

Trichoblastic carcinoma of the pinna. A rare case

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Abstract

Adnexal tumors of the hair follicle can be regarded as lesions that show similar differentiation to one or more portions of the hair follicle. Trichoblastic carcinoma is a rare malignant adnexal tumor, which usually occurs on the scalp. There have been reported cases with regional lymph node metastasis. We report a case of a 65 years old man with a painless irregular mass of the posterior surface of the right pinna, which was slowly growing over a 5-year period. He had a history of a similar lesion on the same site, which was removed 8 years before he presented to us. We excised the lesion and the defect was covered with an advancing flap. Because of the histology result the patient was re-operated and reviewed for over 3 years and there wasn't any sign of recurrence. *Hippokratia* 2007; 11 (2): 89-91

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Trichoblastic carcinoma is a rare adnexal cutaneous tumour which usually occurs on the scalp¹, arising from the pilar sheath epithelium². The tumour develops dermal or subcutaneous fat invasion³ and is occasionally capable of malignant behaviour e.g. metastases^{1,3}.

Clinical history

A 65-year-old male patient presented in the Otolaryngology Outpatient Department with a painless irregular mass of the posterior surface of the right pinna. The mass was slowly growing over a 5 year period causing him moderate itching and bleeding occasionally. There was no lymphadenopathy. The patient reported a history of a similar lesion on the same site 15 years earlier, which was gradually growing and was excised at another Institution 8 years prior to our treatment. No histology result of this excision was available.

The patient underwent complete surgical excision of the lesion to the depth of the auricular cartilage. The skin defect was covered with an advancing flap. Histological examination showed this lesion to be trichoblastic carcinoma with focal central infiltration close to the deep surgical margin while the lateral surgical margins were clear. Because of the histology result the patient was closely reviewed and even though there wasn't any sign of recurrence, a second procedure was carried out 6 months later. The skin of the area was widely excised and the defect was covered with full thickness free skin graft. The new histology was negative for recurrence. The patient is under follow-up for more than 3 years without any sign of recurrence.

Materials and Methods

The resected tumour was fixed in formaldehyde, embedded in paraffin, cut at 2µm, and stained with hematoxylin-eosin.

Results

Histological examination revealed a 2.5x2x1cm circumscribed neoplastic nodule with polylobulated appearance having central cystic and peripheral solid areas. Morphology of the nodule resembled that of malignant epithelial neoplasms related with the basal layer of epidermis, and consisted of micronodular projections of basaloid neoplastic cells without peripheral palisading, while in other areas it demonstrated solid nests of squamous type neoplastic cells or adenoid structures of neoplastic cells without central keratinization. The micronodular masses of neoplasm included bulbar structures resembling the bulbar parts of the hair follicles without trichogenesis. The neoplastic cells were of squamous type with abundant eosinophilic cytoplasm and hyperchromatic and occasionally pleomorphic nuclei with atypical mitotic figures (Figures 1 & 2).

Discussion

Many authors reviewing series of pilar tumours report as the commonest location of the tumor the scalp (90%), the torso and rarely the extremities. It appears more frequently in elderly female and middle age male. There is evidence that pilar tumours may arise from the

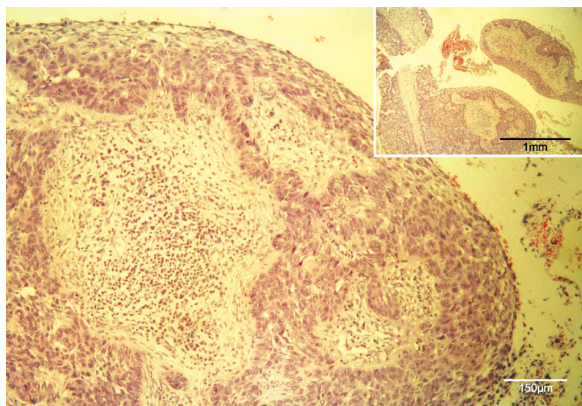


Figure 1. Histology of the dermal tumor nodule. Cystic wall of the tumor nodule with lobular and trabecular structures of squamous epithelial cells. Central cystic and peripheral solid areas (inset). Hematoxylin-eosin stain x128, inset x21

walls of a pre-existing trichilemmal cyst⁴.

Trichoblastic carcinoma is a malignant epithelial adnexal neoplasm arising from the external root sheath of the hair follicle⁵. Its morphological resemblance to trichilemmal neoplasms (e.g. tumors with trichilemmal keratinization)⁵ is responsible for numerous different terms used in the literature⁴ and renders the differential diagnosis from malignant epithelial tumours of the skin difficult, especially from the squamous cell carcinoma^{5,6}. One should be very careful when evaluating the histopathological findings due to the fact that the neoplastic cells of trichoblastic carcinoma express figures of basal cell type as in basal cell carcinoma, without the characteristic peripheral palisading and clefting between tumor and stroma. The principal microscopic criteria for the differential diagnosis between trichoblastic carcinoma and basal cell carcinoma are proposed in Table 1. Furthermore the neoplastic cells demonstrate features of squamous cells without

intercellular bridges formation and without central keratinization. Adnexal differentiation of neoplastic cells, their disposition around a hyalin stroma or bulbar differentiation create problems in differential diagnosis from other histologic types of malignant tumours arising from the hair follicles as basal cell carcinoma, trichilemmal carcinoma and malignant pilomatricoma. In the trichilemmal carcinoma, the neoplastic cells are clear, polygonal ones, most often glycogen-containing with peripheral palisading of cylindric cells and demonstrate keratinization of the isthmic trichilemmal type (as in pilar cysts). In the malignant pilomatricoma, the cells are small basophilic ones; they have keratinization with differentiation of “ghost” cells. The microscopic differential diagnosis from sebaceous carcinoma, clear cell hidradenocarcinoma, and cutaneous metastasis of renal carcinoma is quite easy¹.

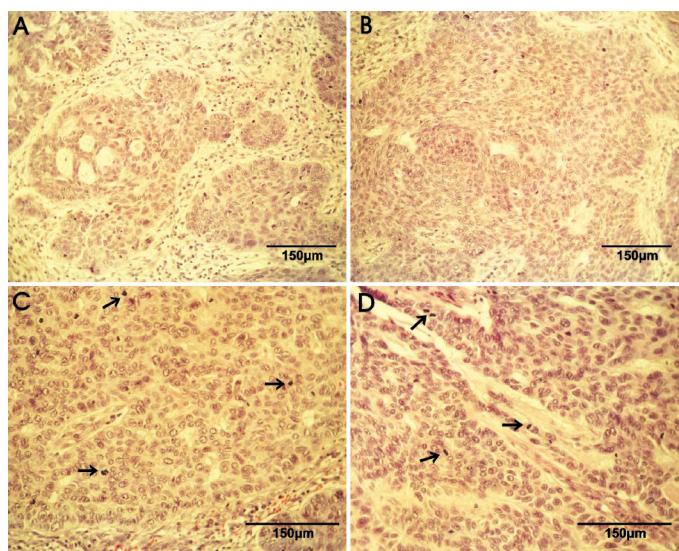


Figure 2. A. Pilar tumor showing lobules and strands of atypical epithelial cells without central keratinization. Hematoxylin-eosin stain x120. B. Lobules of squamous neoplastic cells with atypical features. Hematoxylin-eosin stain x120. C. Neoplastic cells demonstrating nuclear pleomorphism and increased mitotic figures (arrows). Hematoxylin-eosin stain x160. D. Disposition of neoplastic cells around a hyalin stroma. Increased mitotic figures are noticed (arrows). Hematoxylin-eosin stain x160

Table 1: Microscopic criteria for the differential diagnosis between trichoblastic carcinoma and basal cell carcinoma

Trichoblastic carcinoma	Basal cell carcinoma
Lobules of small basal cells without peripheral palisading	Linear palisading of peripheral cells with retraction spaces (clefts) in relation with the fibrous stroma
Focal arrangement of squamous eosinophilic cells without keratinization	Rare foci of squamous metaplasia with keratinization
Bulbar epithelial differentiation without trichogenesis	Lack of organoid adnexal differentiation
Cytonuclear pleomorphism and numerous mitoses	Rare mitoses and pleomorphism except for metatypical forms
Metastatic risk	Exceptional metastatic risk

In the trichoblastic carcinoma, the cells are small as in basal cell carcinoma (which is denominated "malignant trichoblastoma" in the 2nd edition of Ackerman's atlas on "Tumours with follicular differentiation"); they may have an adenoid and/or cystic arrangement (as in the cutaneous counterpart of the adenoid cystic carcinoma), without keratinization ; the foci of large squamous cells are sometimes convoluted but do no more keratinize ; the nuclei are pleomorphic and the mitotic figures increased. The presence of vacuolated glycogen-containing clear cytoplasm in the neoplastic cells is reported as clear cell pilar sheath tumour of the scalp⁷. The architectural pattern of the tumour is cystic, solid or, as in our case, mixed (cystic and solid). Criteria of malignancy in the differential diagnosis from the benign counterpart of the tumour are the presence of cellular and nuclear atypical features, hyperchromatic and polymorphous nuclei, and increased mitotic figures^{3,6}. Trichoblastic carcinoma develops dermal or subcutaneous fat infiltration, while foci of necrosis and/or superficial ulceration of the epidermis may be observed. Immunohistochemical studies demonstrated cytoplasmic expression of cytokeratins. Pilar tumours are completely unreactive (with the exception of sebaceous cells present within or in the vicinity of the tumour or tumours with sebaceous differentiation) with the Thomsen-Friedenreich (T) antigen, a marker that correlates with the prognosis of some carcinomas⁸. In

our case we felt that immunohistochemical studies were not indicated because it is an undifferentiated adnexal tumour without trichogenesis.

Complete surgical excision is considered treatment of choice in order to avoid regional recurrences⁹. Adjuvant radiotherapy have been suggested by some authors³.

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