Desmoplastic trichoepithelioma: an uncommon adnexal tumour with a predilection for cosmetically sensitive sites

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Introduction

Desmoplastic trichoepithelioma is an uncommon adnexal tumour (Rahman et al, 2020) predominantly affecting young women in cosmetically sensitive sites (Mamelak et al, 2010). Detection is often incidental and histological diagnosis can be impeded by biopsies that are insufficiently deep (Mamelak et al, 2010). Clinically and histologically it can mimic morphoeic basal cell carcinoma (Brownstein and Shapiro, 1977). This article presents the case of a 31-year-old woman who presented with a desmoplastic trichoepithelioma on the cheek.

Case report

A 31-year-old white woman presented with a 10-year history of a lesion on her left inner cheek. It had increased in size over the preceding months but had not caused any symptoms. The patient had Fitzpatrick phototype II skin and had lived in Barcelona for 5 years. There was no relevant medical history. Physical examination revealed a 5 mm diameter, pale, white plaque with telangiectasia on the medial left cheek, abutting the nasal sidewall (Figure 1).

A diagnostic punch biopsy was obtained from the lesion and histopathological examination revealed micronodules containing basaloid cells with possible ductal differentiation (Figures 2a and b). There was mild fibrosis of the stroma and no epidermal involvement. The patient was referred for Mohs micrographic surgery and subsequently discharged.

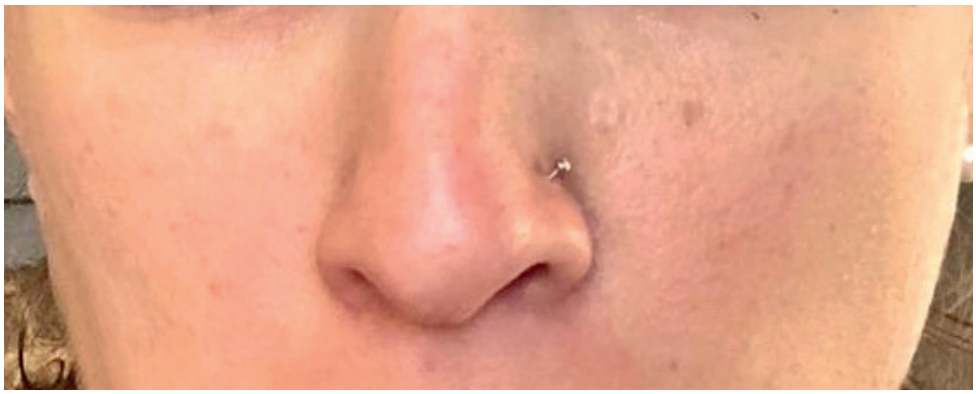

Figure 1. Pale, annular white plaque with central indentation, on the left medial cheek.

Discussion

Desmoplastic trichoepithelioma (aka sclerosing epithelial harmatoma) is an uncommon benign cutaneous neoplasm, affecting 1 in 5000 adults in the UK (Rahman et al, 2020). It was first described by Zeligman in 1960 as a solitary trichoepithelioma. In 1977, Brownstein and Shapiro published a case series of 49 patients with this tumour, reporting the clinical and histopathological features as well as coining the term desmoplastic trichoepithelioma. It most commonly occurs in young to middle-aged women with a predilection for the cheek, forehead and chin (Mamelak et al, 2010). It is a slow-growing, asymptomatic (Brownstein and Shapiro, 1977; Mamelak et al, 2010; Rahman et al, 2020) tumour (although there may be an accelerated growth phase), that is often found incidentally (Moynihan et al, 2011). It typically presents as a solitary 2–18 mm, white, yellowish or skin-coloured, annular papule

How to cite this article:
Phillips D, Chaudhry IH, Morton JD, West EA. Desmoplastic trichoepithelioma: an uncommon adnexal tumour with a predilection for cosmetically sensitive sites. Br J Hosp Med. 2022. https://doi.org/10.12968/hmed.2021.0437

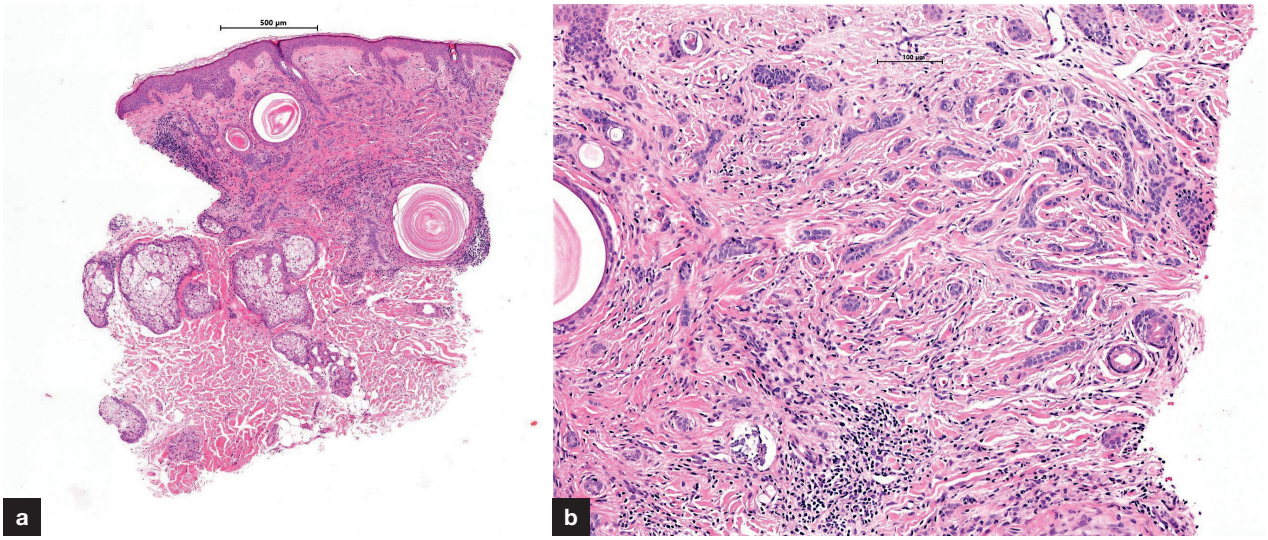

Figure 2. a. A low power view (36x magnification) shows keratocysts and basaloid tumour cells. b. A high power (140 x magnification) section shows strands of basaloid cells of a trichoepithelioma.

or plaque, with central indentation and a raised border (Brownstein and Shapiro, 1977; Mamelak et al, 2010; Rahman et al, 2020). It can be difficult to differentiate these tumours clinically from morphoeic basal cell carcinomas, localised morphoea, sebaceous hyperplasia and trichoepitheliomas (Brownstein and Shapiro, 1977). Dermoscopy may be a helpful diagnostic tool. Desmoplastic trichoepitheliomas and basal cell carcinomas share some dermoscopic features such as arborising telangiectasia and crystalline structures, but the presence of cyst-like structures may be more suggestive of the former (Khelifa et al, 2013).

Diagnosis is made through a full thickness biopsy of the lesion and subsequent histological examination (Rahman et al, 2020). Desmoplastic trichoepitheliomas tend to be confined to the upper two-thirds of the reticular dermis (Rahman et al, 2020). Small or shallow biopsies may hinder accurate tumour differentiation.

Histologically, the tumour mimics morphoeic basal cell carcinomas, microcystic adnexal carcinoma and other adnexal tumours (Mamelak et al, 2010; Rahman et al, 2020). Diagnosis is based on the histological triad originally reported by Brownstein and Shapiro (1977): strands of basaloid cells, keratinous cysts and desmoplastic stroma. Desmoplastic trichoepitheliomas can be differentiated from other adnexal tumours by the lack of loose stroma, the absence of large masses of tumour cells and the lack of ductal differentiation (Mamelak et al, 2010). They also stain positive for CK20 BerEP4, PHLDA1 and CK15 (Rahman et al, 2020).

Desmoplastic trichoepitheliomas predominantly occur in cosmetically sensitive areas and there may be diagnostic uncertainty, both clinically and histologically (Mamelak et al, 2010). Tissue-sparing treatment with Mohs micrographic surgery is recommended to ensure clear margins and reduce the risk of local recurrence (Mamelak et al, 2010). Moynihan et al (2011) argued that experienced dermatologists may be able to clinically diagnose these tumours and manage patients conservatively with observation. This patient was referred for Mohs micrographic surgery and subsequently discharged.

Learning points

- Desmoplastic trichoepitheliomas are uncommon benign cutaneous adnexal tumours predominantly affecting young women, with a predilection for the face.
- Desmoplastic trichoepitheliomas may be difficult to differentiate clinically from morphoeic basal cell carcinomas.
- Diagnosis is achieved with a full thickness skin biopsy.
- Mohs micrographic surgery is the treatment of choice, allowing conservation of skin with adequate surgical margins.

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