Proliferating Tricholemmal Cyst Should Always Be Considered as a Low-Grade Carcinoma

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Key Words

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Proliferating tricholemmal cyst (PTC) is an uncommon append-ageal skin neoplasm undergoing outer root sheath differentiation, showing a compact, abrupt tricholemmal keratinization without granular layer interposition. PTC shares this kind of keratinization with the tricholemmal cyst, with the outer root sheath at the level of the isthmus where the inner root sheath disappears, and with an infundibular sac in catagen and telogen hairs [1]. PTC may also present matrical structures with a pattern similar to that detectable in squamous carcinomas with pilar differentiation [2, 3]. Clinically, PTC usually appears as a

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Table 1. Metastasizing malignant PTC reported in detail in the literature

Metastases Differentiation Died of disease

LN = Lymph node; PDC – poorly differentiated carcinoma; WDC = well-differentiated carcinoma; NA = not available.

firm nodule on the scalp of elderly women. Histology shows a dermal lobular proliferation of squamous epithelium, in some areas constituted by clear cells containing glycogen, circumscribed by a glassy and fairly cellular stroma. Lobules show, usually in their central area, tricholemmal keratinization with occasional calcification. Within the squamous proliferation, one can observe dyskeratotic cells, shadow cells and mitosis as well as a palisading arrangement of the nuclei at the periphery of the lobules [1,2].

To the best of our knowledge, 18 cases of malignant PTC have been reported in detail to date [1, 4-14]. Table 1 shows 11 cases, out of 18 malignant PTC, with a metastasizing behaviour [1, 4-10, 14]. Seven cases arose on the scalp, 2 on the head, i.e. ear and cheek, respectively, one on the arm and one in the inguinal region. All cases but 2 showed locoregional metastases, whilst the

inguinal case gave lung, liver and mediastinum metastases and the case who started on the cheek [5] showed generalized dissemination. A patient observed by us [1] in 1985 had regional metastases and many local relapses with a total of 8 surgical excisions and radiotherapy. In 1992, infiltration of the occipital periosteum was present due to a relapse involving the cervical area at the C1 level. In spite of wider demolition and replacement with a muscular-skin flap from the lateral dorsal muscle, the patient – after some cycles of chemotherapy with cis-platinum, a-interferon and vinorelbine – died of locoregional extension of the tumour in the cervical area, 10 years after nodal metastases, without any other distant spread.

A benign and a malignant form of PTC are distinguished. The differential diagnosis between benign and malignant PTC can be made on the basis of mitotic rate, cytological and architectural atypia, necrosis and stromal infiltration [12, 15]. In some cases, atypical areas were admixed to well-differentiated lobules [12, 14]. However, cases with little or no cytological and architectural atypia may have an aggressive behaviour and vice versa [1, 7, 13]. A morphometric analysis did not show differential criteria between benign and malignant PTC, but rather a homogeneous cell population in each sample, concluding that the biological behaviour of PTC is not correlated with its histological appearance [16]. Ploidy has been studied in PTC [13, 15] and non-diploid cells were found in malignant as well as in histologically benign PTC, the proliferation index being increased in non-diploid cases [15].

Concluding, we suggest that PTC should always be considered as a low-grade carcinoma and an accurate follow-up is recommended in

all cases, as its biological behaviour seems to be actually, histologically unpredictable.

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