A firm plaque on the cheek

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DERMPATH QUIZ

A 22-year-old African-American male patient presented to his dermatologist with an asymptomatic, solitary, firm, plaque on his left cheek of unknown duration [Figure 1]. The lesion had a central depression and raised borders. The patient denied any use of topical medications on the lesion, and the past history was unremarkable. The patient was immunocompetent and denied trauma to the site. No other skin lesions were present elsewhere. An excision biopsy was performed. The dermatopathological sections revealed collections of basaloid cells arranged as small nests and narrow cords, horn cysts, focal areas of calcification, and a desmoplastic stroma. Clefts were present within the stroma, but not between tumor nests and stroma [Figures 2-4]. Of concern were the noted tumor cells seen peripheral to an arrector pili-muscle [Figure 5].

The neoplasm most likely represents:

A. Morpheaform basal cell carcinoma
B. Microcystic adnexal carcinoma
C. Desmoplastic trichoepithelioma
D. Syringoma
E. Folliculosebaceous cystic hamartoma.

ANSWER

C. Desmoplastic trichoepithelioma

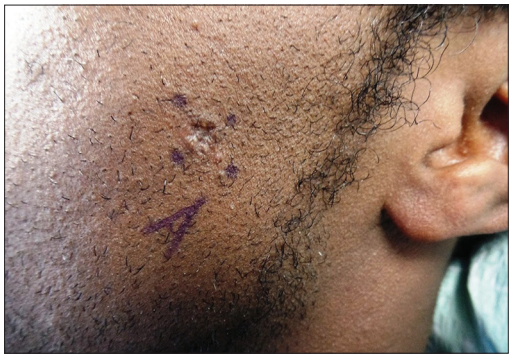

Figure 1: Solitary, firm, white to erythematous, annular plaque on the left cheek

DISCUSSION

Desmoplastic trichoepithelioma (DTE), also known as sclerosing epithelial hamartoma, is an uncommon, benign adnexal tumor with a predominantly infundibular differentiation.[1-2] Follicular, sebaceous, and apocrine differentiation may be present.[2] It occurs most commonly on the face of young to middle-aged women as a firm, annular, skin-colored plaque or nodule with a characteristic nonulcerated central dell.[3] This patient was male, and the site, morphology, and histological features are typical for DTE.

The histopathological characteristics of DTE include central dell formation, proliferation of basaloid cells arranged in small nests and narrow strands in the superficial dermis embedded in a desmoplastic stroma, and presence of small keratinizing cysts. Clefts are present within the stroma, but there are no retraction spaces between tumor islands and the adjacent stroma. Other features that have been reported include granulomatous reactions from cyst rupture, calcification, absence of solar elastosis below the lesion, pseudoepitheliomatous hyperplasia, tumor attachment to the epidermis, and focal bone formation.[2-6] Perineural involvement may occur and could result in misdiagnosis of a malignant neoplasm.[7,8] Because DTE may differentiate toward various parts of the folliculosebaceous apocrine unit, immunohistochemistry staining for glandular structures may be focally positive. Other entities that present histologically with basaloid proliferation and desmoplastic stroma

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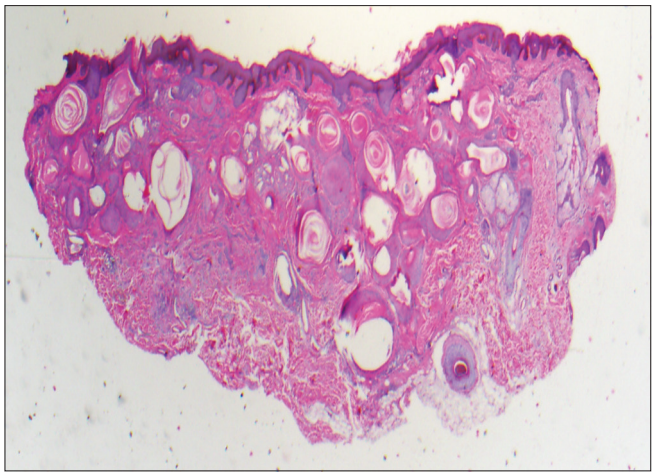

Figure 2: At scanning view, the biopsy shows prominence of horn cysts (H and E, X20)

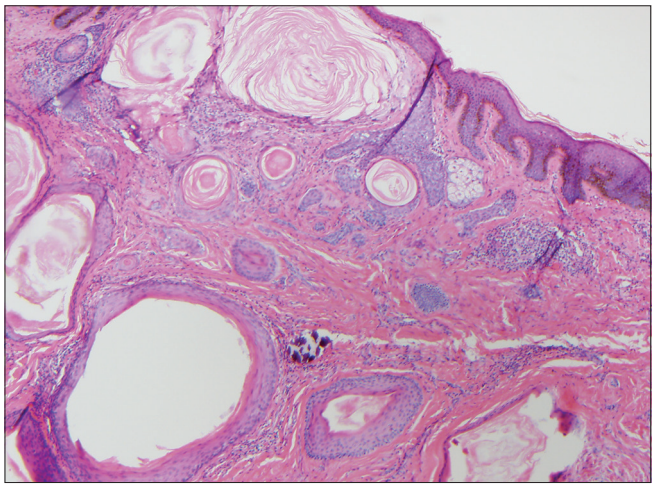

Figure 4: A high-power view shows basaloid cells arranged as small nests and as narrow cords, desmoplastic stroma, and focal calcification (H and E, X100)

include syringoma, microcystic adnexal carcinoma (MAC), and morpheaform basal cell carcinoma (MBCC). In rare cases, breast metastases may also mimic DTE histopathologically.[3] The differentiation of DTE from carcinomas and metastases is important to avoid surgical overtreatment especially in cosmetically sensitive areas.

Syringoma is a benign, sweat gland neoplasm presenting as small papules usually on the eyelids of Asian women and children with Down's syndrome.[9] Histologically, a small biopsy of syringoma and DTE may appear similar, but syringomas, however, are smaller, spherical, have no central dell, and calcification is rare.

MAC is a slow-growing, locally aggressive neoplasm that has a high recurrence rate.[10] It has a predilection for the head and neck area. It may be differentiated from DTE microscopically by the presence of nodular lymphoid aggregates, deeper invasion, and more prominent perineural invasion. Calcification and horn cysts are rare. Within a MAC, cytokeratin (CK) 7 shows intense

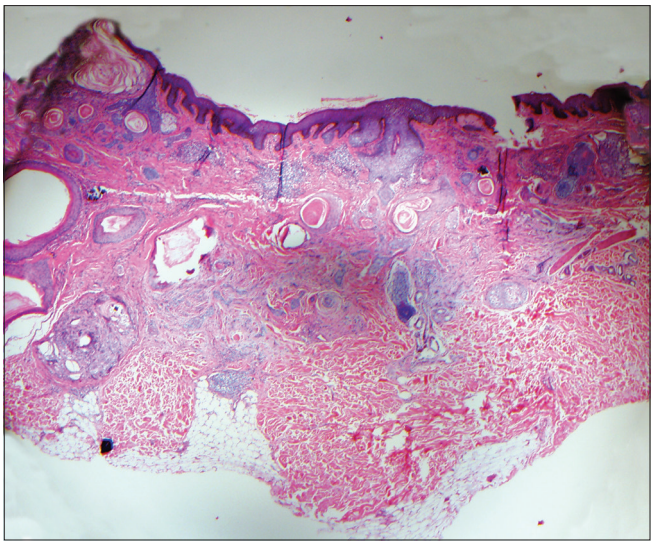

Figure 3: At low-power view, the biopsy again shows prominence of horn cysts (H and E, X40)

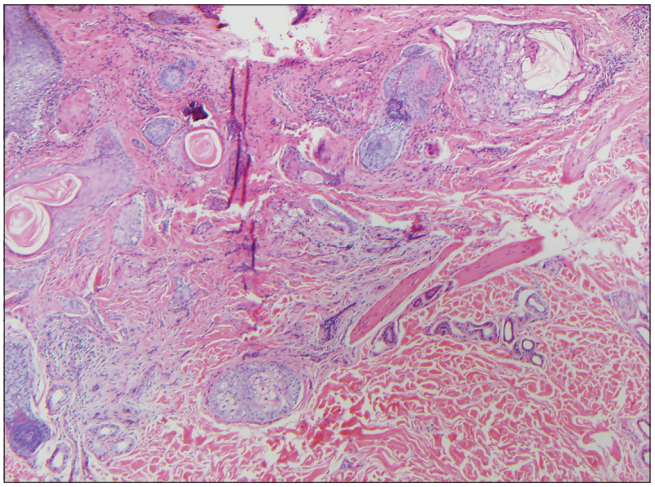

Figure 5: Another high-power view shows tumor cells at the periphery of an arrector pili muscle (H and E, X200)

staining of cells demonstrating glandular differentiation and focal staining of the ductal lumen. Like CK7, carcinoembryonic antigen (CEA) and S-100 staining suggests sweat gland differentiation and as such is useful in differentiating DTE from syringoma and MAC.[11,12]

MBCC clinically presents as a slowly growing scar-like lesion. Histologically, the basaloid islands demonstrate varying shapes and sizes, may be attached to the surface rather than to the infundibula, and may be present in larger aggregates.[9] Ulceration, increased mitotic activity, apoptosis, and clefting between the tumor cells and the stroma may also be seen.[2] Tumor extension into nerves and muscles have been used as criteria to distinguish infiltrative BCC from benign DTE. Ki-67 is a proliferation marker that diffusely stains MBCC.

Folliculosebaceous cystic hamartoma presents with a central infundibular cyst and radiating epithelium with pilosebaceous

differentiation. A surrounding concentric fibroblast-rich or myxoid stroma is often present. Occasionally, the stromal spindle cells may have a wavy neuroid appearance, but the cells stain positive for CD34 and negative for S100 protein confirming follicular fibrous sheath differentiation.

The histopathological findings of a well-circumscribed tumor containing cords and nests of small basaloid cells within a fibrous stroma containing horn cysts, foci of calcification, absence of clefting between tumor cells and stroma, absence of cellular atypia, absence of subcutaneous invasion, and perineural and perimuscular penetration confirm the diagnosis of DTE.

The typical cases of benign DTE does not require surgical treatment; however, if there are features to suggest MAC or MBCC, Mohs micrographic surgery is appropriate.[13]

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Conflicts of interest
There are no conflicts of interest.

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