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Case Report

Desmoplastic trichoepithelioma; a new case and review of the literature with emphasis on its differential diagnosis

結締組織增生性毛髮上皮瘤；新病例一則和文獻綜述，並偏重探討其鑑別診斷

M Concha-Rogazy, S Alvarez-Veliz, P Del Barrio-Díaz, JE Carrasco-Zuber, S Gonzalez-Bombardiere

Trichoepithelioma is a benign neoplasm with hair follicle differentiation that may present as solitary, multiple or desmoplastic forms. This report describes a case of desmoplastic trichoepithelioma, an uncommon adnexal tumour with an incidence of 2 per 10,000. Desmoplastic trichoepithelioma is a benign lesion, clinically and histologically similar to other cutaneous tumours, and presents a true diagnostic challenge. The clinical criteria, histopathological features and immunohistochemical profile are discussed with emphasis on its differential diagnosis.

毛髮上皮瘤爲良性瘤細胞增生，可呈單一、多個或在結締組織中，在向毛髮結構分化過程中形成。此報告描述了一宗結締組織增生性毛髮上皮瘤——一種罕見的皮脂腺上皮瘤，發病率約爲每萬人中兩例。毛髮上皮瘤是一種良性病變，從臨床學及組織學的角度來講與其他皮膚腫瘤類似，因而構成了診斷上的困難。本報告探討其臨床診斷標準、組織病理特點以及免疫組化概 要，並注重研究其鑒別診斷。

Keywords: Cutaneous tumours, adnexal neoplasms, basal cell carcinoma, skin cancer

關鍵詞：皮膚腫瘤、皮脂腺上皮瘤、基底細胞癌、皮膚癌

Department of Dermatology, School of Medicine, Pontificia Universidad Católica de Chile, Santiago de Chile

M Concha-Rogazy, MD
S Alvarez-Veliz, MD
JE Carrasco-Zuber, MD

Department of Pathology, School of Medicine, Pontificia Universidad Católica de Chile, Santiago de Chile

S Gonzalez-Bombardiere, MD

School of Medicine, Pontificia Universidad Católica de Chile, Santiago de Chile

P Del Barrio-Díaz, MD

Correspondence to: Dr. JE Carrasco-Zuber

Department of Dermatology, Centro Médico San Joaquín, Pontificia Universidad Católica de Chile, Santiago de Chile, Av. Vicuña Mackenna 4686, Macul, Santiago, Chile

Introduction

Trichoepithelioma is a benign adnexal tumour of the outer sheath of the pilosebaceous follicle. There are three clinical variants of trichoepithelioma: solitary, multiple, and desmoplastic.

Desmoplastic trichoepithelioma (DTE) was described in 1977 by Brownstein and Shapiro, when it was differentiated from the classic isolated trichoepithelioma.[1] It usually presents as an asymptomatic, slowly growing plaque on the face of young women. Clinical examination reveals an annular, skin-coloured, firm plaque or nodule with a depressed centre and raised border.[2]

On dermoscopy, DTE shows well-defined borders and an ivory-white to yellowish colour, as well as prominent arborising telangiectasias in the central and peripheral areas of the lesion.[3]

Histopathologically, it is characterised by a triad of cords of basaloid tumour cells, small keratinous cysts and desmoplastic stroma.[1]

Making a correct diagnosis of DTE is important because it may be mistaken for morphoeaform basal cell carcinoma (BCC) and microcystic adnexal carcinoma, clinically and histologically.[2]

Case report

A 31-year-old female presented to our clinic with a yellowish papule, which was occasionally pruritic, on her left cheek. The patient claimed that the lesion had been present for 10 years. She had an unrevealing medical history and her current medication included oral contraceptive pills only. The patient had no history of facial sunburn or trauma. She denied any personal or family history of skin cancer and none of her family members had a history of similar lesions.

On examination, an annular, firm papule, yellowish in colour, measuring 0.5x0.5 cm was noted on her left cheek. The centre was depressed but not ulcerated and had a raised border (Figure 1). On dermoscopy, the lesion had well-defined borders and showed a yellowish colour, as well as prominent, arborising telangiectasias in the central and peripheral areas (Figure 2). On physical examination, no other findings were seen on the skin or nails.

An excisional biopsy specimen was taken from the lesion with the provisional diagnosis of sebaceous carcinoma and adnexal tumour. Histopathological examination showed narrow strands of basaloid tumour cells, keratinous cysts and a desmoplastic stroma (Figure 3). A higher power view clearly showed cords and strands of epithelial cells with scanty inflammation, small keratinous cysts and dense collagenous stroma; all findings were consistent with a diagnosis of desmoplastic trichoepithelioma (Figure 4). After establishing the diagnosis, we performed surgical extirpation of the scar with a 2 mm margin for assessment. The biopsy showed no residual tumour and the cosmetic result was good.

Discussion

Desmoplastic trichoepithelioma is an uncommon benign adnexal tumour with an incidence estimated to be 2 per 10,000. It is a variant of trichoblastoma/trichoepithelioma with extensive stromal sclerosis. It appears any time from birth to the fourth decade of life, with a slow growth pattern and no discernible inheritance pattern.[2]

This entity presents as an asymptomatic, solitary, firm, skin-coloured to greyish, sclerotic annular plaque of about one centimetre in diameter.[2] Its centre is frequently depressed, but not ulcerated, and its border is raised, sometimes with a ring of papules, like those of granuloma annulare.[1] It is usually found on the upper cheek or angle of the mouth of a young female and is often mistaken for morphoeaform BCC or microcystic adnexal carcinoma. Multiple lesions are quite rare.[2]

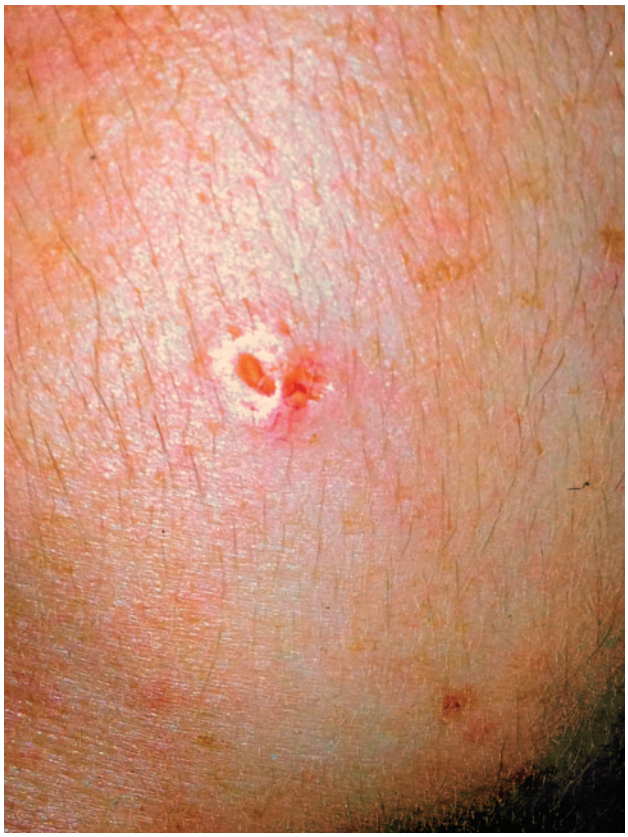

Figure 1. Annular and yellowish papule on the left cheek. The centre was depressed but not ulcerated and had a raised border.

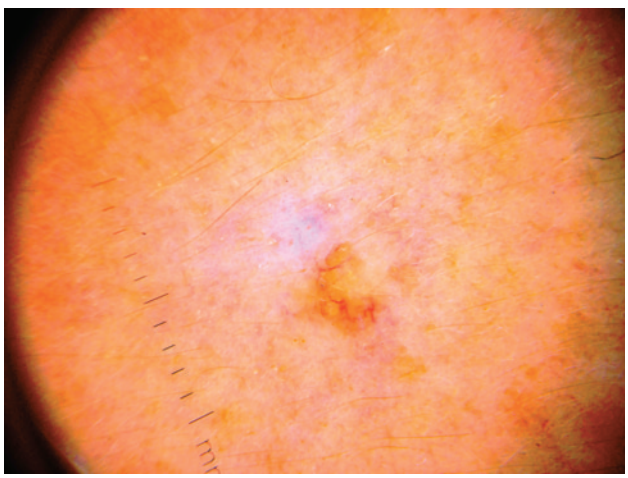

Figure 2. Dermoscopic view of the lesion. There was a well-defined border and yellowish colour as well as prominent arborising telangiectasias in the central and peripheral areas.

On dermoscopy, DTE shows well-defined borders and an ivory-white to yellowish colour, as well as prominent arborising telangiectasias in the central and peripheral areas.[3]

Histopathologically, DTE is characterised by a triad of cords of basaloid tumour cells, small keratinous cysts and a characteristic desmoplastic stroma with tumour-stromal retractions. The tumour shows follicular or sebaceous differentiation, and typically occurs in the upper two-thirds of the dermis.[1]

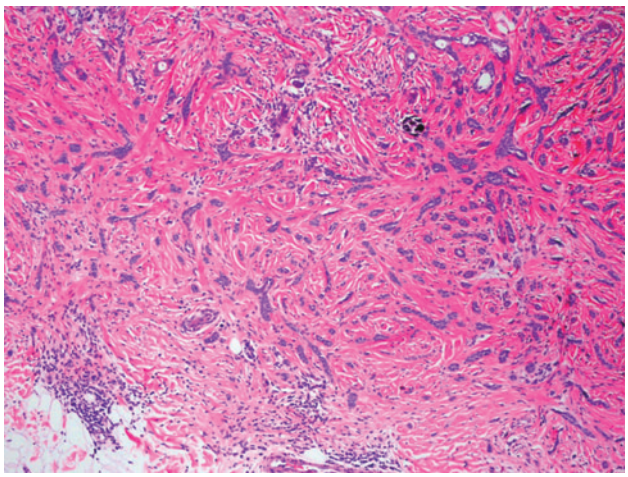

Figure 3. Dermal tumour with narrow strands of basaloid cells, keratinous cysts, and a desmoplastic stroma (H & E, 100 x).

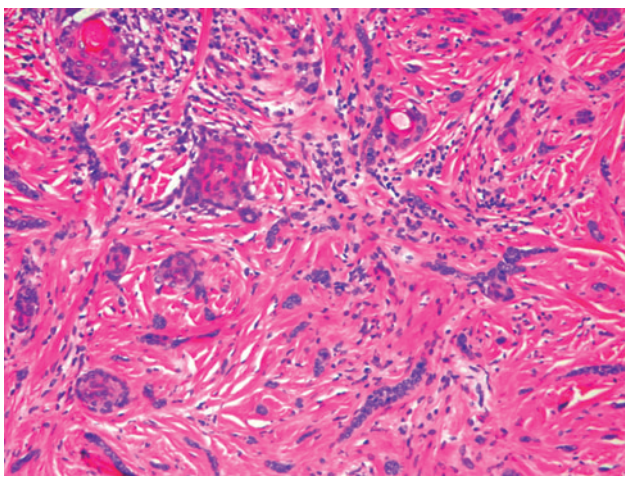

Figure 4. Cords and strands of epithelial cells showing small keratinous cysts and dense collagenous stroma with scanty inflammation (H & E, 200 x).

Clinically, the differential diagnosis includes BCC, sebaceous hyperplasia, conventional trichoepithelioma, granuloma annulare and scleroderma. Histologically, DTE may resemble morphoeaform BCC, microcystic adnexal carcinoma, trichoepithelioma, trichoadenoma and syringoma.[2] It is important to distinguish these neoplasms because they have different disease courses requiring different therapeutic decisions.

Dermoscopy is a non-invasive diagnostic technique which allows the visualisation of morphological features not visible to the naked eye, thus allowing a more accurate diagnosis. Basal cell carcinoma and DTE both have arborising telangiectasias and focal "shiny-white" areas. The difference between them is the ivory-white to yellowish colour of the entire lesion in DTE, with other dermoscopic features of BCC such as leaf-like structures and the absence of ovoid nests.[3]

Immunohistochemistry is often used as another differentiating diagnostic tool in difficult cases. The spindle-shaped cells surrounding the cellular islands in desmoplastic trichoepithelioma are focally but strongly positive for CD34, whereas the stromal cells around BCCs and microcystic adnexal carcinomas are usually negative.[4] Diffuse Bcl-2 positivity is reported in BCC, whereas the basal layer alone is highlighted in DTE.[5] However, some authors have found that this is variable and unreliable in practice.[6]

Immunohistochemical stains for androgen receptor (AR) and cytokeratin 20 (CK20) are useful to differentiate DTE from BCC. The AR-, CK20+ immunophenotype is sensitive (87%) and specific for DTE (100%). The AR+, CK20- immunophenotype is specific (100%) and moderately sensitive (61%) for BCC.[7] CK20 marks Merkel cells dispersed throughout areas of DTE. However, small biopsy size and haphazard distribution of single Merkel cell are factors limiting the utility of CK20 because cells expressing the marker may be absent in the biopsy section by chance.[8]

Since it is a benign lesion, DTE needs no treatment. However, the rapid growth and aggressive histologic features often observed in DTE can cause diagnostic uncertainty and it is often confused with malignancies.[2] Furthermore, this entity has a predilection for cosmetically and functionally sensitive areas where sparing of normal tissue is of the utmost importance. It is for these reasons that Mohs micrographic surgery is the treatment of choice for this "benign" neoplasm to prevent recurrence and subclinical local invasion.[9]

One report suggests that topical 5% imiquimod may be employed as an adjunctive treatment, when combined with surgery.[10] Other alternatives such as dermabrasion and laser surgery may be associated with a considerable rate of recurrence.[11,12]

Conclusion

In summary, we have presented a female patient with desmoplastic trichoepithelioma with a slow-growing yellowish papule on her left cheek. The clinical and histological differentiation between desmoplastic trichoepithelioma, morphoeaform basal cell carcinoma and microcystic adnexal carcinoma is challenging. Because most of these tumours occur in functionally and cosmetically sensitive anatomic locations, accurate diagnosis and distinction between benign and malignant lesion is crucial in making appropriate patient management plan.

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