CASE HISTORY

A 44-year-old woman presented with a one centimeter peri-anal “skin tag” that she first noticed approximately two years ago. The lesion had been stable in size during that time, did not drain, and was non-tender. Her past medical history was significant for constipation and mild hemorrhoids. She requested surgical excision of the peri-anal lesion. Photomicrographs of representative histologic sections of the lesion are shown below.
What is your diagnosis?

a. Apocrine adenocarcinoma  
b. Colorectal adenocarcinoma  
c. Hidradenoma papilliferum  
d. Syringocystadenoma papilliferum
e.  Tubular apocrine adenoma

(SCROLL DOWN FOR ANSWER AND DISCUSSION)
Answer and Discussion:

C. Hidradenoma papilliferum

Hidradenoma papilliferum is an uncommon benign tumor that occurs primarily in women. It usually arises as a solitary, painless mass in the anogenital region, but may develop elsewhere, including the head and neck. Microscopically, the hidradenoma papilliferum is a well-circumscribed nodule situated in the dermis with an arborizing or “mazelike” papillary architecture. The papillary structures within the lesion are lined by two layers of cells, including an inner myo-epithelial cell layer and a luminal tall columnar/cuboidal epithelial cell layer. Apocrine differentiation with “snouting” (decapitation secretion) is seen in the epithelial cells, which tend to have oval, pale-staining nuclei situated near the cell base. While mild cytologic atypia may be present, mitotic activity and tumor necrosis are not seen. Like the hidradenoma, the hidradenoma papilliferum usually shares no connection with the epidermis.

Epithelial cells in hidradenoma papilliferum express keratin, EMA, and CEA. As an indicator of apocrine differentiation, GCDFP-15 is typically also positive. Estrogen receptors and progesterone receptors are often expressed in the epithelial cells, suggesting this entity may have a mammary gland-like origin. Interestingly, morphologic alterations that are commonly seen in benign breast diseases can be present in hidradenoma papilliferum. These changes include oxyphilic metaplasia (similar to apocrine metaplasia in breast), epitheliomatosis with a streaming growth pattern, sclerosing adenosis-like changes, atypical apocrine adenosis-like changes, changes corresponding to usual ductal epithelial hyperplasia, lamprocyte-like changes, clear cell change of the myoepithelium, foamy histiocyte reaction, and stromal fibrosis.

Rarely, hidradenoma papilliferum may transform into its malignant counterpart (apocrine adenocarcinoma), which demonstrates marked cytological atypia, mitotic activity, and necrosis.

References


Case contributed by:

Nima Amini, M.D. (Pathology Resident, UC Davis Medical Center)

Kristin Olson, M.D. (Assistant Professor, UC Davis School of Medicine)