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## Case Presentation

### Microcystic Adnexal Carcinoma of the scalp in an african-american male

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## Abstract

Microcystic adnexal carcinoma (MAC) is a rare adnexal neoplasm that typically presents in Caucasians. We report a rare case of MAC in a 68 year old African American male that presented as a large asymptomatic scalp mass. The clinical and histologic features of MAC are discussed. A summary of all reported cases of MAC in African American patients is presented, and treatment options are discussed.

**Keywords: Dermatology; Skin Cancer; Microcystic Adnexal Carcinoma**

## Introduction

Microcystic adnexal carcinoma (MAC) is a rare, locally aggressive and infiltrative sweat gland neoplasm with occasional hybrid follicular differentiation that is associated with high morbidity. While the majority of MACs occurs in Caucasians [1], to our knowledge, there have been eight prior reported cases of MAC in African American patients. In Caucasian predominant cohorts, MAC has been found primarily in sun-exposed regions of the head and neck of elderly patients [1,2]. Given the paucity of cases reported in African Americans, little is known about potential differences in presentation or tumor behavior in skin of color. This case is presented to increase the body of knowledge of this rare tumor in African American patients.

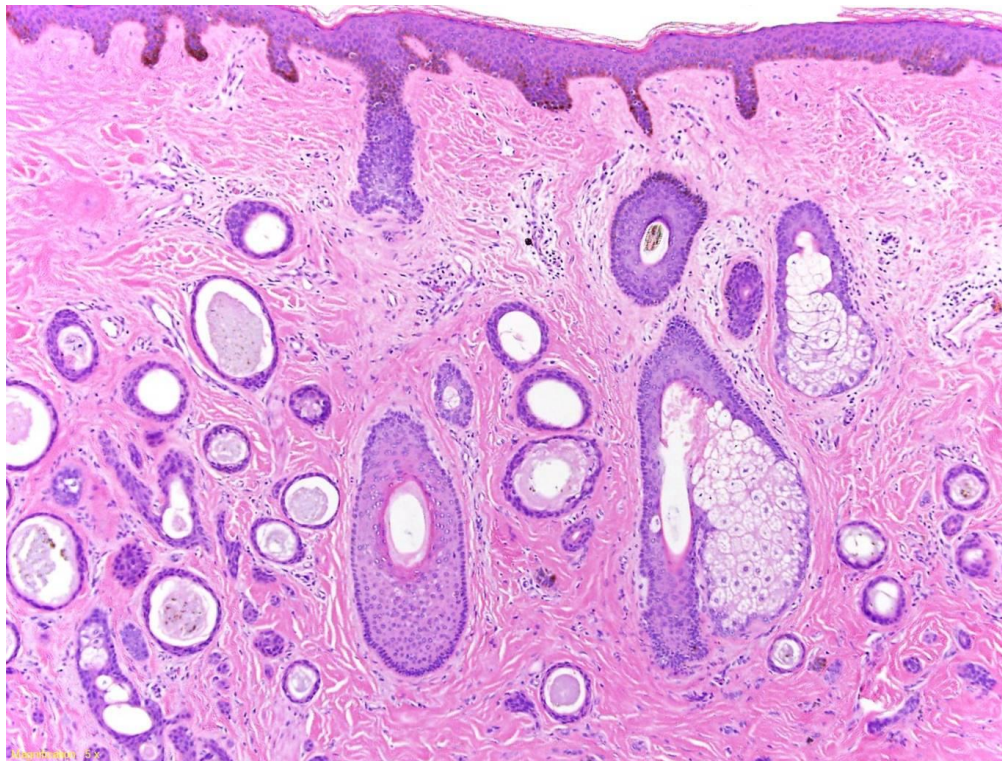
## Case Report

A 69-year-old Fitzpatrick skin type V African American male presented with a multiple year history of a large asymptomatic scalp mass. He denied pruritus, bleeding, paresthesia, or pain at the site. His past medical history was significant for hypertension and a cerebrovascular accident, but he denied any personal or family history of skin cancer, radiation therapy, or immunosuppression. Physical examination revealed an ill-defined 6 x 8 centimeter (cm) hyperpigmented firm plaque with scattered nodularity, erosions, keratin plugs, and a small fluid-filled cystic component (Figure 1).

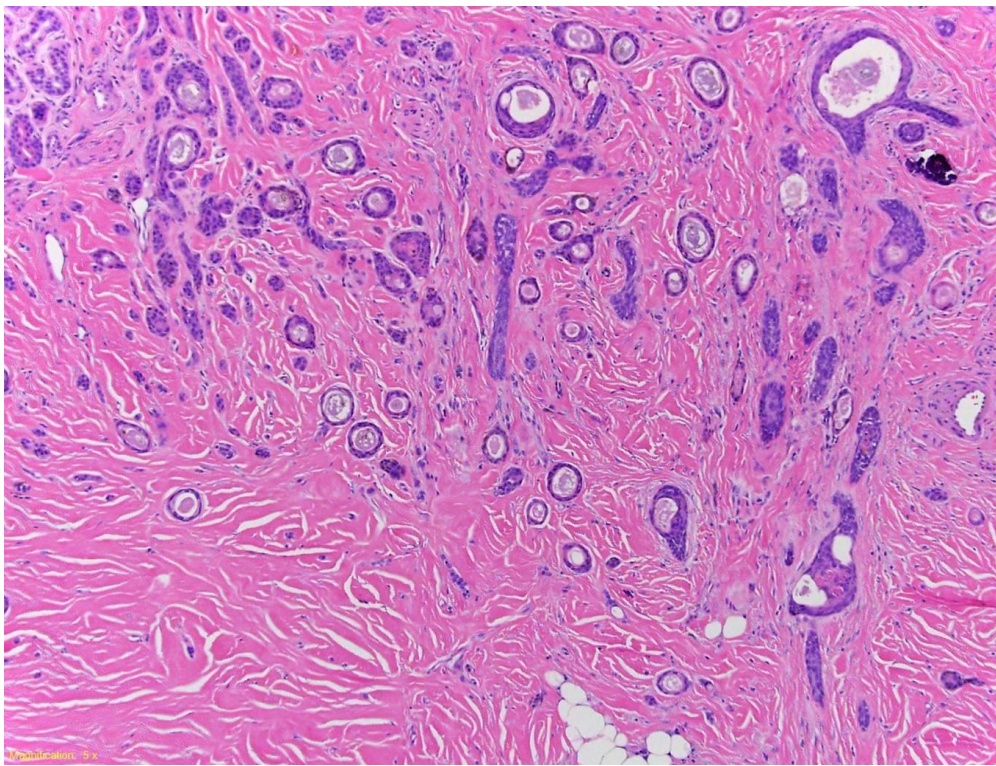


**Figure 1:** Clinical appearance of the lesion at the time of presentation demonstrating a hyperpigmented firm plaque with scattered nodularity and keratin plugs.

No other lesions were found on exam. No palpable lymphadenopathy was appreciated. Two punch biopsy specimens of the scalp were obtained (Figure 2 and Figure 3).



**Figure 2:** H+E stain of punch biopsy demonstrating the dermis and superficial dermis at 10x magnification. Note the round tubular-like structures embedded within fibrous stroma. At this level of the epidermis, the distinction between benign syringoma and MAC is not possible.



**Figure 3:** H+E stain of punch biopsy demonstrating the deep dermis at 10x magnification. Note that the tubules become progressively smaller and form cords as they descend into the dermis representing invasion by a malignant tumor and differentiation from a benign syringoma.

Histopathologic examination revealed a deeply infiltrative dermal tumor with no connection to the epidermis or hair follicles. Numerous small to medium-sized keratocysts were found superficially that merged into smaller cysts. The deep dermis consisted of small solid strands of cells with a highly infiltrative growth pattern and a dense fibrous stroma. A combined S100/PanCK stain revealed focal perineural invasion. These features confirmed the diagnosis of microcystic adnexal carcinoma. A CT scan visualized the soft tissue lesion extending to the calvarium without definite evidence of bony invasion, and a PET scan demonstrated no evidence of metastatic disease.

Because of the size of our patient's lesion and the potential challenge in achieving adequate local anesthesia, wide local excision with two cm margins to the level of the periosteum was performed by otolaryngology under general anesthesia, and the defect was repaired with INTEGRA®. The margins were noted to be negative for carcinoma and the patient continues to be followed closely by dermatology for clinical monitoring of the surgical site.

## Discussion

Microcystic adnexal carcinoma (MAC), also known as sclerosing sweat duct carcinoma (SSDC), is a rare, locally aggressive and deeply infiltrative adnexal neoplasm. While MAC only rarely metastasizes [3], it causes significant morbidity through local invasion of bone, muscle, blood vessels, cartilage, and nerves. Clinically, the lesion's borders are often difficult to discern with unpredictable subclinical extension. In one series of twenty-five cases of MAC, the clinically apparent tumor size averaged 3 cm<sup>2</sup>, while the total defect size after excision measured 18 cm<sup>2</sup> [2]. Similarly, Chiller et al. reviewed twenty-two cases of MAC excised with Mohs micrographic surgery and found the defect size after surgical removal to be four times larger than the clinically apparent tumor [4].

Histopathologically, infiltrative cords, squamous or basaloid nests, and ductal structures are noted in a desmoplastic stroma. These findings can be difficult to differentiate from syringomas and desmoplastic trichoepitheliomas on superficial biopsies [5]. The tumor cells are typically cytologically bland, lack evidence of cell necrosis, and demonstrate rare or absent mitoses. Perineural invasion is a helpful clue in differentiating MACs from more benign proliferations and may present with numbness, burning, or paresthesia [6].

In predominately Caucasian cohorts, 60-85% of MACs occur in the head and neck at a median age of onset of sixty-eight years old [1,2,7]; however, pediatric and congenital cases have also been reported [8,9]. Like most cutaneous neoplasms, MAC is more

common in fair skinned individuals with around 90% of MACs occurring in Caucasian patients [1]. Although lesions are not strictly restricted to sun-exposed sites, its predilection for the head and neck regions of fair skinned individuals suggests ultraviolet (UV) exposure may be a risk factor. MAC has also been known to arise in areas of previous radiation therapy [10].

There have been few reports of MAC in skin of color; hence, little is known about potential differences in clinical presentation, patient epidemiology, or tumor aggressiveness in patients of varying skin types. To our knowledge, there have been only eight reported cases of MAC in African American patients prior to this case. A brief summary of each of these cases is presented in Table 1. The median age at presentation in this small cohort is 43 years old, in contrast to the median age of 68 reported in Caucasian-predominant patients. Seven of the 9 cases (78%) were on the head and neck, which is similar to the distribution reported in non-African Americans. One case was on a non-sun-exposed region (vulva), and one case had a mix of sun-exposed and non-exposed regions.

**Table 1 - Microcystic Adnexal Carcinoma in African American Patients**

Year	Author	Patient Age & Gender	Location of lesion	Clinical Size	Post-op Defect Size
1991	Buhl et al [11]	43 year old female	Vulva	1.5 x 2.0 cm,	Vulvectomy
1998	Park et al [12]	31 year old male	Vertex of scalp	1.1 x 1.2 cm	3.0 x 4.2 cm
2001	Peterson et al [13]	64 year old male	Lip philtrum	1.5 cm	Size not listed
2001	Gardner et al [14]	52 year old female	Posterior scalp	8.0 x 7.0 cm	12.2 x 11.1 cm
2007	Nadiminti et al [15]	54 year old female	Right eyebrow	Size not listed	7.2 x 7.0 cm
			Left axilla	1.0 x 1.0 cm	Size not listed
2007	Nadiminti et al [15]	26 year old male	Scalp	Size not listed	2.2 x 1.5 cm
2007	Page et al [16]	34 year old male	21 sites on hands, arms, back, and legs. No lesions on face.	Multiple	Multiple
2008	Nelson et al [9]	6 year old female	Left preauricular	Size not listed	Size not listed
2016	This case	68 year old male	Vertex of scalp	6.0 x 8.0 cm	10.0 x 12.0 cm

Mohs micrographic surgery is considered the first-line management option [2]. With proper margin control, tumor recurrence rates are between 0-2% following excision via Mohs surgery [4,17]. In contrast, margins are positive in over half of cases following standard excision and therefore require multiple excisions (and several office visits) to clear subclinical spread [4,18]. Occasionally, large tumor size may necessitate wide excision under general anesthesia [2,19]. Adjuvant radiotherapy appears effective at decreasing the risk of recurrence in patients treated with wide excision. In one case-series of patients with advanced MAC (56% with perineural invasion) treated with wide local excision and post-operative adjuvant radiotherapy, 12/14 patients remained free of disease with a median follow-up time of five years while one patient had local recurrence and one had nodal recurrence [18]. Overall, the data is limited regarding the role of radiotherapy in the management of MAC.

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