Desmoplastic Trichoepithelioma: Report of a Case Illustrating Its Natural History

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First described more than 30 years ago, desmoplastic trichoepithelioma is a rare but benign adnexal neoplasm. Most often identified in middle-aged individuals and females, desmoplastic trichoepithelioma usually is a solitary annular plaque. Though the tumors are benign, the possibility of malignant neoplasm may spark both clinical and histologic concern. A full-thickness skin biopsy is advisable when desmoplastic trichoepithelioma is suspected. A patient's clinical history may provide some clues to help guide diagnosis, as the tumors may be present for years and slow growth is commonly reported. We present a patient with desmoplastic trichoepithelioma that uniquely documents and supports the typical natural history of this tumor, as demonstrated by annual school photographs.

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Desmoplastic trichoepithelioma is a rare but benign adnexal neoplasm that was first described in 1976.[1,2] This tumor is most often identified in middle-aged individuals, and 85% of those affected are female.[2] Desmoplastic trichoepithelioma usually is solitary and measures less than 2 cm, appearing as a firm, white, often annular plaque.[2-4] The rim of this plaque may consist of coalescent papules.[2] Most cases of desmoplastic trichoepithelioma are sporadic; however, familial cases involving both solitary and multiple lesions have been reported.[5,6] Histologically, the tumors are characterized by a triad of narrow cellular strands, desmoplastic stroma, and keratinous cysts.[3,4]

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The natural history of desmoplastic trichoepithelioma is well-defined. Based on the histories of affected patients, the tumor often slowly expands over years and even decades.[3] We present a case of desmoplastic trichoepithelioma that uniquely documents this progression over time in annual school photographs.

Case Report

A 29-year-old woman presented postpartum for evaluation of changing melanocytic nevi and was incidentally noted to have a concerning lesion on her mid left cheek. She reported that the lesion had slowly expanded over time and recalled first being aware of its presence 24 years earlier while in kindergarten.

Physical examination revealed a 0.8×1.2-cm, firm, annular plaque with central depression on the mid left cheek (Figure 1). The nevi appeared unremarkable and no noteworthy actinic damage was identified elsewhere on the skin. After examination, she provided all of her annual school photographs from kindergarten through the 12th grade, which

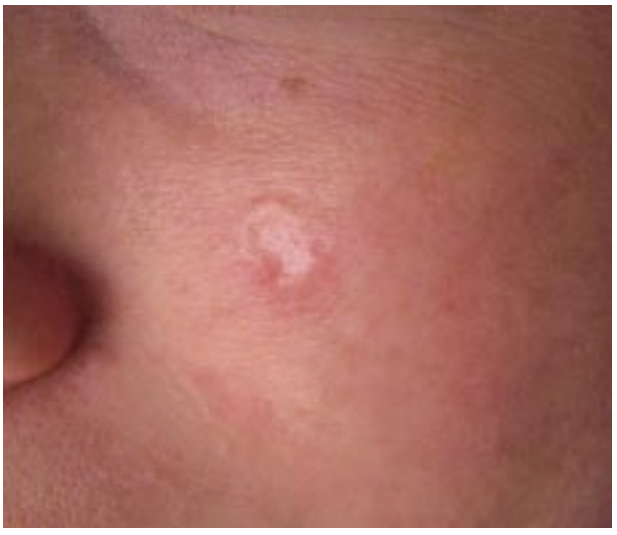

Figure 1. Annular plaque on the patient's mid left cheek at presentation (aged 29 years).

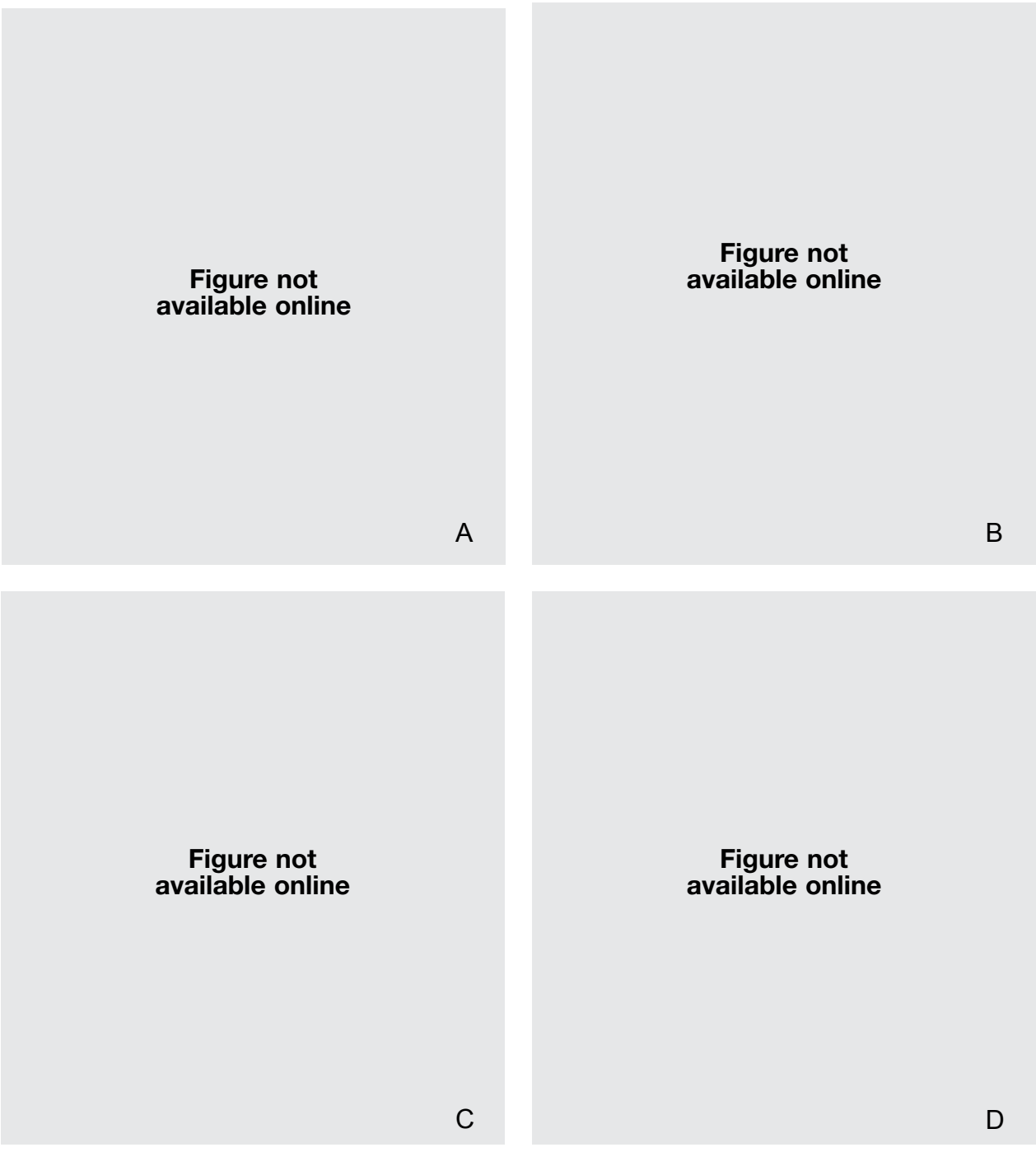

Figure 2. Annular plaque on the patient's mid left cheek demonstrating slow growth of the lesion (aged 6 years [first grade][A]; aged 8 years [third grade][B]; aged 13 years [eighth grade][C]; aged 16 years [11th grade][D]).

clearly demonstrated the slow growth of the lesion (Figure 2). Based on the patient's history and physical examination findings, a diagnosis of desmoplastic trichoepithelioma was strongly suspected, but a 4-mm punch biopsy was performed for confirmation. The histopathologic examination of the biopsy specimen revealed narrow strands of cells, desmoplastic stroma, and keratinous microcyst formation in the upper dermis, which were consistent with desmoplastic trichoepithelioma (Figure 3).

Given the benign and asymptomatic nature of this tumor, a collective decision was made to proceed with longitudinal observation. Upon reexamination 6 months later, the lesion was unchanged.

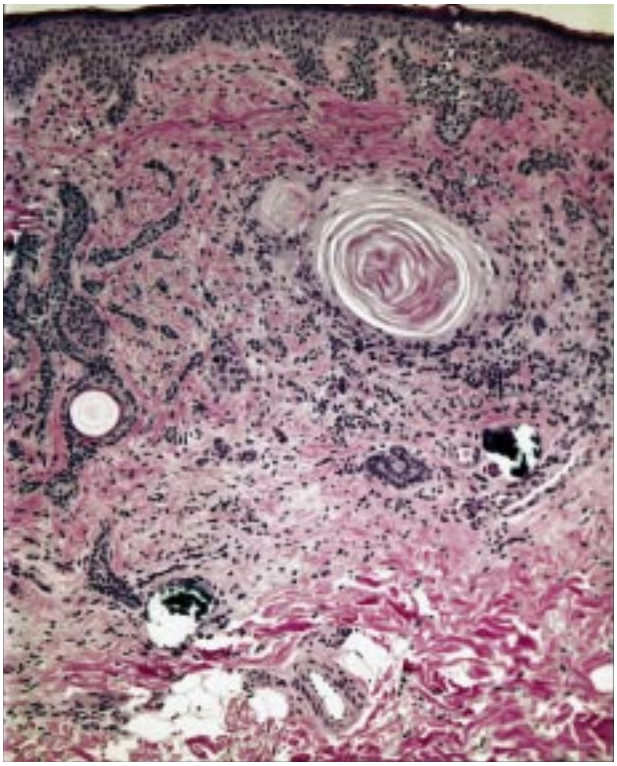

Figure 3. Photomicrograph of the biopsy specimen showing narrow cellular strands, desmoplastic stroma, and keratinous microcysts (H&E, original magnification ×10).

Comment

Although desmoplastic trichoepithelioma is benign, a full-thickness skin biopsy usually is required to exclude other possibilities, especially certain malignancies. The clinical differential diagnosis includes morpheaform basal cell carcinoma, microcystic adnexal carcinoma, granuloma annulare, sebaceous hyperplasia, sarcoidosis, and scar tissue.[2,4] Histologically, desmoplastic trichoepithelioma may share some features of the following tumors: morpheaform basal cell carcinoma, syringoma, desmoplastic tumors metastatic to the skin, microcystic adnexal carcinoma, trichoadenoma, and basaloid follicular hamartoma[2,3]; however, the histology usually is distinctive. In the seminal work by Brownstein and Shapiro[1,2] describing desmoplastic trichoepithelioma, the critical histologic triad of tumor cells in narrow strands, desmoplastic stroma, and keratinous cysts was identified. Of critical importance is a full-thickness skin biopsy to help differentiate desmoplastic trichoepithelioma from microcystic adnexal carcinoma.[3] All 3 of these standard microscopic features were present in the histopathologic examination of our patient; the patient's clinical history and photographic documentation of the lesion further solidified the diagnosis.

Reviewing a patient's clinical history may be helpful in diagnosing desmoplastic trichoepithelioma, given the reported tendency of these tumors to slowly expand. Case reports from the literature demonstrate that desmoplastic trichoepithelioma may be present from 1 to 40 years or more prior to diagnosis.[1-7] Furthermore, slow growth often is reported.[3-5,7] Our patient uniquely demonstrated the potential for slow progression of the lesion by means of her annual school photographs.

Treatment of this rare neoplasm is not standardized but generally involves either longitudinal observation or surgical approaches. In the original cases presented by Brownstein and Shapiro,[2] electrodesiccation and curettage were used to treat most tumors, with no reported adverse occurrences. Excision, dermabrasion, and laser ablation also may be considered.[4] In a few other cases, longitudinal observation alone was selected and no substantial changes were reported over variable follow-up intervals.[6] If malignancy is excluded, tailoring treatment on a case-by-case basis after a thorough discussion with the patient is recommended.

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