Case 1: Metastatic adenoid cystic carcinoma

I. Adenoid cystic carcinoma (ACC) can present as a primary or metastatic tumor in the skin
   a. 2nd most common salivary gland carcinoma
   b. Also can develop in sinuses, breasts, respiratory tract,

II. Histopathologic features
   a. Cribriforming, with gland and pseudo gland formation, “swiss cheese” appearance
      i. Other subsets include solid and tubular
   b. Mucin within glands
   c. Cells are rounded, small, without single-cell necrosis
   d. No epidermal connection
   e. Can exhibit epithelial and myoepithelial cells.

III. Differential diagnosis
   a. Adenoid basal cell carcinoma- single cell necrosis, epidermal connection, varied sizes of pseudoglands. Peripheral palisading.
   b. Spiradenoma or cylindroma with cribriforming- should be only focal. No atypia. Better circumscribed.
   c. Microcystic adnexal carcinoma- cysts and ducts, deeply infiltrating, bland cytology.
   d. Desmoplastic trichoepithelioma- calcifications, pseudohorn cysts, no deep infiltration, central dell, folliculosebaceous differentiation

IV. Immunohistochemical stains
   a. S100/SOX10—remember myoepithelial cells can express these markers, not just cells of neural origin
   b. Cytokeratin 7
   c. P63
   d. CD117
   e. MYB-- MYB-NFIB translocation in 90% salivary and cutaneous ACC’s

REFERENCES:


Case 2: Follicular lymphomatoid papulosis (“Type F”)

I. Clinical features
   a. Crops of papules <2 cm, resolving +/- scarring over 3-12 weeks
   b. No predilection for anatomic location
   c. May occur in setting of malignancy in 15-20% of cases
      i. Most common malignancy—MF
      ii. Other tumors—Hodgkin lymphoma, ALCL, rarely CLL, Sézary

II. Histopathologic subtypes
   a. LyP A- Wedge shaped infiltrate, variable numbers of atypical enlarged
      lymphocytes admixed with smaller cells, eosinophils, +/- vacuolar changes,
      erythrocyte extravasation
   b. LyP B- MF-like. Epidermotropic infiltrates of small lymphocytes, occasionally
      forming Pautrier microabscesses.
      i. Can lose CD30 expression
   c. LyP C- ALCL-like. Sheets of enlarged, atypical lymphocytes, resembling ALCL.
      CD30+. Usually without other ALCL markers.
   d. LyP D- CD8+. Epidermotropic intense infiltrates of CD8+ cells, with superficial
      and deep infiltrates. On morphology, can resemble an aggressive
      epidermotropic CD8+ lymphoma. Clinical correlation is crucial
   e. LyP E- angiotropic subtype. + Angiotropism, with associated necrosis. Resolves
      with scarring
   f. LyP F- follicular subtype- see below
   g. LyP with TCR gamma expression
   h. DUSP22 rearrangement

III. Histopathologic features of follicular lymphomatoid papulosis
   a. Perifollicular atypical lymphocytes with CD30 positivity
   b. Folliculotropism and infiltration into sebaceous lobules
   c. Other manifestations:
      i. Follicular mucinosis
      ii. Granulomatous infiltrate and follicular rupture
      iii. Peri-eccrine infiltrates

IV. Differential diagnosis
   a. Papular MF- clinical information of regressing lesions is essential. Complicated
      by concurrent MF in LyP patients. Lesions of MF will tend to persist without
      intervention. Histologically, lacks folliculotropism
   b. Folliculotropic MF- Clinical history again is paramount. CD30 positivity can be
      seen in FMF, but this is more likely with large cell transformation. Clinically, will
   c. Pityriasis lichenoides (for LyP A)—erythrocyte extravasation, vacuolar interface
      changes, no atypical CD30+ cells. Can sometimes be difficult to differentiate,
      and there may be some overlap.
   d. Cutaneous anaplastic large cell lymphoma
      i. Clinical- larger, persistent lesions, +/- ulceration. Can be oligolesional,
         can result in local LN involvement
      ii. Histopathologic and molecular- Sheets of atypical, enlarged lymphocytes
         (like LyP C), may resemble carcinoma.
1. Can be + for CD56, EMA, negative for CD3, TCR beta
2. DUSP22 rearrangement (only 3% of LyP, ~20% of ALCL)
e. Benign mimics- arthropod bite reaction, especially nodular scabies

REFERENCES
