A 9-year-old boy presented with a slowly progressive lesion of 5 years’ duration affecting only the left side of the face in a dermatomal pattern. The patient denied any symptoms and had no other anomalies or family history of similar lesions. On physical examination the lesion was found to span a 12×7-cm area of the lateral half of the left cheek and was composed of multiple variable-sized, pinkish to flesh-colored papules that coalesced in some areas to form small plaques. Few milialike cysts were present. One papule was biopsied.

**THE BEST DIAGNOSIS IS:**
- linear eccrine poroma
- linear nevoid basaloid follicular hamartoma
- linear spiradenoma
- unilateral dermaltrichomaihroepithelioma
- unilateral neurocutaneous basal cell carcinoma

Please turn to page 201 for the diagnosis.
Aderenal lesions presenting with a linear and/or dermatomal pattern rarely have been reported. Bolognia et al\textsuperscript{1} performed a comprehensive review of Blaschko lines and skin conditions that follow such a pattern. The authors found that adnexal-related lesions included linear nevus comedonicus, linear basal cell nevus with comedones (linear basaloid follicular hamartoma), unilateral nevoid basal cell carcinoma (BCC), linear trichoepithelioma, linear trichodiscoma, linear hamartoma of the follicular infundibulum, nevus sebaceous, syringocystadenoma papilliferum, porokeratotic eccrine ostial and dermal duct nevus, linear eccrine poroma, linear spiradenoma, linear syringoma, and linear eccrine syringofibroadenoma.\textsuperscript{1}

Trichoepithelioma is a hair follicle–related neoplastic lesion presenting most commonly as the autosomal-dominant multiple familial type with lesions mainly centered on the face. Initial genetic studies associated the disease with loss of heterozygosity in the 9p21 region and further studies identified mutations in the \textit{CYLD} (cylindromatosis [turban tumor syndrome]) gene on chromosome 16q12-q13.\textsuperscript{2,3} Unilateral, linear, and dermatomal forms of trichoepithelioma rarely are reported. In 1986, Geffner et al\textsuperscript{4} reported a case of linear and dermatomal trichoepithelioma in a 10-year-old girl. In addition to discrete solitary lesions affecting the face, she developed lesions on the left shoulder, left side of the trunk, and left lower leg following dermatomal distribution. In 2006, 2 cases of dermatomal trichoepitheliomas affecting the face in children, as in our case, were reported.\textsuperscript{5,6} Another case involving the neck was reported in 2016.\textsuperscript{7} Although classic multiple familial trichoepithelioma can be part of conditions such as Brooke-Spiegler\textsuperscript{8} and Rombo syndromes,\textsuperscript{9} no syndromal association has been reported thus far with the unilateral, linear, or dermatomal variants.

Our case showed typical histopathologic features of trichoepithelioma, including discrete islands of basaloid cells in the dermis set in a conspicuous fibroblastic stroma. Focal connection with the epidermis was present. Most of the islands showed peripheral palisading and horn cysts lined by eosinophilic cells. The fibroblastic component was tightly adherent to the epithelial component, and only stromal clefts were detected. Papillary mesenchymal bodies also were detected as oval aggregates of fibroblastic cells invaginating into epithelial islands to form hair papillae.

Histopathologically, the 2 most important differential diagnoses of trichoepithelioma include BCC and basaloid follicular hamartoma. In differentiating BCC from trichoepithelioma, the presence of dense fibroblastic stroma and papillary mesenchymal bodies characterize trichoepithelioma, while a fibromucinous stroma with mucinous retraction artifacts and clefting between the basaloid islands and the stroma characterize BCC (Figure 1).\textsuperscript{10} Immunohistochemical studies using antibodies against Bcl-2, CD34, CD10, androgen receptor, Ki-67, cytokeratin 19, and \textit{PHLDA1} (pleckstrin homology domain family A member 1) have reportedly been utilized to differentiate trichoepithelioma from BCC.\textsuperscript{11,12}

Basaloid follicular hamartoma is characterized by thin anastomosing strands and branching cords of undifferentiated basaloid cells that replace or associate hair follicles in a latticelike pattern (Figure 2). The strands usually

\textbf{FIGURE 1.} Basal cell carcinoma. Basaloid islands with peripheral palisading and peritumoral mucinous retraction artifacts (H&E, original magnification ×100).

\textbf{FIGURE 2.} Basaloid follicular hamartoma. Strands of immature basaloid cells replacing and associating follicular structures in a latticelike pattern (H&E, original magnification ×40).
are vertically oriented perpendicular to the epidermis. Peripheral palisading is possible, and the basaloid strands are surrounded with cellular connective tissue stroma. Tumor islands in eccrine poroma show broad connections with the epidermis and are composed of poroid cells that show evident ductal differentiation with eosinophilic cuticles (Figure 3).\(^\text{14}\) Spiradenoma is characterized by capsulated deep-seated tumorous nodules not connected with the epidermis and composed of light and dark cells with ductal differentiation and vascular stroma (Figure 4). Scattered lymphocytes within the tumor lobules and in the stroma also are seen. Eosinophilic hyaline globules rarely can be present.\(^\text{15}\)

Many pathologists consider trichoepithelioma as the superficial variant of trichoblastoma. According to the recent World Health Organization classification of benign tumors with follicular differentiation, trichoepithelioma is considered synonymous with trichoblastoma.\(^\text{16}\)

Trichoepitheliomas are benign tumors, and therapy is mainly directed at removal for cosmetic purposes. Several methods of removal are available including electrocautery, laser therapy, and surgery. Awareness of the possible dermatomal distribution of hair follicle and other adnexal-related conditions is important, and such lesions should be thought of in the differential diagnosis of unilateral and/or dermatomal lesions.

**REFERENCES**


**FIGURE 3.** Eccrine poroma. Tumor island composed of poroid cells with evident ductal differentiation with eosinophilic cuticles (H&E, original magnification ×100).

**FIGURE 4.** Spiradenoma. Light and dark cells with evident ductal differentiation and lymphocytic infiltrate within the tumor (H&E, original magnification ×400).