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REVIEW

Treatment of Malignant Cutaneous Adnexal Neoplasms[☆]

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KEYWORDS

Malignant cutaneous adnexal neoplasms; Treatment; Surgery; Mohs micrographic surgery

PALABRAS CLAVE

Neoplasias anexas cutáneas malignas; Tratamiento; Cirugía; Cirugía de Mohs

Abstract Malignant cutaneous adnexal neoplasms form a group of rare, typically low-grade-malignancy carcinomas with follicular, sebaceous, apocrine, or eccrine differentiation or a combination of the first 3 subtypes. Their clinical presentation is usually unremarkable, and biopsy is required to establish the differentiation subtype and the definitive diagnosis. Due to their rarity, no clear consensus has been reached on which treatment is most effective. Mohs micrographic surgery is considered to be the best option to prevent recurrence in the majority of patients. Radiotherapy and chemotherapy have been studied in very few cases and have rarely been shown to be effective.

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Tratamiento de las neoplasias anexas cutáneas malignas

Resumen Las neoplasias anexas cutáneas malignas constituyen un grupo de carcinomas poco frecuentes, habitualmente de bajo grado de malignidad, que muestran diferenciación folicular, sebácea, apocrina o ecrina o una combinación de las 3 primeras. Clínicamente suelen ser neoplasias con características poco distintivas, siendo necesaria una biopsia que permitirá establecer el tipo de diferenciación y el diagnóstico definitivo. Al tratarse de una enfermedad poco frecuente, no existe un claro consenso sobre el tratamiento más eficaz. En la mayoría de casos se considera la microcirugía de Mohs como la opción más efectiva para prevenir recidivas. La radioterapia y quimioterapia han sido escasamente estudiadas y solo se han mostrado eficaces en escasas ocasiones.

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Introduction

Malignant cutaneous adnexal neoplasms are an uncommon group of low-grade carcinomas. Although most of these tumors have very limited ability to spread to distant sites, they are locally aggressive and must be treated with surgical excision to ensure tumor-free margins. Malignant cutaneous adnexal neoplasms have distinctive histopathologic characteristics, but their clinical characteristics are largely nonspecific. The type of differentiation present in each tumor is recognizable through histopathologic characteristics that resemble certain findings present in the corresponding normal adnexal structures. Generally speaking, as these tumors are malignant, they show few signs of differentiation. Investigation through serial cuts or immunohistochemical staining is therefore necessary to establish the type of differentiation in a given adnexal tumour. Many malignant cutaneous adnexal carcinomas display only ductal differentiation, and as eccrine and apocrine ducts are currently indistinguishable both immunohistochemically and ultrastructurally, all that can be clearly established in such cases is that the tumour is a ductal carcinoma. No further differentiation is possible. Although differentiation is minimal, however, certain histopathologic features, summarized in [Table 1](#), can suggest malignancy. In this article, we review the malignant cutaneous adnexal neoplasms listed in [Table 2](#).

Pilomatrix Carcinoma

Pilomatrix carcinoma presents as a solitary nodule on the upper body in the vast majority of cases.¹ It is more common in middle-aged and elderly men. Most publications to date have described a largely nonaggressive biologic behavior, but there have been reports of metastasis to regional lymph nodes and internal organs with fatal outcomes.²

Histopathologically, pilomatrix carcinoma is seen as an asymmetric, poorly circumscribed tumor with frequent ulceration of the epidermal surface. It consists of a proliferation of immature basaloid cells, reminiscent of follicular matrical cells, that form either solid basaloid islands or cords of cells penetrating the deep dermis, subcutaneous tissue, and even the fascia and muscle ([Fig. 1](#)). The most common finding is the presence of islands of shadow cells indicating matrical differentiation.³

Treatment

Pilomatrix carcinoma is a malignant tumor that frequently shows aggressive local behavior. It invades adjacent structures, and incomplete excision of the primary tumor results in disease persistence in 60% of cases.⁴ Metastasis generally occurs through hematogenous or lymphatic spread and there have been reports of distant metastasis resulting in death.⁵ Most publications recommend surgical excision with margins of between 5 mm and 2 cm.^{6,7} Mohs micrographic surgery (MMS) is a good option as it frequently achieves clear margins. Adjuvant radiation therapy has produced varying results, but has shown no clear improvement in recurrence rates. Intravenous chemotherapy has not proven

Table 1 Histopathologic Differential Diagnosis Between Benign and Malignant Adnexal Neoplasms.

Benign Tumors	Neoplastic Tumors
Symmetric	Asymmetric
Well circumscribed	Poorly circumscribed
Upturned V-shape often present	Upturned V-shape often absent
Frequent vertical orientation	Frequent horizontal orientation
Smooth borders	Serrated borders
Condensed peripheral fibrous tissue	Noncondensed peripheral fibrous tissue
Clefting between the tumor stroma and the adjacent healthy dermis	Clefting between the tumor stroma and the epithelium
Enucleation often easy following incision	Enucleation often difficult following incision
Stroma predominating over epithelium	Epithelium predominating over stroma
Tends to be located in superficial layers	Tends to invade deep layers
No epidermal ulceration	Frequent epidermal ulceration
Tumor islands separated by abundant stroma	Tumor islands separated by scanty stroma
Tumor islands with a relatively uniform shape and size	Tumor islands of varying shapes and sizes
Small individual tumor islands	Sheets of confluent tumor islands
Well differentiated	Poorly differentiated
Conservation of existing adnexal structures	Destruction of existing adnexal structures
General absence of massive necrosis	Massive necrosis common
Non-neoplastic cells in perineural location	Perineural neoplastic cells common
Absence of intravascular neoplastic cells	Occasional presence of intravascular neoplastic cells
Absence of cords of epithelial cells among collagen bundles	Cords of epithelial cells among collagen bundles
Tumor islands tend to become smaller as they penetrate the dermis	Tumor islands do not tend to become smaller as they penetrate the dermis

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