An unusual cystic lesion on the helix

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DERMPATH QUIZ

A 73-year-old male presented to his dermatologist with a cyst over the left helix, which had been present for an unknown duration. The clinical impression was an epidermoid cyst, and the lesion was excised and sent for histopathological evaluation.

Dermatopathological sections revealed the cystic, basaloid neoplasm pictured in Figures 1 and 2.

The cystic neoplasm most likely represents:

A. Cutaneous cyst of Gardner's syndrome
B. Cystic basal cell carcinoma
C. Cystic panfolliculoma
D. Pilomatricoma
E. Trichoblastoma

Answer: C. Cystic panfolliculoma

DISCUSSION

Panfolliculomas represent a family of benign pilar neoplasms initially described by Ackerman *et al*. in 1993 who coined the term to describe the neoplasm's differentiation toward all segments of the follicular unit–the follicular infundibulum, isthmus, stem, and bulb.[1] The most common pattern is that of a cystic neoplasm, although intraepidermal and sebaceous variants also occur.[2,3] The characteristic histopathologic findings of panfolliculoma include evidence of differentiation toward both the upper and lower segments of the hair follicle.[4] Figure 1 demonstrates a partially ruptured cystic neoplasm with areas of differentiation toward the matrix and infundibulum. Figure 2 demonstrates pilomatrical keratinization with an adjacent infundibular cyst. Trichohyalin granules indicating inner root sheath differentiation are noted in Figure 3.

Panfolliculomas occur in males and females with equal frequency and with a reported age range from 30 to 81 years.[3,5] The tumor is usually located on the head or trunk with a few reports of leg involvement.[5] The most common clinical impression is that of an epidermoid cyst, but clinical presentations mimicking a trichoepithelioma and nodular basal cell carcinoma have been reported.

Histopathologically, panfolliculomas are unique among follicular cysts in their differentiation toward all parts of the normal follicular unit. The components most commonly found include corneocytes in a basket weave and laminated pattern, epidermoid keratinocytes, trichilemmal differentiation of glycogenated cells with peripheral palisading, inner root sheath keratinization with trichohyalin granules, and matrical differentiation with keratinization to shadow cells. Proliferation of follicular bulb, germ, and papilla can also occur.[1]

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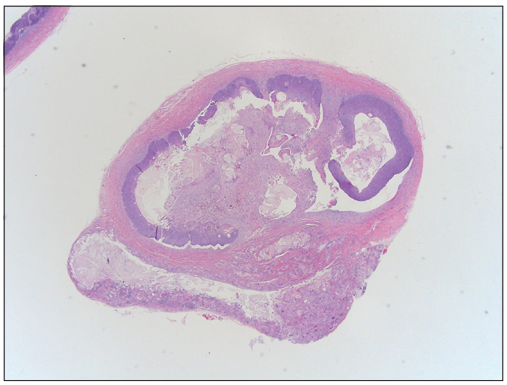

Figure 1: Panfolliculoma, H and E, ×20

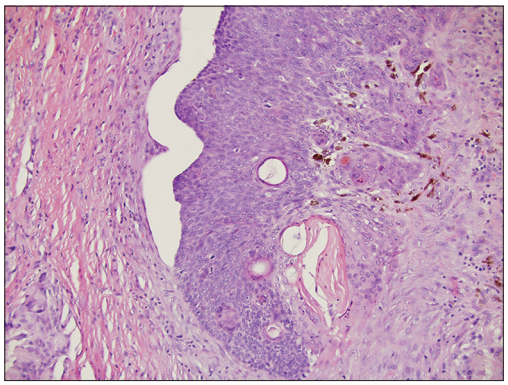

Figure 2: Panfolliculoma, H and E, ×40

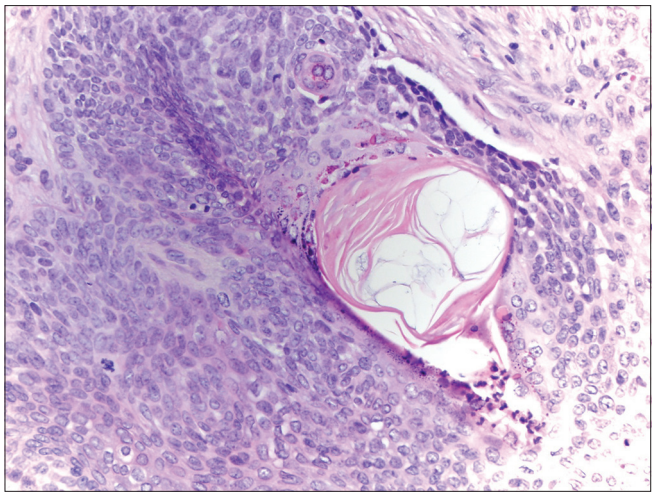

Figure 3: Panfolliculoma, H and E, ×600

Cystic panfolliculoma does not have any known syndromic associations, and it is important to differentiate this neoplasm from mimics, which include malignancy and cysts associated with genodermatoses. Specifically, the differential diagnosis includes cystic basal cell carcinoma, pilomatricoma, the cutaneous cysts of Gardner's syndrome, and trichoblastoma. Cystic panfolliculoma lacks the fibomyxoid stroma of cystic basal cell carcinoma and demonstrates areas of matrical and inner root sheath differentiation. Furthermore, necrosis is not a common finding in cystic panfolliculomas. The presence of trichohyalin granules in panfolliculoma assists in differentiation from benign pilomatricoma and from the cutaneous cysts with pilomatrical differentiation associated with Gardner's syndrome.[6] Trichoblastoma demonstrates differentiation toward the hair bulb, but not toward the isthmus or infundibulum. Trichoblastomas have a prominent fibroblast-rich stromal component with papillary mesenchymal bodies. In contrast, panfolliculomas demonstrate little stromal component and differentiation toward all segments of the follicular unit.[1]

In summary, panfolliculoma is an uncommon follicular neoplasm with a distinct histopathologic appearance. Correct identification and differentiation from its mimics is important to allow for appropriate therapy.

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