Desmoplastic Trichilemmoma

Dr. C. K. Ho

Date:	14 July, 1999
Venue:	Yaumatei Skin Centre
Organizer:	Social Hygiene Service, DH; Clinico-pathological Seminar

CASE SUMMARY

History

A 73-year-old lady presented with a one-year history of a pruritic papule on the chin. This was resistant to treatment with topical steroids and topical antibiotics. There was no significant past medical history or drug history.

Physical examination

There was a slightly keratotic papule on the left cheek measuring 1.5cm in diameter with a central punctum on the left angle of the mouth (Figure 1). There was no other associated lesion. No regional lymph node was palpable.

Differential diagnosis

The differential diagnoses included benign adnexal tumour, keratoacanthoma, squamous cell carcinoma, and basal cell carcinoma.

Investigations

An excisional skin biopsy was performed which showed a well-circumscribed epithelial proliferation at the epidermis surrounded by a thick basement membrane. There was a distinct cell membrane around the tumour cells with a moderate amount of glycogen and peripheral palisading. Occasional mitosis and cellular fibrous stroma were seen. Excision was complete. The histopathology was consistent with desmoplastic trichilemmoma (Figure 2). Positive CD34 expression confirmed outer root sheath differentiation of tumour cells (Figure 3).

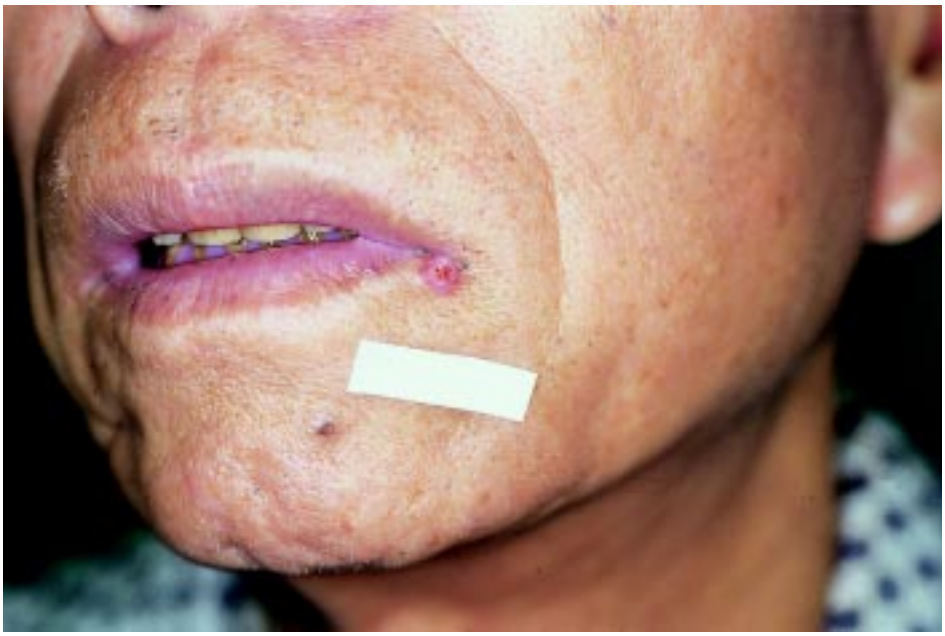

Figure 1: A slightly keratotic papule with a central punctum on the left angle of the mouth

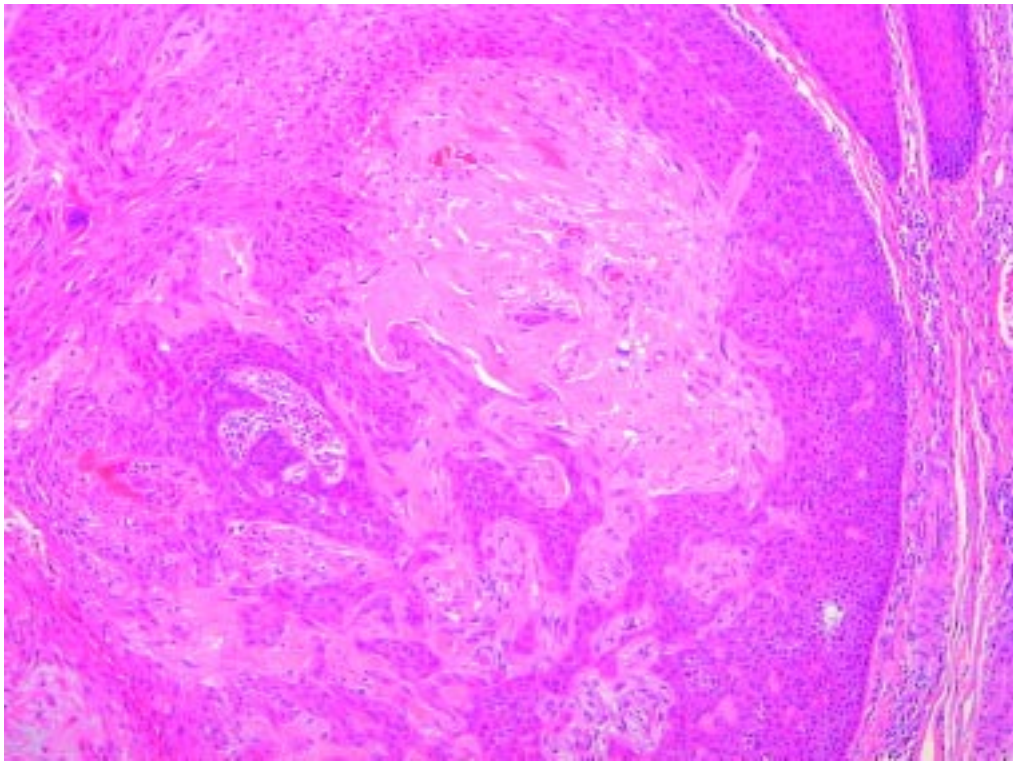

Figure 2: Circumscribed tumour with peripheral palisading and focally thickened basement membrane (right lower field). In the left, there are sclerosis and strangulated epithelial islands. (By courtesy of Dr. K. C. Lee, Department of Pathology, QEH)

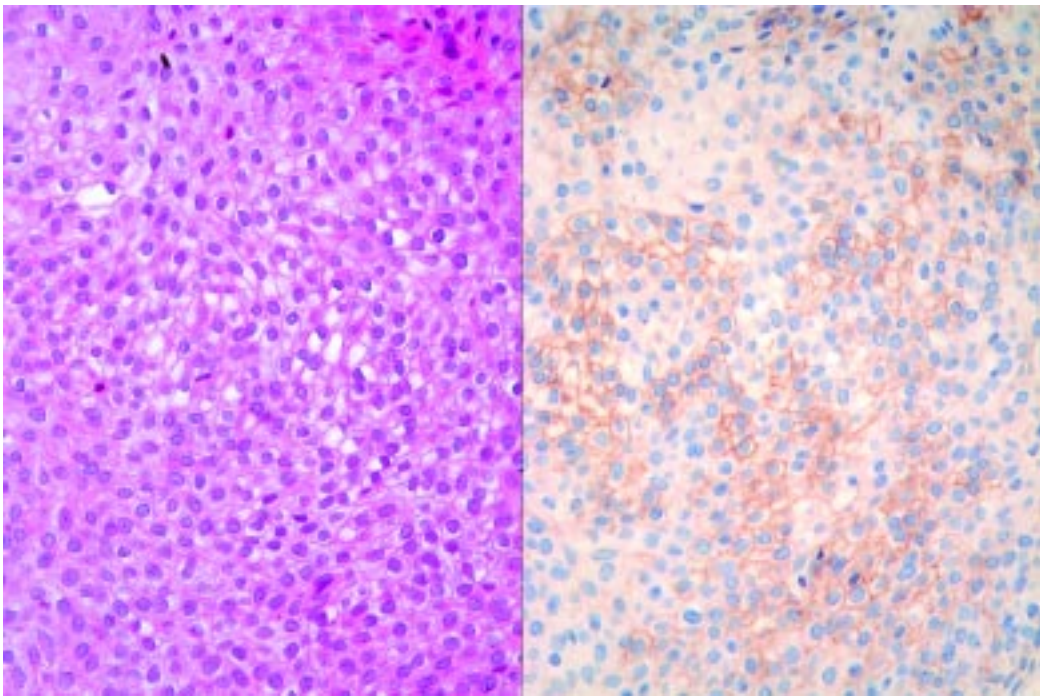

Figure 3: High power view showing clear polygonal cells with sharp border (left) and CD34 expression (right) confirming outer root sheath differentiation. (By courtesy of Dr. K. C. Lee, Department of Pathology, QEH)

Diagnosis

The diagnosis of desmoplastic trichilemmoma was made.

Follow up

There was no recurrence of the excised lesion.

REVIEW ON DESMOPLASTIC TRICHILEMMOMA (DT)

Definition

Trichilemmoma is a benign adnexal tumour of the outer sheath of the pilosebaceous follicle. It is characterised by a superficial lobular proliferation of glycogen-rich clear cells and peripheral palisading surrounded by a thick basement membrane. This may occur as a solitary lesion or in a generalized form (Cowden's syndrome) which consists of multiple trichilemmomas occurring on the face. Patients with Cowden's syndrome are often in the third or fourth decade. It is inherited as an autosomal dominant trait. There are also benign keratoses of the extremities, fibromas and papillomas of the oral mucosa (cobblestones on the lip). More importantly, this may be associated with benign and malignant tumours of the stomach, gastrointestinal tract, thyroid, ovaries and uterus (carcinoma of the breast in up to 29% of women).

There are four less common histological variants: (1) keratinizing trichilemmoma, with central keratinization and extensive squamous eddy formation; (2) an intermediate form between the keratinizing and typical forms, (3) malignant trichilemmoma which has invasive and metastasizing ability and (4) desmoplastic trichilemmoma which is the most recently described variant of trichilemmoma.

Desmoplastic trichilemmoma

Histology

This is characterised by a superficial lobular growth with central desmoplastic stroma. Typical features of trichilemmoma are present at the periphery while at the centre there is a random pattern of cords and nests of basaloid cells with peripheral palisding in a dense stroma. This may be mistaken for invasive carcinoma especially if only the centre is biopsied or when the tumour is highly asymmetric. In addition, although nuclear atypia is usually not present, individual dyskeratosis or cell necrosis may be seen, further increasing the difficulty in diagnosis. The distinguishing features of DT and basal cell carcinoma (BCC) are shown in table 1. In these cases, the presence of a hyaline membrane around the tumour lobules is diagnostic. The cellular stroma typically contains abundant eosinophilic PAS and Alcian blue positive material. There may also be acanthosis of the overlying epidermis. In a study of

Table 1. Contrasting features of desmoplastic trichilemmoma and basal cell carcinoma

	Desmoplastic trichilemmoma	Basal cell carcinoma
Histological features	· Typical features of trichilemmoma at the periphery · Loss of the lobular pattern and desmoplastic stromal changes at the centre · Irregular cords of tumour cells with basophilic and irregular cytoplasm · Occasional dyskeratosis and cell necrosis	· Atypical basaloid cells with peripheral palisading · Mitotic figures · Neoplastic cell with hyperchromatic nuclei
Clinical features	· Smooth telangectatic papule · Pearly border occasionally present · Occasionally ulcerated · Lip, eyebrow, nose · No recurrence reported · Benign lesion	· Nodular BCC most common · Asymptomatic translucent pearly nodule with telangectasia · Occasional ulceration · Lip, eyebrows, nose, ears · May recur after excision · Malignant

seven cases of DT, the problem of misdiagnosis can be seen: three had been previously diagnosed as desmoplastic basal cell carcinoma with features of trichilemmoma, two as malignant trichilemmoma and trichilemmoma with hyaline stromal changes.[2]

The presence of a perilobular hyaline mantle also enables DT to be differentiated from BCC and malignant trichilemmoma. In malignant trichilemmoma, there is a reverse pattern in which the invasive lobules are located at the periphery while the centre consists of well-differentiated trichilemmoma cells with peripheral palisading and a hyaline membrane. In a study of seven cases of DT, immunohistochemical analysis of the stromal cells was negative for epithelial antigen (EMA), carcinoembryonic antigen, and HPV.[1] Only vimentin was positive, suggesting that the stromal cells are of mesenchymal origin rather than of epithelial origin. In another study of 22 cases of DT, the proportion of desmoplasia varied between 20 and 60% of the lesion and was closely associated with ulceration.[2] Human papilloma virus sequences were detected by polymerase chain reaction in typical trichilemmoma but these were not specified to be DT.[3]

The histogenesis of the desmoplastic reaction is unknown. Suggested mechanisms include degeneration of the epithelial lobule or a mesenchymal response to ulceration. It has also been suggested that desmoplasia is due to soluble factors secreted by DT cells

Clinical features

DT is an uncommon lesion and is most often found in men over 50 years of age. It has a non-specific appearance and as it may present as a smooth, asymptomatic, telangiectatic papule, it is often diagnosed as a BCC. It may also have a keratotic surface and may be misdiagnosed as a papilloma. It is most often found on the lip, eyebrow, and nose, that is, similar distribution to a BCC. Lesions vary between 0.5 and 1. 0cm in diameter and may be present for 6 months to 6 years before diagnosis. No recurrence had been reported after excision.

Differential diagnosis

The differential diagnoses includes papilloma, BCC, other benign adnexal tumours and plane warts.

Treatment

Surgical excision is the most effective treatment.

Learning points:

Desmoplastic trichilemmoma is an uncommon benign lesion which may be misdiagnosed as basal cell carcinoma both clinically and histologically.

References

1. Tellechea O, Reis JP, Poiares Bapitista A. Desmoplastic Trichillemmoma. Am J Dermatopathol 1992;14(2):104-14.
2. Hunt SJ, Kilzer PB, Santa Cruz D. Desmoplastic Trichilemmoma: histologic variant resembling invasive carcinoma. J Cutan Pathol 1990;17:45-52.
3. Rohwedder A, Keminer O, Hendricks C, Schaller J. Detection of HPV DNA in trichilemmomas by polymerase chain reaction. J Med Virol 1997; 51(2):119-25.