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Eyelid Microcystic Adnexal Carcinoma

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Microcystic adnexal carcinoma is an uncommon cutaneous tumor with multiple synonyms. On cursory microscopic examination, the tumor mimics syringoma and other benign skin adnexal tumors. However, the asymmetric, infiltrative growth pattern clearly sets the lesion apart as carcinoma. The tumor is locally aggressive, with recurrences common, but regional metastases are rare. Histogenesis is controversial. Optimal treatment consists of complete surgical excision with clear surgical margins.

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Microcystic adnexal carcinoma (MAC) is an uncommon tumor known by a variety of names, including combined adnexal tumor of the skin,[1] sclerosing sweat duct carcinoma,[2,3] microcystic carcinoma,[4] sweat gland carcinoma with syringomatous features,[5,6] and malignant syringoma.[7] It is a locally aggressive lesion with a strong tendency for local recurrence if incompletely excised. Histologically, MAC can be confused with benign skin adnexal neoplasms. The face is the most common site.[8] Glatt et al[7] and LeBoit and Sexton[9] each described two periocular cases. We treated one patient with an upper-eyelid–eyebrow lesion.

REPORT OF A CASE

A 35-year-old woman had a 0.2-cm nonulcerated cutaneous nodule with a 1-cm circumferential zone of induration involving her right upper eyelid and eyebrow. She did not have excessively sun-damaged skin for her age, and she had not had facial radiotherapy. Punch biopsy disclosed a sweat gland neoplasm, not otherwise specified.

Complete surgical excision with the Mohs micrographic technique left a substantial defect (**Figure 1**). The defect was closed by a combination of Z-plasties and rhomboid flaps (**Figure 2**). The initial cosmetic result was acceptable (**Figure 3**). Eyebrow tattooing and scar dermabrasion followed, with good effect. There has been no recurrence after 2 years.

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PATHOLOGIC FINDINGS

The surgical excision of skin and subcutaneous tissue displayed an asymmetric, poorly circumscribed tumor that was broader than it was deep. There was no continuity of the lesion with the surface epithelium. Tumor extended into the deeper dermis, subcutaneous tissues, and muscle (**Figure 4**).

The tumor was composed of small epithelial cells. In the superficial portion of the lesion, small keratocysts were noted (**Figure 5**). At middermal levels, microtubules and thin trabeculae predominated. There was focal invasion of the perineural space (**Figure 6**). The epithelial constituents in the dermis and subcutis were surrounded by abundant, dense, hyalinized stroma. Mitoses and necrosis were not features of the tumor.

A battery of immunostains demonstrated that the tumor cells were positive with antibodies to high- and low-molecular weight keratin, and negative with antibodies for epithelial membrane antigen and S100 protein. The tumor cells and intraluminal contents did not react with antibodies to carcinoembryonic an-

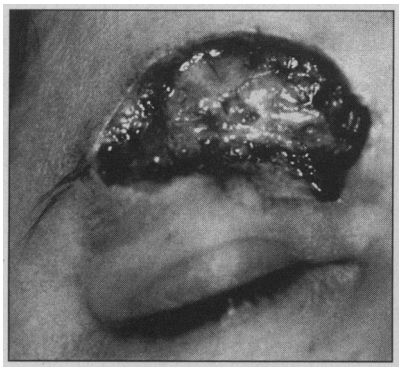

Figure 1. *Initial defect after Mohs surgical excision.*

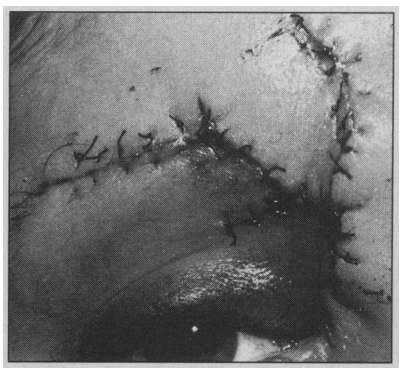

Figure 2. *Appearance after closure by Z-plasties and rhomboid flaps.*

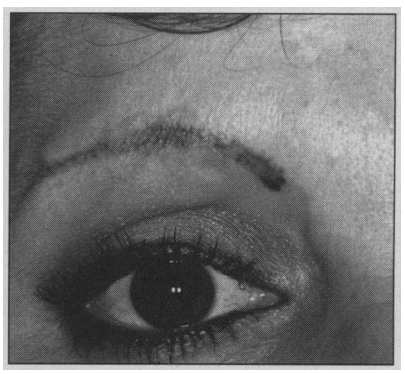

Figure 3. *Final appearance 1 year postoperatively.*

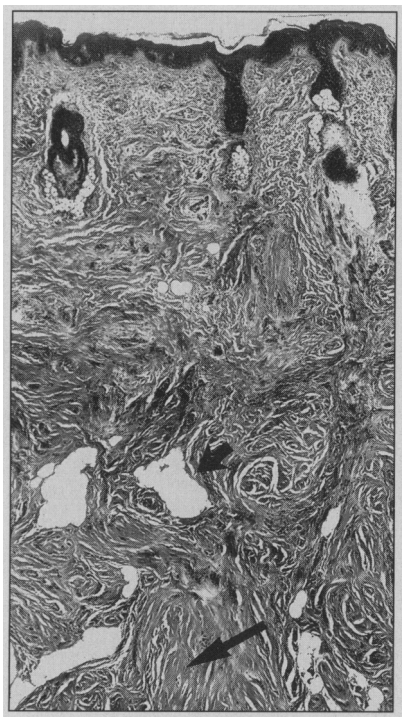

Figure 4. *Overview of the tumor displaying absence of epidermal involvement. There is deep subcutaneous extension. The subcutaneous fat (short arrow) is partitioned by bands of dense hyalinized tumor-containing stroma (long arrow) (hematoxylin-eosin, original magnification ×25).*

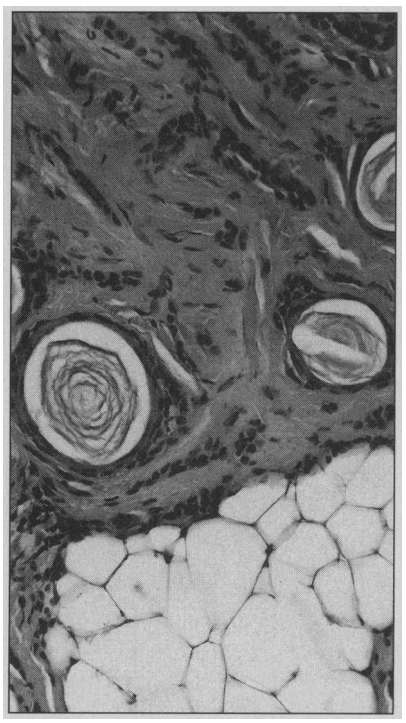

Figure 5. *Small keratocysts in the upper portion of the lesion (hematoxylin-eosin, original magnification ×400).*

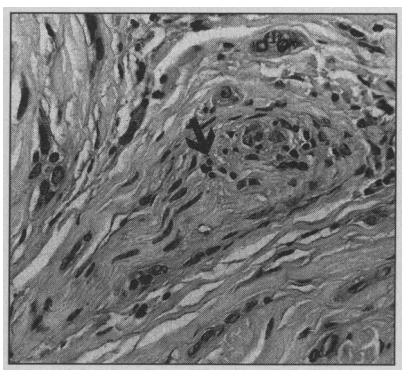

Figure 6. *Tumor invading the perineural space (arrow) (hematoxylin-eosin, original magnification ×400).*

tigen on paraffin-embedded sections. Intact, uninvolved eccrine elements served as a striking built-in positive control.

COMMENT

Microcystic adnexal carcinoma is a relatively uncommon skin appendage tumor. Gender distribution is equal. Age of the patients ranges from 18 to 76 years.[2] The face was involved in 31 of 36 cases in one major series.[8] The case reported herein represents the fifth one reported to occur in a periocular location.[7,9] Growth is indolent, and the time ranges from 1 to 17 years.[4] Lupton and McMarlin[10] noted that two cases in the literature occurred after radiation therapy for acne. Lober and Larbig[11] reported a case of MAC in a 48-year-old man who had received thymic irradiation in childhood and radiation after removal of a thyroid carcinoma at 35 years of age.

The lesion is usually solitary and occurs as a nodule or an indurated, deep-seated plaque. The size ranges from 1 to 3 cm. On superficial microscopic exanimation, the appearance mimics that of several benign skin appendage tumors. Benign skin appendage tumors tend to be symmetric and deeper than they are broad. Like most malignant skin appendage tumors, the silhouette of MAC is asymmetric and broader than it is deep. The MAC infiltrates the dermis, subcutaneous fat, and underlying tissues. Perineural space invasion was noted in 80% of cases in one series.[5] Cellular atypia, abnormal mitoses, and necrosis are not features of MAC. The epithelial elements are set in abundant hyalinized sclerotic stroma.

In one series,[9] nine of 17 lesions were initially misdiagnosed because of the small size of the biopsy specimen. In general, the most common misdiagnosis is syringoma. Clinically, syringomas are usually small and multiple. Microscopically, they are symmetric and do not display perineural invasion.

The histogenesis of MAC is disputed. To some investigators, the

presence of small keratocysts in the upper portion of the lesion implies pilar differentiation, and the presence of microtubules at deeper levels implies eccrine differentiation. These investigators postulate origin from pluripotential adnexal keratinocytes, capable of differentiating toward pilar and eccrine duct structures to a variable degree within a given tumor. Most of these investigators designate the tumor as MAC.[1,9,11-14]

Alternatively, there are those who consider that the small keratocysts, microtubules, and solid strands of tumor are all within the spectrum of eccrine neoplasms. For this group, the keratocysts are likened to the acrosyringium of the normal eccrine apparatus. These investigators prefer to designate the tumor as sclerosing sweat duct carcinoma.[3-5,8] Lipper and Peiper[6] reported that by electron microscopy, the tumor corresponds to benign syringoma and normal eccrine ducts.

Inmunohistochemistry has not been definitive in settling the question of histogenesis. Some have used carcinoembryonic antigen expression by tumor cells as evidence of eccrine histogenesis.[3,15,16] Others consider the absence of carcinoembryonic antigen expression in the areas of keratin microcysts as evidence of pilar differentiation.[12] In the case under discussion, all elements of the tumor failed to express carcinoembryonic antigen. The data from an immunohistochemical study of MAC by Wick and colleagues[17] are consistent with sudoriferous and partial pilar differentiation. Because we support the pluripotential adnexal keratinocytic concept, we have designated our tumor as MAC.

In the largest series,[8] one or more local recurrences occurred in 47% of cases within 2 to 29 years after initial therapy. Lupton and McMarlin[10] reported a recurrence after a 30-year interval, emphasizing the indolent biologic behavior of MAC.

Treatment requires complete surgical excision. Tumor-free margins in the original specimen portend a favorable prognosis.[5] One reported tumor involved an underlying lymph node, probably by direct extension.[5] The risk for regional lymph node metastasis is apparently low.

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