMEC in 1959, and since then, only 20 cases have been reported in the literature.⁵ PCMEC is more common in men and seen in the sixth to eighth decade and very rarely has been seen in children.⁷ Head and neck are the usual sites involved, though the axilla, vulva, finger and foot may rarely be involved. Clinically, PCMEC presents as a slowly growing nodular erythematous to skin-coloured nodule. Secondary ulceration may be present. It mimics basal cell carcinoma clinically. Histopathologically, there are components of epidermoid cells showing atypia along with clear cells and mucin, which can be demonstrated by PAS staining.⁸ ASC and PCMEC were thought to be identical conditions, but Riedlinger *et al* in the early 2000s explained that the two conditions have a different clinical course and histopathology.⁹ PCMEC are low-grade tumours with less risk of metastasis contrary to ASC. Histopathologically, MEC are more dermal-based tumours with abundant mucin-producing cells, papillary features and peritumorous fibrosis while ASC are predominantly intraepidermal with less mucin-producing cells and more carcinomatous cells. Histopathologically, the aggressiveness of the tumour is determined by the proportion of various type of cells (squamous, mucin secreting, intermediate and clear cells) and their maturation.

MEC of the skin may be primary, secondary to contiguous spread or metastatic from a distant site. Clinical and radiological examinations are inconclusive in PCMEC, but immunohistochemistry shows p63 positivity which helps in confirming the diagnosis.¹⁰ Depending on the clinic histopathological aggressiveness of tumour, the various treatment

Learning points

- ▶ Primary cutaneous mucoepidermal carcinoma (PCMEC) is a very rare mucoepidermal carcinoma.
- ▶ Primary and metastatic MEC have to be differentiated.
- ▶ PCMEC is a variable grade malignancy and treatment needs to be tailored accordingly.

modalities are excision or in a high-grade tumour radiotherapy may be required.¹¹

Contributors MG has done definition of intellectual content and manuscript review, AnG has searched the literature, KV did manuscript editing and ArG has prepared the manuscript.

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