# Ulcerated Nodule on the Scalp

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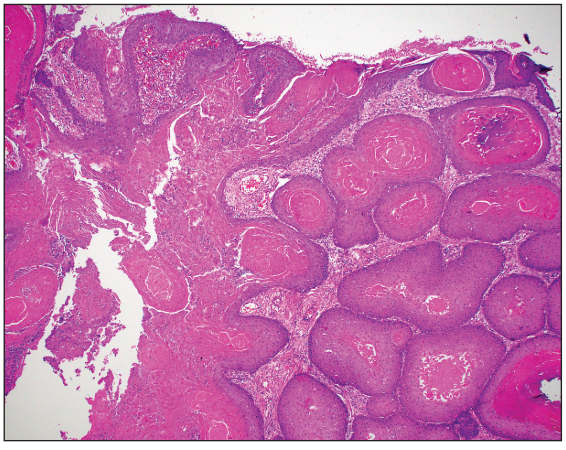

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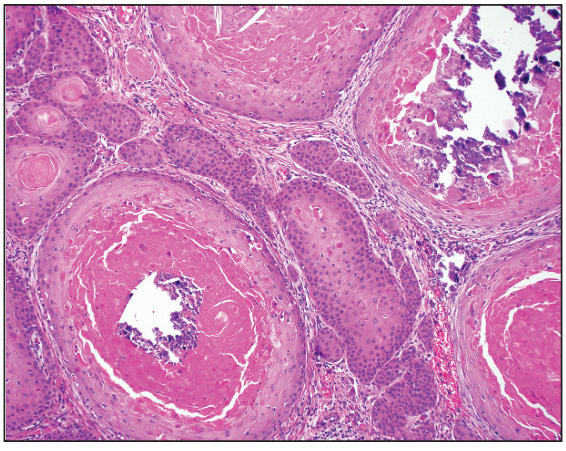

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A 66-year-old woman presented to the dermatology clinic with a rapidly enlarging, draining lesion on the scalp. The lesion seemed to enlarge over the last 3 months from a lesion that had been there for years. Physical examination revealed a 2.2-cm ulcerated nodule on the right parietal scalp. A shave biopsy was obtained.

## THE BEST **DIAGNOSIS IS:**

a. desmoplastic trichilemmoma
b. inverted follicular keratosis
c. keratoacanthoma
d. pilomatricoma
e. proliferating pilar tumor

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The authors report no conflict of interest.

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# THE **DIAGNOSIS:** Proliferating Pilar Tumor

Proliferating pilar tumor (PPT), or cyst, is a neoplasm of trichilemmal keratinization first described by Wilson-Jones[1] in 1966. Proliferating pilar tumors lie on a spectrum with malignant PPT, which is a rare adnexal neoplasm first described by Saida et al[2] in 1983. The incidence of PPT is unknown given the paucity of cases and the possible misdiagnosis as squamous cell carcinoma (SCC). Proliferating pilar tumors tend to present on the head and neck of older females as a multilobular and sometimes ulcerating nodule.[3] Although PPT can occur de novo, the majority of cases are thought to develop progressively from a benign pilar cyst. Histopathologically, PPT is characterized by cords and nests of squamous cells that display trichilemmal keratinization (quiz images).

Classification of PPT as benign or malignant is challenging, though criteria have been proposed.[3-7] Lesions with minimal infiltration into the surrounding dermis and scant mitosis typically behave in a benign manner, while lesions showing nuclear atypia, atypical mitosis, and irregular infiltration into the surrounding dermis can have up to a 50% locoregional recurrence rate.[3] In addition, distinguishing a PPT from an SCC or trichilemmal carcinoma also can be difficult; however, SCC is favored when there is a lack of trichilemmal keratinization or when squamous atypia is present in the adjacent epidermis.[8] Trichilemmal carcinoma is a rare tumor that has been questioned as a distinct entity.[9-12]

Pilomatricoma, also known as calcifying epithelioma of Malherbe, is a benign pilar tumor that presents as a slowly growing nodule on the head or neck area or arms.[13,14] Most pilomatricomas develop by the second decade of life. Multiple lesions may be present in association with myotonic dystrophy or Gardner syndrome among other syndromes.[15-17] Similar to PPT, pilomatricomas present as large dermal nodules; however, they tend to be circumscribed and have a trabecular network that consists of basophilic cells and eosinophilic keratinized shadow cells (Figure 1).[18] Calcification may be seen and bone formation subsequently may occur.[19]

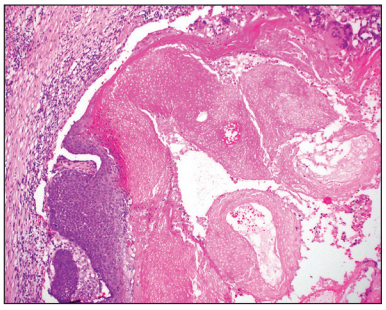

**FIGURE 1.** Pilomatricoma. Dermal tumor with peripheral basaloid cells, ghost cells, focal keratin debris, and focal multinucleated foreign body giant cells (H&E, original magnification ×100).

Most sources now consider keratoacanthoma (KA) as a well-differentiated SCC.[20] The typical presentation consists of a rapidly growing erythematous to flesh-colored nodule with a central keratinous plug that develops over a period of weeks. If untreated, KAs may resolve over a period of months and leave a depressed scar. Local destruction can result from KAs, and they have the potential to transform into a more aggressive SCC. Accordingly, most clinicians use tissue destructive methods, excision, or Mohs micrographic surgery for treatment based on location. Histologically, a well-circumscribed proliferation of glassy cytoplasm is noted. A depressed keratin-filled center is surrounded by a lip of epithelium extending over the lesion (Figure 2).[20,21] Pseudoepitheliomatous hyperplasia accompanied by hypergranulosis is seen in the center of KAs rather than at the periphery, which is typical of non-KA SCCs. Typical KAs lack acantholysis, a feature suggesting a non-KA type of SCC. Neutrophilic microabscesses and eosinophils commonly are seen in KAs.[20,21]

Inverted follicular keratosis is a benign tumor that gained traction as its own entity in the 1960s.[22] These

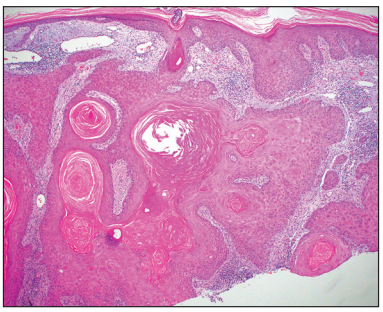

**FIGURE 2.** Keratoacanthoma. Crateriform squamous proliferation with keratin debris in the center of the cystic spaces and squamous cells toward the periphery with eosinophilic and glassy cytoplasm (H&E, original magnification ×40).

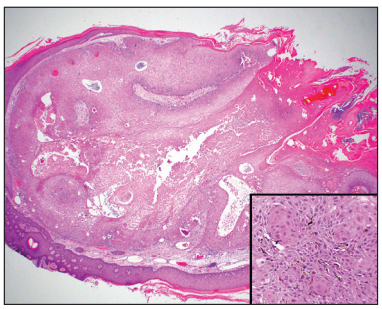

**FIGURE 3.** Inverted follicular keratosis. Endophytic, slightly verrucous squamous proliferation with central cyst formation and keratin debris (H&E, original magnification ×20). Squamous eddies and dendritic melanocytes can be seen on higher magnification (H&E, original magnification ×100 [inset]).

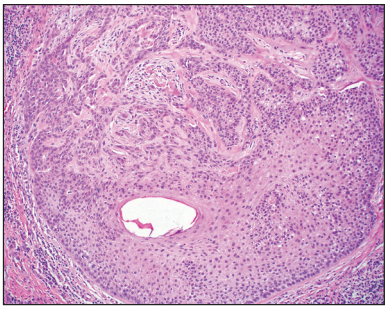

**FIGURE 4.** Desmoplastic trichilemmoma. Proliferation of lobules of squamous cells with clear cytoplasm, peripheral palisading of more basaloid keratinocytes, and a thickened basement membrane (H&E, original magnification ×100).

lesions typically develop from the follicular infundibulum, but some consider them a version of a wart or seborrheic keratosis.[23] They generally are flesh-colored nodules on the upper cutaneous lip or face. Treatment usually consists of complete excision. There are many different growth patterns described, but they typically are endophytic tumors with eosinophilic squamous cells in the center and more basophilic cells at the periphery (Figure 3).[24] Characteristically, there are squamous eddies throughout the tumor (Figure 3 [inset]). There also may be a scant lymphohistiocytic infiltrate within the dermis surrounding the lesion.

Trichilemmomas are flesh-colored adnexal neoplasms that may present as a solitary lesion or in clusters on the face. They have been reported to occur on all nonglabrous skin sites.[25] Multiple lesions may occur in association with Cowden syndrome or with nevus sebaceous.[26] A desmoplastic variant of trichilemmomas has been reported.[27] Desmoplastic trichilemmomas appear as well-circumscribed tumors of outer root sheath differentiation with lobules extending down into the dermis.[28] Vacuolated glycogen-filled keratinocytes are scattered throughout the lesion but are most prominent at the base. At the periphery of the lobules, peripheral palisading of basaloid cells is accompanied by a thickened eosinophilic basement membrane that is periodic acid–Schiff positive. Typical trichilemmomas also can display these features; however, the main differentiating feature of a desmoplastic trichilemmoma is the pink hyalinized stroma separating small islets of basophilic cells (Figure 4). Differentiation from an invasive malignant carcinoma sometimes can be challenging without a focus of typical trichilemmoma or if the biopsy specimen is too superficial.[29]

Pilar cysts are common tumors that typically arise on the scalp and sometimes are proliferating. Proliferating pilar tumor should be kept on the differential when secondary changes such as ulceration occur in the primary lesion of the scalp. Microscopically, and sometimes clinically, PPT can be difficult to differentiate from other mimickers.

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