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Material submitted: onehistology H&E slide
Title of Case: ***ENDOCRINE MUCIN PRODUCING SWEAT GLAND CARCINOMA (EMPSGC); Aduty of care to rule out a metastasis?***

CLINICAL HISTORY: 70-year-old Caucasian male. Nodular lesion right upper lid.

MACROSCOPY: A triangular smooth piece of skin (6x5x3mm) with a central nodular area

MICROSCOPY
Dermis extensively infiltrated by a nodular epithelial neoplasm with solid, cribiform and nested arrangement. Tumour cells show a bland uniform appearance withmedium-sized round to oval nuclei featuring fine granular or stippledchromatin. One (1) mitotic figure was seen in the entire lesion. No necrosis is seen. Some peripheral palisading with no clefting,No lymphovascularor perineural invasion is seen. There was no tumour connected with surface epithelium.

Stains: Mucin deposition seen withAlcian bluewith intracytoplasmic and extracellular deposit.

Immunohistochemical stains:
Positive: NSE, Chromogranin, Estrogen, Progesterone, Androgen, GATA3, GDFP, Berp4
Negative: S100, Ck20, Inhibin, SOX10, P40, Adipophilin, CD117, P63, PLAG1, Ckit

DIAGNOSIS- Endocrine mucin producing sweat gland carcinomaEMPSGC (in the right clinical and radiological context).

DISCUSSION–
Endocrine mucin-producing sweat gland carcinoma (EMPSGC) is a rare,low-grade, cutaneous adnexal lesion with neuroendocrine differentiation first described in 1997

It has a predilection for the skin of the eyelid, but has also been reported in the face and rarely extra-facial locations cheek, scalp, peri auricular, temple, chest wall). The tumour is seen more frequently in women and on average affects the elderly.

It is considereda precursor of invasive neuroendocrine type primary cutaneous mucinous carcinoma (PCMC), which is associated with a low-grade malignant behaviour. Up to 2020 there are about 190 cases reported in the literature.

It is histologically and immunohistochemically analogous to solid/papillary carcinoma of the breast/endocrine ductal carcinoma in situ with a nodular, solid, papillary, and/or cribriform architecture, neuroendocrine differentiation, and mucin production.

The morphological and immunohistochemical profile may be indistinguishable from a breast and salivary gland primary tumour.

EMPSGC is considered indolent in nature; though recurrences may occur, metastases have not been reported so far.
As sweat glands and breast tissue share a common embryological origin, it is not uncommon to find analogous tumours at these sites.
Recently it has been shown by next generation sequencing analysis suggesting a varied multistep mutational pathogenesis or EMOSGC MUC2 positivity suggesting a conjunctival origin.

DIFFERENTIAL DIAGNOSES:
Nodular mucinous hidradenoma (scalp): variable cystic and solid conformation but an absence of neuroendocrine differentiation, attachment to the epidermis and vascular spaces favour hidradenoma and a greater diversity of cell types including eosinophilic polygonal cells, squamous cells, clear cells and goblet cells
Paraganglia: Features against: S100-ve, inhibin –ve, Cytokeratin +
Granular cell tumour: Features against CD68-ve, S100-ve
Alveolar soft tissue sarcoma/Melanoma: S100, Melan A, SOX10
Metastases from common locations including breast, salivary gland and pancreas.

OBJECTIVE:

A. Awareness of a rareeyelid lesion; consideration needs to be given to the possibility of a metastases rather than a primary lesion (mainly breast &salivary gland).
B. Eyelid biopsies sometimes only include a limited amount of superficial tumour material. Basal cell carcinoma/Merkel cell carcinoma and benign adnexal tumours could beconsidered as differentials in a small biopsy.

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