European Journal of Ophthalmology / Vol. 14 no. 6, 2004 / pp. 562-564

SHORT COMMUNICATION

Case report

Desmoplastic trichilemmoma: a rare tumor of the eyelid

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PURPOSE. *To report an upper eyelid mass which proved to be a desmoplastic trichilemmoma.*
METHODS. *A 60-year-old man had a slowly enlarging upper eyelid mass. The tumor was excised. The pathologic evaluation of the tumor was centered on the differential diagnosis.*
RESULTS. *The clinical appearance of this lesion is nonspecific and can simulate a verruca, follicular keratosis, or basal cell carcinoma. Central desmoplasia, outer root sheath differentiation of the tumor cells, and CD34 positivity are the main characteristics that allow differentiation from basal cell carcinoma.*
CONCLUSIONS. *Proper recognition of a benign neoplasm that may be misdiagnosed as basal cell cancer can prevent aggressive surgical treatment. (Eur J Ophthalmol 2004; 14: 562-4)*

KEY WORDS. *Trichilemmoma, Desmoplasia, Eyelid tumor, Basal cell cancer*

Accepted: June 14, 2004

INTRODUCTION

Desmoplastic trichilemmoma is a benign neoplasm of the hair follicle that is derived from the outer root sheath or infundibular epithelium. It was first recognized as "a special variant of trichilemmoma" by Hunt et al (1).

It usually presents as a solitary nodular lesion of the face in patients over the fifth decade (1-3). The sites of predilection are the lip, eyebrow, and nose (1). There is a recent report concerning a case located on the upper eyelid (4). The importance of this lesion is that it can be misdiagnosed both clinically and pathologically. The most common clinical misdiagnoses are papilloma and basal cell carcinoma (BCC) (2). The characteristic yet worrisome histopathologic feature for the inexperienced pathologist is the desmoplastic stromal reaction, which closely mimics an infiltrating squamous cell carcinoma or a morphea-like BCC (1, 3, 5). A failure to properly make this distinction can result in excessive treatment. Because the eyelid is one of the locations where this tumor can present, ophthalmologists should bear in mind this possibility in order to avoid unnecessary treatment and consequent functional impairment.

Case report

A 60-year-old man with a slowly enlarging and painless mass on his left upper eyelid referred to our clinic. He first noticed the lesion 10 months ago. Clinically the lesion was a small nodular tumor of 11 mm of greatest dimensions, located in the middle of left upper eyelid. A central small scale-crust was noticed on the surface (Fig. 1). The clinical diagnosis was papilloma.

A complete excision under local anesthesia was performed. During the operation it was noticed that the tumor underlying a slightly thickened skin was well circumscribed, pink-gray, and soft.

Histopathologic examination of the specimen showed

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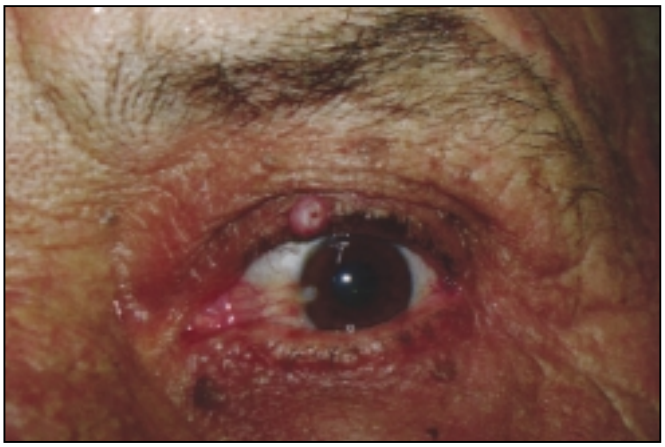

Fig. 1 - *The clinical appearance of the lesion.*

a tumor in close contact with surface epidermis, growing toward the dermis to form a nodule (Fig. 2). The tumoral cells closely resembled the outer root sheath cells of a hair follicle, with clear cytoplasm and eccentric round uniform nuclei. There was a clear palisadic arrangement of the cells at the periphery of the lobules (Fig. 2). The second most striking feature was the dimorphic appearance of the tumor. The area composed of well defined lobules with peripheral palisading merged gradually with areas where tumoral cells were embedded in a sclerotic, desmoplastic stroma (Fig. 3). The lobular architecture was lost in these areas. Immunohistochemical examination with anti-CD34 (QBEND 10) antibody showed membranous staining of the tumoral cells in the lobular area, where palisading was prominent, while tumoral cells in the desmoplastic area remained negative (Fig. 4).

The patient has been followed up for 10 months, weekly during the first month and then with monthly intervals. The patient had no symptoms and the operation area was stable without any sign of recurrence. Follow-up continues.

DISCUSSION

Desmoplastic trichilemmoma is a benign skin tumor with features of outer root sheath differentiation. It is a very infrequent lesion, representing only 7 cases among 21,198 consecutive skin biopsies as reported by Tellechea et al (2). Twenty-eight cases were found among 120,000 accessioned skin biopsies over an 18-month period by Crowson and Magro (6). It is typical-

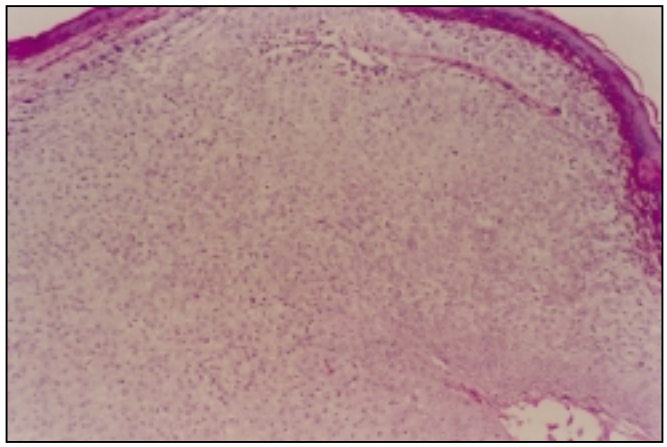

Fig. 2 - *The lobular part of the tumor covered with the thin epidermis on the surface. Tumoral cells exhibit external root sheath differentiation with eccentric uniform nuclei, vacuolated cytoplasm. There is a clear palisadic arrangement at the periphery of the lobulus (hematoxylin and eosin x125).*

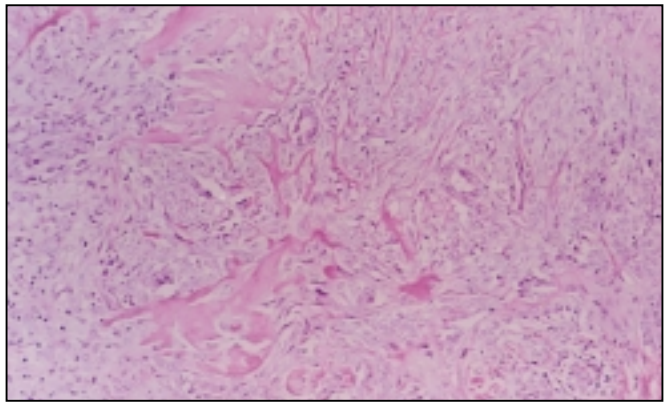

Fig. 3 - *Area showing the prominent stromal desmoplasia. Tumoral cells arranged in narrow cords are embedded in the hyalinized, dense fibrous stroma (hematoxylin and eosin x125).*

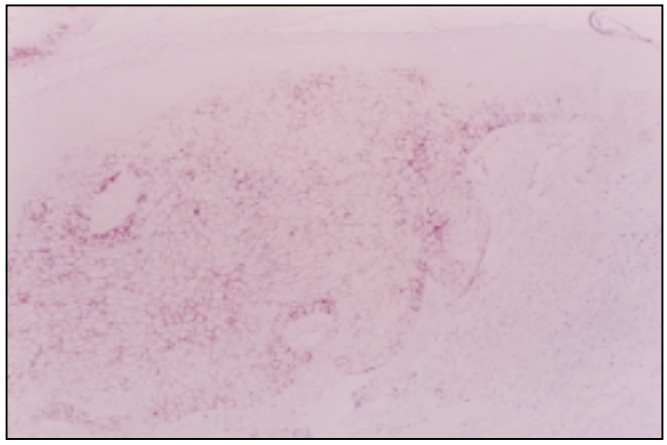

Fig. 4 - *Membranous positive reactivity of tumoral cells in the lobular area of the tumor. Note that the surface epidermis is negative (Anti-CD34 x125).*

ly a slow growing, asymptomatic, solitary dome shaped nodular lesion of 3 to 8 mm in diameter (1). It occurs in middle-aged patients, usually above the fifth decade (1, 2), and seems to be more frequent in men (2). The sites of predilection are the lip, eyebrow, and nose, the head and neck area in general (1, 6). To our knowledge, among the total of 68 desmoplastic trichilemmomas reported in the English literature (1-8), only seven cases were located in the eyelid (1-4). Crowson and Magro reported the largest series with 28 cases and among them, seven were located on the face, with no further topographic specification (6). Hidayat and Font reported a series with 31 cases with lesions located on eyelid and brow (9). Our patient had a very similar presentation to the cases described in the literature in terms of age and macroscopic appearance of the lesion. Histopathologically the tumor consists of lobules of epithelial cells with outer root sheath differentiation which have glycogen rich clear cytoplasm, eccentric nuclei, and peripheral palisading. Generally in the central part of the tumor the epithelial cells are arranged in irregular cords entrapped in a desmoplastic stroma (1-3, 5). This feature, upon which the tumor is named, can mimic the infiltrative pattern of a malignant tumor. The most common misdiagnoses are squamous cell carcinoma and morphea-like BCC (1-3, 5, 9). Illueca et al reported on the usefulness of CD34 immunostaining of epithelial cells in the differentiation from BCC (3). In concordance of this finding, we also demonstrate the positive immunostaining with anti-CD34 (QBEND10) antibody. An interesting feature was that the lobular part showed definite immunostaining while epithelial cells in the desmoplastic part were negative.

Desmoplasia is the most peculiar histopathologic feature of this tumor and different authors have speculated concerning the possible mechanisms which might play a role in this process. Tellechea et al (2) concluded that desmoplasia was a fibroblast-mediated, dendrocyte-independent stromal reaction. More recently Massi and Franchi (5) tested the content of extracellular matrix and found that type I collagen and tenascin were present in the area of desmoplasia. They suggest that probable secretion of tenascin by epithelial cells could result in desmoplasia (5). Hunt et al (1) reviewing a series of 22 patients reported that the area of desmoplasia varied between 20 and 60% of the total tumoral area. Generally occurring in the central part of the tumor, it may sometimes be seen peripherally (1). The desmoplastic area accounted roughly for 70% of the total area and was seen peripherally in our case.

In conclusion, we would like to stress that, although a rare tumor, desmoplastic trichilemmomas could be seen in the eyelid. If the diagnosis is evoked by the ophthalmologist, this could be an important aid to the pathologist who evaluates the specimen to avoid a misdiagnosis that may result in overtreatment

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