

CUTANEOUS NODULE ON THE FACE: ADAMANTINOID TRICHOBLASTOMA - A RARE, UNIQUE TUMOR

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Abstract

Adamantinoid trichoblastoma is a rare adnexal tumor which clinically masquerades as various benign and malignant lesions. Less than 50 cases have been documented so far. In this report we discuss the clinicopathological features of this rare and fascinating tumor with a brief review of literature.

Key words: cutaneous lymphadenoma; adamantinoid trichoblastoma; tumor infiltrating lymphocytes

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Introduction

Adamantinoid trichoblastoma (AT) is an uncommon benign skin adnexal (follicular) neoplasm with a prominent lymphocytic infiltrate and an adamantinoid appearance (similar to dental adamantinoma). It was originally described as “lymphoepithelial tumor of the skin” by Santa Cruz and Barr in 1987 and was later renamed as “cutaneous lymphadenoma” (CL) in 1991 [1]. Currently many authors believe it to be a variant of trichoblastoma, a benign follicular tumor with both epithelial and mesenchymal components. It is a rare tumor with fewer than 50 cases reported in the world literature.

Case Report

A 42 year old lady presented with a firm, nodular, slowly progressive swelling in the face of 1 year duration. Clinically it had the appearance of a keloid /neurofibroma. Surgical excision of the lesion was performed and received in our laboratory for histopathological study.

Pathological findings:

Grossly, the excised skin covered tissue measured 1.3 x 0.8 x 0.5 cm and showed grey white areas on cut section. Sections showed thinned out epidermis overlying an unencapsulated, well circumscribed dermal tumor composed of irregular islands of epithelial nests with palisading basaloid cells at the periphery. Central areas within the islands showed large polygonal cells with clear cytoplasm with a dense lymphocytic infiltrate, edema, histiocytes, giant cells, focal ductal and

follicular differentiation along with Reed Sternberg like cells. Surrounding stroma showed dense fibrosis with lymphocytic infiltrates (Fig. 1-4).

Histopathological diagnosis of *Nodular Adamantinoid trichoblastoma (Cutaneous lymphadenoma)* was made.

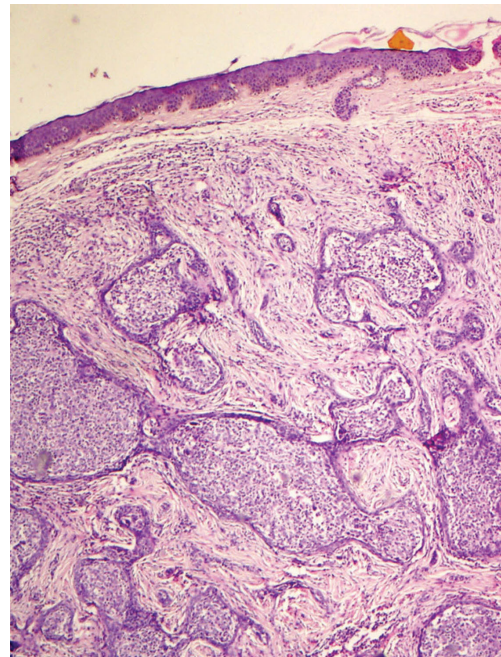


Figure 1. Section shows thinned out epidermis overlying a tumor composed of irregular epithelial nests with peripheral palisading basaloid cells. H&E, 100X

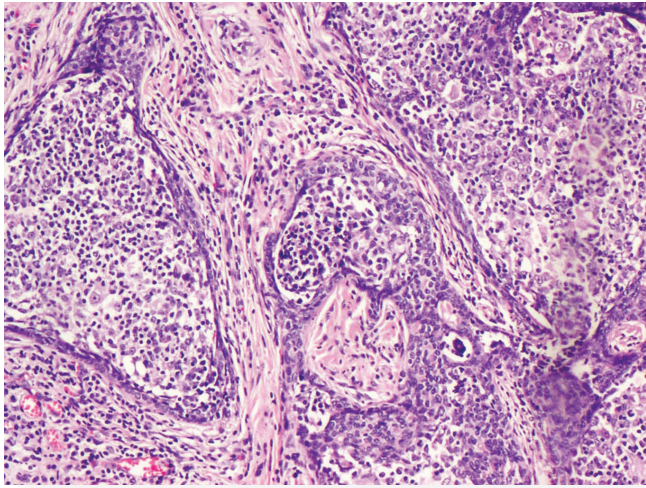


Figure 2. The epithelial nests showing lymphocytic infiltrates surrounded by desmoplastic stroma. H&E, 200X

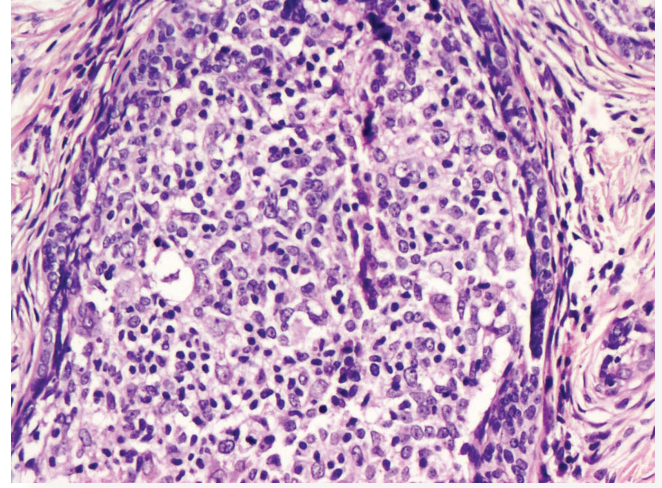


Figure 3. Higher power view of the epithelial islands showing cells with clear cytoplasm, lymphocytes and histiocytes. H&E 400X

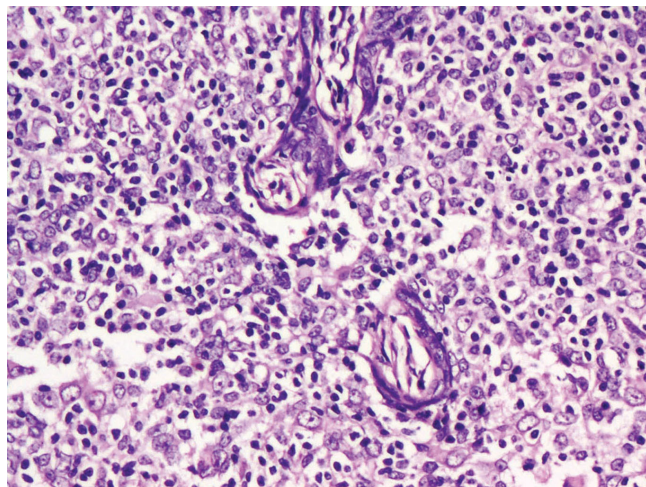


Figure 4. Higher power view of the epithelial nests shows scattered large Reed Sternberg-like cells. H&E, 400X

Discussion

AT/CL is an uncommon skin adnexal tumor usually presenting as an asymptomatic, small, dome-shaped, flesh-colored papule or nodule, typically in the head and neck region. They are commonly seen in young to middle aged adults with a predominance of male patients. Unlike other skin adnexal tumors, the description of these tumors is limited to random case reports [2]. Less than 50 such cases have been reported in the world literature.

Possibly because of their rarity, ATs are generally misinterpreted clinically as basal cell carcinoma, adnexal tumor, nevus or dermatofibroma [2]. The present case was clinically thought to be a keloid/neurofibroma.

Microscopic examination demonstrates a triphasic tumor composed of cell nests with palisading basaloid cells at the periphery (epithelial component), lymphoid infiltrate and desmoplastic stroma (mesenchymal component). There can also be large cells that resemble Reed Sternberg cells within the lobules. Focal ductal, follicular and sebaceous differentiation, central keratinization and stromal mucinosis have been described [3]. The tumor is unencapsulated and generally well circumscribed, but may have infiltrating outlines making way for tumor recurrence [2].

The histological differential diagnosis includes - clear cell

Basal cell carcinoma, clear cell syringoma, trichoepithelioma and malignant lymphoepithelioma - like carcinoma. However, a lymphoid cell infiltrate within the tumor lobules is not prominent in such tumors [4]. In younger patients it has to be differentiated from dermal thymus, which is an aberrant location of thymic tissue in the skin, due to defective migration, and presents as linear symmetric ulcerated scar-like lesions in the neck in patients with other facial - branchial abnormalities [2].

Several proposals have been made regarding the possible histogenesis of this tumor. Santa Cruz et al and Santa Cruz and Barr originally suggested immature pilosebaceous differentiation [5,6]. Others proposed an eccrine origin suggesting the term “lymphotropic solid syringoma” and “lymphotropic eccrine benign tumor” [7,8]. Dahill and Seywright [9] reported a case of synchronous occurrence of cutaneous lymphadenoma and syringoid eccrine carcinoma in a single patient providing additional evidence to support eccrine differentiation. Fillipo et al [10], argued that AT/CL is not a distinct entity but may represent a basal cell carcinoma with possibly pilar or eccrine differentiation. Mc Niff et al [11], evaluated the immunohistochemical staining patterns of CK20 (merkel cells), bcl2 (epithelial), S100 and CD1a (Langerhans cells) and CD34 (stromal cells) in CL, trichoblastomas and nodular basal cell carcinomas. They observed similar staining patterns for the first three markers in CL and trichoblastomas (importantly peripheral staining of bcl2 in tumor lobules) and opined that CLs are in-fact a variant of trichoblastomas. Most recently, these tumors have been classified as an adamantinoid variant of trichoblastoma, considering its resemblance to dental adamantinoma, a benign epithelial and mesenchymal tumor of oral cavity [2,12,13]. The marked lymphocytic infiltrate in these tumors is thought to be a result of either defective lymphocyte-epithelial interaction or an exuberant host response to the tumor cells [14,15].

A benign clinical course has been described in the literature for these tumors [1-15]. Surgical excision of the tumor is the definitive treatment. However the tendency to local infiltration may result in tumor recurrence in cases of incomplete removal. To conclude, AT is a rare benign adnexal tumor which can clinically masquerade as a variety of benign and malignant lesions.

Histologically, it is a triphasic tumor with epithelial islands, lymphocytic infiltrates and desmoplastic stroma. AT has been called by different names in the past which include, lymphoepithelial tumor of the skin, lymphotropic solid syringoma, lymphotropic eccrine benign tumor and cutaneous lymphadenoma. The term AT describes the close relationship of this tumor with trichoblastoma and its peculiar adamantinoid appearance in tissue sections, but it does overlook the prominent intra-tumoral lymphocytic infiltrate, which is a characteristic feature of this tumor. The authors thus feel that 'Lymphotropic AT' would be a better terminology to describe this rare tumor.

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