Slide diagnosis/topic: Endocrine mucin-producing sweat gland carcinoma

Case number: MTT 4

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Image link:

Case vignette: 68 year old woman presents with a slowly enlarging lesion on the left lower eyelid. Clinical impression was ‘hidrocystoma’.

Discussion of slide diagnosis: Sections reveal skin with a proliferation of basaloid cells arranged as expansile multinodular nests in the deep dermis with adjacent small pools of mucin. The tumor cells appear cytologically bland with round-oval nuclei with finely granular stippled chromatin and variable amounts of pale eosinophilic cytoplasm. Adjacent to this, there are small pools of mucin containing smaller islands of tumor cells floating within these mucin pools.

Endocrine mucin producing sweat gland carcinoma (EMPSGC) most commonly occurs in older women as a slow growing dermal/subcutaneous nodule around the eyelid. Histologically, EMPSGC are well-circumscribed multinodular tumors containing variable compositions of solid and cystic components with occasional papillary projections. Both intracytoplasmic and extracellular mucin is typically present in EMPSGC. Immunohistochemical studies are informative as EMPSGC express at least one neuroendocrine marker, including synaptophysin or chromogranin. In addition, EMPSGC expresses estrogen and progesterone receptors and cytokeratin 7 and are negative for cytokeratin 20, melanocytic antigens. The common co-existence of EMPSGC and mucinous carcinoma has led to the proposal that EMPSGC is a precursor of or on a continuum with mucinous carcinoma, and stains for myoepithelial markers may show surrounding myoepithelial cells surrounding the nests of EMPSGC.(1-3)

Question 1: The best diagnosis is:

A. Basal cell carcinoma
B. Squamous cell carcinoma
C. Invasive sebaceous carcinoma
D. Xanthoma
E. Endocrine mucin-producing sweat gland carcinoma

Question 1: The best diagnosis is:

A. Basal cell carcinoma—Incorrect. Basal cell carcinoma is uncommon in the anus, but consists of nests of basaloid cells (with peripheral palisading of the basaloid cells) infiltrating the underlying dermis with a characteristic mucinous stromal reaction and retraction artifact.
B. Squamous cell carcinoma—Incorrect. Squamous cell carcinomas (SCC) typically arise from the overlying epidermis and consist of variably sized islands of atypical keratinocytes (usually with evidence of keratinization) invading the underlying dermis.
C. Invasive sebaceous carcinoma—Incorrect. Sebaceous carcinoma most commonly arises with an associated in situ component, and the tumor cells variably show intracytoplasmic vacuoles which impinge/scallop the nucleus. Nuclear atypia is usually prominent in sebaceous carcinoma, and islands of tumor cells floating in extracellular mucin is uncommon in sebaceous carcinoma.
D. **Xanthoma—Incorrect.** Xanthomas are usually multiple small papules around the eyelids and consist of sheets of foamy histiocytes.

E. **Endocrine mucin-producing sweat gland carcinoma—Correct.** Sections reveal skin with a proliferation of basaloid cells arranged as expansile multinodular nests with pushing borders in the deep dermis with adjacent small pools of mucin. The tumor cells appear cytologically bland with round-oval nuclei with finely granular stippled chromatin and variable amounts of pale eosinophilic cytoplasm. Adjacent to this, there are small pools of mucin containing smaller islands of tumor cells floating within them.

**References:**