

Desmoplastic Trichepithelioma: A Case Report

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Abstract

Desmoplastic trichoepithelioma is an uncommon benign skin cancer, considered to have follicular differentiation. It is a well-defined version of trichoepithelioma due to its unique clinical and histopathological characteristics. It occurs in an incidence of two per 10,000 and corresponds to less than 1% of all skin tumors. It is usually seen in middle-aged women, but it has been reported in all age groups and genders. Although clinically rare, desmoplastic trichoepithelioma is difficult to distinguish from other skin lesions caused by certain diseases, such as basal cell carcinoma. Therefore, it is important to assist in the proper recognition, diagnosis and treatment of this disease.

Keywords: *Desmoplastic trichoepithelioma; Trichoepithelioma; Dermoscopy*

1. Introduction

Desmoplastic trichoepithelioma is an uncommon benign skin cancer deriving from basal cells in the outer root sheath of hair follicles. Its unique clinical and histopathological features turn it into a well-defined version of trichoepithelioma [1].

This disease affects 2 in 10,000 individuals and accounts for less than 1% of all skin tumors. It is oftentimes diagnosed in middle-aged women, although it has been reported in individuals belonging to all age groups and to both sexes [2].

It overall emerges as asymptomatic, skin-colored, solitary, annular, indurated and centrally depressed papules or plaques. The most often affected body sites comprise those exposed to the sun, mainly facial areas such as cheeks, chin and forehead; whereas the least affected ones comprise the upper trunk, neck and scalp [3].

Trichoepithelioma presents clinical and histological features similar to those of basal cell carcinoma; thus, differentiating these tumor types can be a diagnostic challenge [4].

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These tumors must be surgically removed due to their fast growth, as well as to aggressive histological features that can be misdiagnosed as basal cell carcinoma and to their trend to appear in cosmetically and functionally sensitive body sites [3].

2. Case Description

A 32-year-old female patient, with no significant pathological history, has sought medical attention due to facial skin lesion associated with pruritus, which persisted for approximately six years. Dermatological examination has evidenced a plaque (approximately 9 mm), with raised edges and atrophic core, in the left infrazygomatic region (FIG. 1).



FIG. 1. Plaque with raised edge and atrophic core measuring approximately 9 mm in the left infrazygomatic region.

Dermatoscopy has shown tumor with well-defined edges, ivory-white color, cystic structures on the upper edge, brownish pigmentation nests on the lower edge and central atrophy (FIG. 2). Incisional biopsy of the lesion was carried out after diagnostic hypotheses of desmoplastic trichoepithelioma, sebaceous nevus and granuloma annulare centrifugum; patient's return was scheduled. Histopathological examination has evidenced epithelial neoplasm connected to the epidermis. It presented narrow cords of small basaloid cells, keratin cysts and in-between fibroblastic stroma; it did not show atypia or necrosis (FIG. 3 and 4).

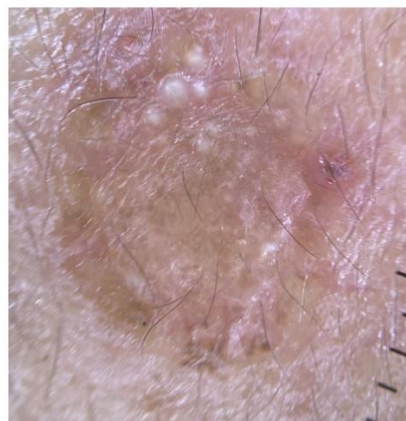


FIG. 2. Tumor with well-defined edges, ivory-white color, cystic structures on the upper edge, brownish pigmentation nests on the lower edge and central atrophy.

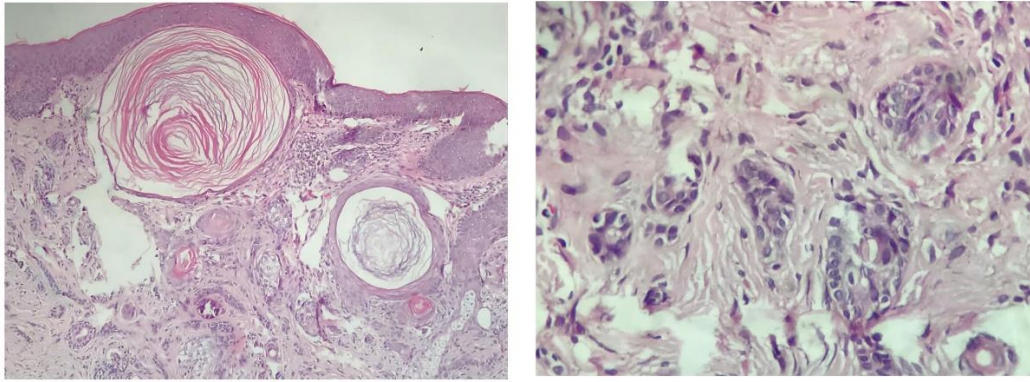


FIG. 3 & 4. Epithelial neoplasm connected to the epidermis. It presents narrow cords of small basaloid cells, keratin cysts and in-between fibroblastic stroma, but it does not present atypia or necrosis.

Thus, the skin lesion was subjected to excisional biopsy in the operating room, without complications, after patient's consent. Biopsy results have confirmed the diagnosis of desmoplastic trichoepithelioma (measuring approximately 9 mm) affecting both the superficial and middle dermises.

3. Discussion

Desmoplastic trichoepithelioma is a relatively rare benign skin neoplasm, whose microscopic and histological features were first described by Brownstein and Shapiro [2]. It often emerges as slow-growing, asymptomatic, solitary and indurated plaques or papules. These lesions present raised annular edge and non-ulcerated depressed core. Assumingly, it most often affects the right side of the face (measuring 3 to 8 mm, in diameter), mostly on the cheek, which is followed by the nose, chin, forehead, periorbital region and lips [5]. It is often diagnosed in middle-aged women, although it has been reported in individuals belonging to all age groups and to both sexes [5].

Differentiating the clinical diagnosis of desmoplastic trichoepithelioma from that of basal cell carcinoma, mainly from its morpheaform variant, can be a hard task to be accomplished [6]. Associating dermatoscopy with naked eye examination can help making the correct diagnosis. Focused arboriform vessels are features often seen in basal cell carcinomas during dermatoscopy examinations, although similar findings can be observed in desmoplastic trichoepithelioma cases. However, more specific criteria are available in the literature for the diagnosis of desmoplastic trichoepithelioma, such as the incidence of ivory-white background throughout the lesion, with multiple arboriform vessels and whitish globules [7-9]. Findings in the herein reported case - whose lesion presented ivory-white background, cystic structures and brownish pigmentation nests - were more specific to desmoplastic trichoepithelioma, as shown in FIG. 2.

Narrow tumor cords with one to three rows of basaloid tumor cells, prominent oval core and sparse cytoplasm; corneal cysts, some of them with drop-shaped epithelial projections resembling eccrine ducts; and desmoplastic stroma with dense and hypocellular collagen are the three histopathological features used to diagnose desmoplastic trichoepithelioma. Furthermore, calcification sites can be seen in corneal cysts and foreign body granulomas [10]. All fundamental aspects associated with the diagnosis of desmoplastic trichoepithelioma were found in the histopathology of the herein reported case, as shown in FIG. 3 and 4.

Differential diagnosis must be conducted to rule out morpheaform basal cell carcinoma, classic trichoepithelioma, sebaceous hyperplasia, annular granuloma, scarring and scleroderma. Histopathological differentiation must also be conducted, mainly to rule out morpheaform or fibrosing basal cell carcinoma, microcystic adnexal carcinoma, syringomas, basaloid follicular hamartoma and trichoadenoma [11].

Local surgical excision associated with frozen section margin control is the treatment of choice for desmoplastic trichoepithelioma [12]. Despite the benign diagnosis, these tumors can penetrate deeper skin layers and present aggressive histological features. Furthermore, they mostly affect cosmetically sensitive sites, whose healthy tissue preservation is of utmost importance. These were the reasons why excisional biopsy was the treatment of choice to prevent disease recurrence and subclinical local tumor invasion in the present case. Dermabrasion, electrosurgery and laser stand out among therapeutic options with high risk of disease recurrence.

4. Conclusion

Desmoplastic trichoepithelioma is clinically hard to be differentiated from other skin lesions. Therefore, it is important conducting the proper identification, diagnosis and treatment of this disease. Nowadays, dermatoscopy appears to be a valuable tool to assess pigmented skin lesions; consequently, the application of this technique has been increasingly indicated in different contexts. The current study has confirmed observations available in the literature and it can be used to help differentiating desmoplastic trichoepithelioma from basal cell carcinoma. However, further case studies should be conducted to permanently validate its differentiation criteria.

5. Conflicts of Interest

The authors declare no conflicts of interest.

6. Financial Support

The current study did not receive any financial contribution, grant or scholarship.

7. Patient Consent

Granted

REFERENCES

1. Rahman J, Tahir M, Arekemase H, et al. Desmoplastic Trichoepithelioma: Histopathologic and Immunohistochemical Criteria for Differentiation of a Rare Benign Hair Follicle Tumor from Other Cutaneous Adnexal Tumors. *Cureus*. 2020;12(8):e9703.
2. Brownstein MH, Shapiro L. Desmoplastic trichoepithelioma. *Cancer*. 1977;40(6):2979-86.
3. Mamelak AJ, Goldberg LH, Katz TM, et al. Desmoplastic trichoepithelioma. *J Am Acad Dermatol*. 2010;62(1):102-06.

4. Wee SJ, Park MC, Chung CM. Basal cell carcinoma misdiagnosed as trichoepithelioma. *Arch Craniofac Surg.* 2020;21(3):202-05.
5. Wang Q, Ghimire D, Wang J, et al. Desmoplastic trichoepithelioma: A clinicopathological study of three cases and a review of the literature. *Oncol Lett.* 2015;10(4):2468-76.
6. Poniecka AW, Alexis JB. An immunohistochemical study of basal cell carcinoma and trichoepithelioma. *Am J Dermatopathol.* 1999;21(4):332-6.
7. Ardigo M, Zieff J, Scope A, et al. Dermoscopic and reflectance confocal microscope findings of trichoepithelioma. *Dermatology.* 2007;215(4):354-8.
8. Liebman TN, Jaimes-Lopez N, Balagula Y, et al. Dermoscopic features of basal cell carcinomas: differences in appearance under non-polarized and polarized light. *Dermatol Surg.* 2012;38(3):392-9.
9. Khelifa E, Masouyé I, Kaya G, et al. Dermoscopy of desmoplastic trichoepithelioma reveals other criteria to distinguish it from basal cell carcinoma. *Dermatology.* 2013;226(2):101-04.
10. Takei Y, Fukushima S, Ackerman AB. Criteria for histologic differentiation of desmoplastic trichoepithelioma (sclerosing epithelial hamartoma) from morphea-like basal-cell carcinoma. *Am J Dermatopathol.* 1985;7(3):207-21.
11. Wang Q, Ghimire D, Wang J, et al. Desmoplastic trichoepithelioma: A clinicopathological study of three cases and a review of the literature. *Oncol Lett.* 2015;10(4):2468-76.
12. Behroozan DS, Goldberg LH, Glaich AS, et al. Mohs micrographic surgery for deeply penetrating, expanding benign cutaneous neoplasms. *Dermatol Surg.* 2006;32(7):958-65.