Dermpath Quiz

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A 52-year-old woman with no significant past medical history presented to her dermatologist with a long-standing history of multiple, small papules on her face. These had remained stable in size. The remainder of the physical examination and laboratory investigations was unremarkable. A biopsy of one of the papules was taken from the right side of her hairline.

THE LESION MOST LIKELY REPRESENTS?

A. Trichoepithelioma
B. Trichoblastoma
C. Basaloid follicular hamartoma (BFH)
D. Fibroepithelioma of Pinkus (FOP)
E. Infundibulocystic basal cell carcinoma (IFBCC)

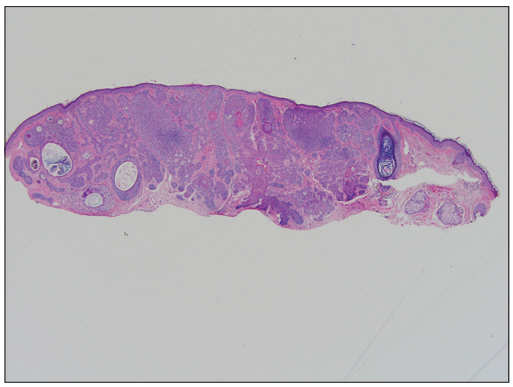

Figure 1: The lesion is composed of basaloid cells with horn cysts and peripheral palisading (H and E, ×40)

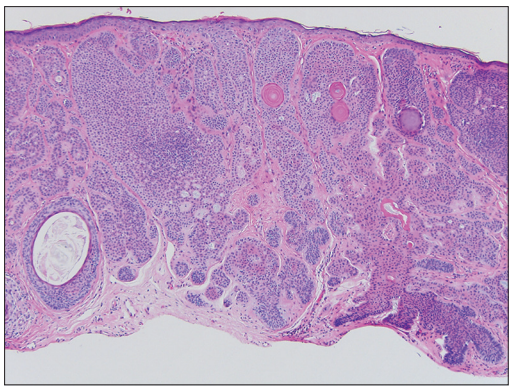

Figure 2: Anastomosing strands of basaloid cells are noted. The intervening stroma is scant with myxoid areas and clefting within the stroma (H and E, ×100)

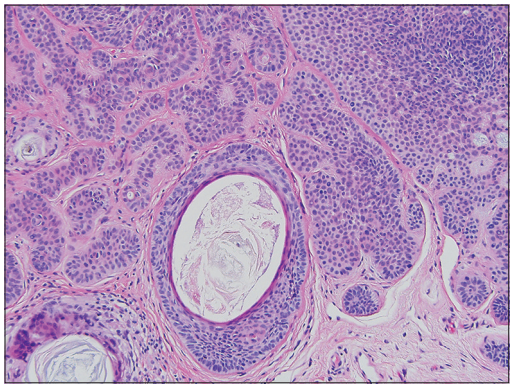

Figure 3: Prominent horn cysts with keratin are identified. Peripheral palisading is prominent (H and E, ×200)

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ANSWER

C. Basaloid follicular hamartoma

DISCUSSION

Basaloid follicular hamartoma is a rare, benign follicular tumor that recapitulates portions of the normal hair follicle. It demonstrates a spectrum of clinical presentations, including multiple lesions with a generalized or localized distribution, blaschkoid distribution, or as a solitary stable lesion. The generalized form may be sporadic, acquired, familial or congenital. The sporadic form consists of multiple lesions without systemic disease. The acquired form occurs in females and is associated with alopecia, myasthenia gravis and systemic lupus erythematosus. The familial form is an autosomal dominant disease and can be found in association with hypotrichosis and hypohidrosis. Finally, the generalized congenital form is associated with cystic fibrosis and alopecia.[1,2] Patients have been described with overlap between BFH syndrome and Gorlin's syndrome and both are associated with PTCH mutations.

Histopathological examination reveals nests and cords of basaloid cells within the superficial dermis, embedded in a scant to abundant fibromucinous stroma [Figures 1 and 2]. Horn cysts and hair follicle-like structures are present [Figure 3]. And clefting may be noted within the stroma, but unlike other benign pilar tumors, BFH rarely demonstrates a concentric fibroblast-rich stroma or papillary mesenchymal bodies.

The differential diagnosis includes trichoepithelioma, trichoblastoma, FOP and IFBCC.[2,3]

Trichoepitheliomas are usually larger than BFH and are composed of tumor islands of basaloid cells in a distinct lace-like or "swiss-cheese" pattern. In addition, the stroma of trichoepithelioma is more cellular with concentric fibroblasts, papillary mesenchymal bodies and more prominent stroma clefting.[1]

Trichoblastomas are circumscribed, large, basaloid neoplasms which, unlike BFH, are located in the deep dermis or subcutis, without any connection to the surface epithelium. Trichoblastomas, like related trichoepitheliomas, have a cellular stroma with prominent papillary mesenchymal bodies.

Fibroepithelioma of Pinkus is a variant of basal cell carcinoma, which is characterized by arborizing cords of basaloid cells from the epidermis. Islands of fibromucinous stroma are surrounded by the reticular epithelial network, and eccrine ducts are present within the pink strands of epithelium. Blue basaloid buds are noted at the tips of the pink strands.[1,4]

The most important differential diagnosis of BFH is IFBCC. IFBCC is characterized by cords and strands of basaloid cells embedded in a scant, fibromyxoid stroma, as in BFH. Horn cysts, clefts within stroma and no epidermal changes are also common to both lesions.[2,3] While IFBCC can have a greater degree of cellular atypia, mitotic activity, necrosis, deep infiltration, and epidermal ulceration, the two tumors may be identical histologically and often can only be distinguishable based on clinical features and biological behavior. At the molecular level, both lesions have abnormal PTCH (patched) signaling pathways.[1,3,5]

Surgical excision is usually not required for BFH, except for cosmetic reasons or to exclude other skin tumors. When multiple BFH lesions are present, clinicians should be aware of the possibility of systemic disease.[1-3]

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