Follicular neoplasms in 15 minutes!

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Adnexal neoplasms are fertile soil for lumper-splitter wars

- No uniform accepted terminology
- No “consensus conference police”
- Neoplasms frequently redescribed with new names, especially in non-peer-reviewed books!
- Some variants are like snowflakes (one of a kind)
- “Differentiation toward” follicles, sweat ducts, sebaceous glands, not “origin from”
Trichofolliculoma

- Solitary comedo-like papule
- Tufts of vellus hairs sometimes seen clinically
- Multiple mature vellus follicles open into big plugged mother of all follicles
- Dilated pore of Winer: Buds off comedo without mature follicles or hair shafts
- Pilar sheath acanthoma: Upper lip, more massive epithelial proliferation
Trichofolliculoma
Plug in upper lip
Pilar sheath acanthoma
massive
Folliculosebaceous cystic hamartoma
Trichoepithelioma

- **Fleshy** (not pearly) papules, usually central face
- Often **solitary**, sometimes hereditary multiple
- Basaloid aggregates, often reticulated
- Often **horn cysts**
- **Characteristic stroma** with many fibroblasts; no stromal retraction from tumor
- **Papillary mesenchymal bodies**
- **Trichoblastoma**: in vogue term now, splitters say trichoep is superficial form of trichoblastoma with cribriform or germinative pattern
Trichoepithelioma
Trichoepithelioma
Trichoepithelioma
Trichoepithelioma
Desmoplastic trichoepithelioma

- Annular plaque on face, resembles BCC or scar
- Like a sclerosing BCC (basaloid strands), but has horn cysts, trichoep-type stroma, less atypia, and no stromal retraction
Desmoplastic trichoepithelioma

• **Microcystic adnexal carcinoma** also similar, but is large and has sweat ducts as well as horn cysts.

• Like solitary trichoep, in the setting of severe sun damage, “when in doubt, cut it out” and treat like a BCC, because sometimes impossible to distinguish “BCC with follicular differentiation”
Desmoplastic trichoepithelioma
Trichoadenoma of Nikolowski

• Annular solitary papule or plaque on face with central depression resembling BCC or scar

• A variant (?) of trichoblastoma or trichoepithelioma with many horn cysts surrounded by mature (less basaloid) “squamatized” epithelium
Trichoadenoma
Pilomatricoma (pilomatrixxoma)  
Calcifying epithelioma of Malherbe

• Solitary “cystic” nodule, chalky keratin inside
• Elevates the skin: “tent sign”
• Often face, neck, upper arm of children, also second peak in older adults
• Basaloid cells, squamous epithelial lining, shadow (ghost) cells, transitional pyknotic cells
• Often calcify or ossify
• Often granulomatous reaction
Pilomatricoma

- Called a “cyst” by the less astute clinician
- Shards of keratinous material like a cyst
- Often chalky due to calcification.
- Sometimes mutation in the beta-catenin gene (CTNNB1).
- May be associated with Gardner syndrome
- Multiple pilomatrixomas are seen with myotonic dystrophy.
Pilomatricoma
Pilomatricoma
Pilomatricoma
Pilar tumor =
Proliferating trichilemmal cyst =
Proliferating follicular cystic neoplasm

- Proliferating wall of pilar cyst (trichilemmal cyst, isthmus-catagen cyst) with abrupt “trichilemmal keratinization”
- Some cases are malignant and can metastasize (“malignant pilar tumor”); traditional dogma is that most are a pseudomalignancy. Ackerman feels they all are SCCs!
Scalp nodule
Proliferating trichilemmal cyst

Epidermis

Squamous proliferation within "cyst"
Proliferating trichilemmal cyst
Proliferating trichilemmal cyst
Trichilemmoma = Trichoolemmoma

- **Solitary** verrucous papule on face
- **Multiple** lesions in cases of **Cowden’s disease**
- **Lobule** of epithelium with pale cells projects downward into the dermis
- **Not a viral wart** by molecular methods
- **Clear cell acanthoma** is usually on the leg and is more psoriasiform
Trichilemmoma
Trichilemmoma
Tumor of follicular infundibulum (TFI)

- Flat or barely papular hyperkeratotic papule of face
- Horizontally oriented pale epidermal shelves that connect adjacent follicles and surface epithelium
- Variant of reticulated seborrheic keratosis?
- Ackerman has proposed that seborrheic keratosis might be a follicular neoplasm!
Tumor of follicular infundibulum

- Slightly pale keratinocytes
- Horizontal bridge between follicles
Fibrofolliculomas and trichodiscomas

• Multiple smooth facial papules: Hornstein-Birt-Hogg-Dubé syndrome: folliculin FLCN gene, skin tags, renal tumors, spontaneous pneumohorax.

• Closed related to perifollicular fibroma or angiofibroma but fibrofolliculoma has delicate strands of epithelium that project into the perifollicular loose stroma.

• Trichodiscoma is probably the same tumor without the follicular strands; probably not related to the mythical Haarscheibe (hair disc).
Birt-Hogg-Dubé syndrome
Trichodiscoma
Trichodiscoma
Trichoblastoma – in vogue!

• When in doubt, call it a BCC – **immunostains similar to trichoep, overrated:**
• **Bcl-2** more positive in BCC
• **CK20** Merkel cells more often populate trichos
• **CD34** stains stroma right up against follicular tumor – spares margin around BCC - worthless
• Splitters say trichoep is a superficial form of trichoblastoma with cribriform or germinative pattern
• Is trichoblastoma the most common tumor arising in nevus sebaceus?
Trichoblastoma
Trichoblastoma
Malignant Follicular Tumors:
Pilomatrix carcinoma
Trichilemmocarcinoma

• Lumpers would say these are all just squamous cell carcinomas with follicular differentiation (just SCCs that have shadow cells or abrupt trichilemmal keratinization)
Trichilemmocarcinoma