Case Report of 74-Year-Old Female with Microcystic Adnexal Carcinoma, a Rare and Locally Invasive Neoplasm, with Multiple Recurrences and Review of the Literature

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Abstract

Microcystic adnexal carcinomas (MAC) are a very rare form of locally invading neoplasms. This paper reports a case of a 74-year-old female diagnosed with a MAC neoplasm localized to the left upper cheek; she was treated with both surgery and radiotherapy. The patient had two recurrences after surgical excisions with the second excision being coupled with radiotherapy. This report assesses the rarity of this disease, which may lead to misdiagnosis, and the optimal treatment to minimize the possibility of known high recurrence rates. MAC is not considered a highly metastatic neoplasm; however, potential for metastasis exists, as discussed in this case.

ABBREVIATIONS

MAC: Microcystic Adnexal Carcinoma; BAP: Black/Asian/Pacific-Islander; Other: All Races Not Encompassed Under White or BAP; SEER: Surveillance, Epidemiology, and End Results; MMS: Mohs Micrographic Surgery

INTRODUCTION

Microcystic adnexal carcinoma (MAC), also known as sclerosing sweat duct carcinoma, malignant syringoma, syringoid carcinoma, and trichofolliculoma with perineural invasion, is a rare type of skin cancer. It typically presents with aggressive local invasion of eccrine sweat glands and exhibits small keratinizing cysts, apocrine differentiation, and well-defined ducts [1]. They are considered to have keratinocytic origins, thanks to in-vitro studies, but this still remains a question of further investigation [2]. Sweat gland malignancies are considered to be rare and represent solely 0.005% of all malignant epithelial neoplasms [2]. 85% of MAC cases present in the head and neck region, exhibiting preference for the per orbital skin and centro facial region (with an inclination towards the left side of the face)[2,1]. It is commonly presented in women in their sixth and seventh decade of life [3]. MACs are capable of infiltrating muscle tissues and peripheral nerve fibers; typical treatment consists of surgery since radiotherapy results in increased probability of recurrence [1]. Due to its rarity, this cancer is commonly misdiagnosed; Rustemeyer et al., described a case of MAC as desmoplastic trichoepithelioma for over four years due to the indistinguishable features [3]. Recurrence has been reported to occur even after a period of 30 years [4].

CASE PRESENTATION

We report a case of MAC in a 74-year-old Caucasian woman; she presented with a slow growing, crusty, painless, plaque like 2 cm lesion on the left upper cheek. A small biopsy was obtained and pathological evaluation showed nests and cysts of cytologically uniform, squamoid cells randomly dispersed in a dense edematous collagenous stroma (Figure 1). In some areas, branching of tubular structures and aggregation of neoplastic cells gave commalike shapes. The microcysts contained lamellated keratinaceous-like material resembling hair matrix, and some areas demonstrated extensive squamous metaplasia. Despite MAC was suspected, the limited biopsy sample did not allow definitive pathological diagnosis and additional sampling was requested for further investigation. The diagnosis was consistent with a sclerosing adnexal neoplasm, the differential diagnosis included: desmoplastic trichoepithelioma, sclerosing or morpheaform basal cell carcinoma, and syringoma, rare form of basosquamous or even squamous cell carcinoma, and epithelial-myoepithelial carcinoma. An excisional biopsy was...
performed and the 2 cm mass was removed, which showed a depth of 8mm from the skin surface; it was 1mm from the deep margin. Pathological evaluation of the excised mass showed all features noted in the initial biopsy, but in addition, clear cellular and architectural malignant features were noted. Moderately differentiated squamous cell carcinoma in the superficial layers was seen merging with a syringomatous carcinoma focally. Perineural invasion was present. Immunohistochemical analysis was consistent with syringomatous carcinoma (positive for p63, CK5/6, CK14 and questionable positive reaction with MUC1; negative for S100, calponin, actin, and mucin). Seven months later, the patient experienced recurrence at the same site and was surgically removed; nine months later, the patient experienced another local recurrence. She declined any further surgery. Radiotherapy of the involved area was performed, and the patient was lost to follow-up.

**DISCUSSION**

The case presented here highlights common epidemiological similarities within reported MAC patients and shows the rarity of the disease. Literature presents the median age at diagnosis at 67.5 years [5] and the mean age at diagnosis at 61.2 years [1]; the patient presented in this case was 74. MACs are considered a relatively rare condition; Chen et al., concluded that approximately 350 cases are present within the English literature. MACs are more common amongst females, favouring whites over others: 54.6% of cases reported in Surveillance, Epidemiology, and End Results (SEER) were in white female patients with another 36.4% being in white male patients (Table 1 and Figure 2) [5]. The remaining 9% of cases are characterized in Table 1 and Figure 2. A case report was published in 2016 exhibiting MAC in a Chinese patient from the Hainan province of China; this is one of three cases ever reported in the Chinese population [2] as the other 2 cases were reported by Fang et al., in 2009 [6]. Similarly, the first ever reported case of MAC in an African-American man was in 2001 by Peterson et al. [7].

While the disease is more prominent in whites (particularly females), the lesion tends to occur in specific regions; the left side of the face is favoured. Our patient exhibited this as she had a lesion on her left upper cheek; 74% of lesions of the 164 patients in the SEER database analysis presented with head/neck lesions, with the occurrence being highest in white females (Table 2 and Figure 3). White males have the second highest occurrence in the head and neck region (Table 2 and Figure 3). This anatomical distribution pattern is also similarly reported by Chiller et al., in their case series review of 48 case reports of MAC patients, which is considered to be the largest case series review in the English literature [1].

Despite aggressive local invasion, metastasis is not common within MAC patients. The present patient did not have metastasis; however, the patient was lost to follow-up after two local recurrences. SEER database analysis also confirms that MAC metastasis is uncommon as none of the 164 patients exhibited metastasis [5]. Despite the rarity, there have been a few cases of metastasis; one case series review found one case of lung

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**Figure 1** Histopathology of microcystic adnexal carcinoma, skin biopsy of the mass, taken from the left cheek of a 74 year old woman

A: Nests and cysts of squamoid cells scattered in background of dense edematous collagenous stroma, H&E at x20 magnification

B: Tubular structures and aggregates of neoplastic cells, H&E at x50 magnification

C: Lamellated keratinous-like material resembling hair matrix, H&E at x50 magnification

D: Atypical pleomorphic squamous cells, H&E at x100 magnification
metastasis out of seven cases [8] while another case report found lymph node metastasis in a 63-year-old male [9].

Surgery is typically used for treatment; radiotherapy is not as popular due to the potential of recurrence [1]. The patient presented in this case also was surgically treated and radiotherapy was used due to recurrence post-surgery. Leibovitch et al., recommend that for treatment, radiotherapy should be avoided since MAC tumours are relatively radiotherapy resistant; surgical excision is the best method [10]. However, standard wide local excision has a recurrence rate of 40-60% [10]. Similarly, a study of 15 patients by Cooper et al., reported a recurrence of 47% with wide excision surgery and a study of 27 patients by Sebastian et. al reported a recurrence rate of 60% [11,12]. Leibovitch et al., suggests that this high reoccurrence may be because standard histologic methods of margin control are based on "bread-loaf" sectioning, therefore, only examining 0.2% of the margins. This is problematic because there may be residual undetected tumours [10].

For this reason, Mohs micrographic surgery (MMS) is the preferred method of excision because MMS utilizes horizontal en-face sectioning of the outer surface of the excised tissue. This method allows for examination of both peripheral and deep margins [10]. This is backed by the fact that several case reports have been published that utilized MMS and had low recurrence rates; Snow et al., performed a literature review from 1972 for all MAC cases treated with MMS (39 cases with more than two years of follow up). Their review reported that only four out of 39 cases (10.3%) had recurrence [13]. Friedman et al., published a similar review for MAC treated cases that were reported from 1984 to 1998; it was found that no recurrence was noted in 17 cases over a period of six weeks to 36 months [14]. Leibovitch et al., however, did perform MMS and had one case of recurrence in their 20 patients study (5%) [10].

SEER database analysis further confirms that surgery is the preferred treatment as 92% of the patients received surgery, only 10 patients of their 164 received radiotherapy [5]. Chiller et al., showed that after excisions of the primary tumour, 16 of the 23 patients that were treated with wide-excision surgery were fully cured [2]. The remaining 7 were either given micrographic surgery or another wide local excision was preformed; only 1 patient required 4 excisions until they were cured [2]. Based on the data and recommendations of the examined literature, MMS is the preferred treatment as it has low recurrence rates.

As MAC is a rare tumor, and the differential diagnosis includes a wide spectrum of benign and malignant entities, it can be misdiagnosed as reported in the case presented by Rustemeyer et al., which misdiagnosed MAC as trichoepithelioma for over four years [3]. It is our hope that this case report and review will increase the awareness of clinicians and pathologists to this rare tumor and the wide variety of possible differential diagnosis; minimizing the probability of misdiagnosis of this entity is essential, particularly for understanding the optimal treatment.

### Table 1: Distribution of MAC Occurrence within various Races; Data collected from SEER Database Analysis by Patel et al.

<table>
<thead>
<tr>
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<th>Male Occurrence (%)</th>
<th>Female Occurrence (%)</th>
<th>Total (%)</th>
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<tbody>
<tr>
<td>White</td>
<td>36.4</td>
<td>54.6</td>
<td>91.0</td>
</tr>
<tr>
<td>BAP</td>
<td>1.6</td>
<td>2.4</td>
<td>4.0</td>
</tr>
<tr>
<td>Other</td>
<td>2.0</td>
<td>3.0</td>
<td>5.0</td>
</tr>
<tr>
<td>Total (%)</td>
<td>40.0</td>
<td>60.0</td>
<td>100.0</td>
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**Figure 2** Graph showing MAC distribution by race and gender; Data collected from SEER Database Analysis by Patel et al.
It can be seen here that females have a higher presentation of MAC, particularly white females. White males have a higher presentation of MAC in contrast to other males. This preference for white males and females indicates a potential link between MAC and skin colour.

It can be seen here that the head and neck regions present with the highest incidence, particularly in white females and white males. The second most common location for MAC lesions was the trunk.

REFERENCES