Panfolliculoma: report of two cases

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ABSTRACT
Panfolliculoma is a distinctive and unusual benign follicular neoplasm. It shows differentiation towards all components of the hair follicle, including the upper and the lower segments. We report two cases of this rare disease. The first patient presented with a painful nodule on the scalp and the second patient had an asymptomatic skin-colored nodule on his left eyebrow. Histopathologic examination of both cases revealed lobular proliferation of solid-cystic follicular structures. The cystic structures contain laminated keratin, indicating infundibular differentiation. The solid tumor part was composed of basaloid germinative cells, matricial cells, and cells with trichohyalin granules, representing inner root sheath differentiation. The range of differentiations was highlighted using immunohistochemical stains, including labeling of cytokeratin 1, 5, 10, 14 (using 34βE12), cytokeratin 5/6, CD34 (outer root sheath), and Ber-EP4 (lower segment of hair follicle). Panfolliculoma should not be confused with trichofolliculoma or trichoblastoma.

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Introduction

The term panfolliculoma was first described by Ackerman and colleagues in 1993. This term refers to a follicular neoplasm characterized by differentiation into all components of a follicle. Clinically, the neoplasm usually presents to dermatologists as a slow-growing cyst or a translucent dome-shaped papule resembling a cyst, trichoepithelioma or basal cell carcinoma. We report two cases of panfolliculoma and discuss the histopathologic differential diagnosis with regard to other similar follicular tumors.

Case reports

Case 1

A 41-year-old female patient presented with a tender and inflamed nodule on the scalp (Figure 1A). She had been aware of the lesion for several months. It was initially asymptomatic, but gradually became mildly painful. Excision biopsy was performed, under the impression of an epidermal cyst or pilary cyst.

Microscopic examination revealed a well-demarcated and multicystic lesion in the deep dermis (Figure 1B). The wall of the cystic structure was composed of both corneocytes arranged in a basket-weave and laminated array, resembling infundibular differentiation (Figure 1C), and blue-gray corneocytes containing pinkish trichohyalin granules (Figure 1D), indicating hair matricial differentiation. Cells with clear cytoplasm indicative of outer root sheath were also found at the periphery of the trichohyalin-rich corneocytes. Distorted or incompletely formed hair shafts were scattered in the cystic structure. Total excision was performed and no recurrence was noted at subsequent follow-up.

Case 2

A 51-year-old man had noted an asymptomatic, flesh-colored subcutaneous nodule on his left eyebrow for several years. A second pinkish protruding papule had developed above the original nodule over the preceding year (Figure 2A). He became concerned and visited the dermatological clinic at...
Figure 1  (A) Tender and inflamed nodule on the scalp. (B) The tumor was composed of connecting basaloid tumor islands with multiple cystic structures of variable sizes (H&E, 20x). (C) There was laminated keratin content in the cystic structure with hair shaft formation, showing complete hair follicle differentiation (H&E, 100x). (D) Lower segment differentiation of hair follicle characterized by trichohyalin granules and matrical cells (H&E, 200x).

Figure 2  (A) Pedunculated pinkish nodule on the left eyebrow. (B) The tumor was composed of solid cystic nodules (H&E, 20x). (C) Each tumor nodule demonstrated full hair follicle differentiation, from infundibulum to the hair matrix (H&E, 100x). (D) Close view showing laminated keratin in the cystic structure and matrical differentiation (H&E, 200x).
Panfolliculoma is rarely reported, with only 14 cases reported in the literature to date.\(^1\)–\(^5\) However, the small number of reported cases suggests that panfolliculoma affects both sexes equally, with an age range between the second and seventh decades. This neoplasm can be located on the eyebrow, buttock, trunk or scalp. It usually presents as a slow-growing cystic nodule, and may mimic basal cell carcinoma. The distinct histopathologic features of this tumor often confuse pathologists, and it may be misdiagnosed as trichofolliculoma, trichoepithelioma, or trichoblastoma. By definition, the constituents of this neoplasm include germinative cells, matrical cells, cells of the outer sheath, trichohyalin granules indicative of inner root sheath differentiation, shadow cells suggestive of hair differentiation, and corneocytes in a basket-weave and laminated array and showing infundibular differentiation. This differentiation towards all elements of a hair follicle is unique, and differs from other benign follicular tumors.

Trichofolliculoma is a relatively common benign follicular tumor. It is characterized by a cystic structure with secondary follicular differentiation of the cyst wall, and the formation of small vellus hairs inserted into the cystic...
cavity. However, trichofolliculoma shows mature hair follicle differentiation, and therefore no proliferation of immature basaloid tumor cells.

Trichoblastoma and trichoepithelioma (superficial variants of trichoblastoma) are characterized by lobular proliferation of basaloid cells arranged in reticular, racemiform, or cribriform patterns. Panfolliculoma resembles these two tumors at lower microscopic magnifications. However, trichoblastoma and trichoepithelioma have greater tendencies toward follicular germs and hair papillae formation, and do not have complete hair follicle differentiation.

There is usually no differentiation toward infundibulum, isthmus, outer root sheath, or hair shaft formation in trichoblastoma or trichoepithelioma.

Immunohistochemical staining can also help to identify the differentiation towards different parts of the hair follicle. Hoang and Levenson reported the immunohistochemical profile of panfolliculoma, that is the uniform labeling of tumor cells by 34βE12 (high molecular cytokeratin) and CK 5/6 immunostaining. The 34βE12 antibody stains keratinocytes in the follicular infundibulum and isthmus, and the CK 5/6 stains outer root sheath. Therefore, we suggest that this immunohistochemical profile supports the existence of both upper and lower segments in panfolliculoma. Moreover, Ber-EP4 highlights germinative cells, while immunostaining for CD34, an indicator of trichilemmal keratinization and perifollicular fibrous tissue, are able to identify fibrotic stroma and focal epithelial components in the panfolliculoma.

Although the immunohistochemical staining is helpful, it is not essential for diagnosis, because the characteristic findings are usually easily recognizable following hematoxylin and eosin staining.

The nomenclature of benign follicular tumors is based on their architecture, tumor cell composition and their association with hair follicle differentiation. It has been suggested that the development of different follicular components in these tumors can be affected by the perifollicular stroma. Various types of follicular or sebaceous differentiation were observed in the epidermis of dermatofibromas or the cystic wall of epidermal cysts. It is possible that the induction of different follicular structures occurs more commonly in hair-rich areas, such as the face and scalp, leading to the formation of panfolliculoma and other benign follicular tumors.

In summary, panfolliculoma is a rare neoplasm exhibiting differentiation towards all components of the hair follicle. Clinical differentiation between panfolliculoma and other follicular tumors can be difficult, but it exhibits characteristic histologic features that should prevent its confusion with other benign follicular tumors.

References

5. González-Guerra E, Requena L, Kutzner H. Immunohistochemical study of calretinin in normal hair follicles and tumors with follicular differentiation. Actas Dermosifiliogr 2008;99:456–63. [In Spanish]