A Case Report of Microcystic Adnexal Carcinoma Treated with Mohs Micrographic Surgery

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Microcystic adnexal carcinoma is a rare cutaneous appendageal malignancy, which is characterized by local aggressiveness. Histopathologically and immuno-histopathologically, the tumor reveals mixed eccrine and pilar differentiation. To date, Mohs micrographic surgery provides a more effective therapy and better outcome than traditional wide excision. We report a 66-year-old male with a slowly growing whitish indurated plaque approaching his left nasolabial area for at least 20 years. A diagnostic biopsy showed deeply infiltrating cords and strands of tumor cells with eccrine-like and pilar-like differentiation surrounded by a sclerotic stroma. Microcystic adnexal carcinoma was diagnosed and Mohs micrographic surgery was performed consequently. This is the first reported case treated with Mohs micrographic surgery in Taiwan. (Dermatol Sinica 22 : 301-305, 2004)

Key words: Microcystic adnexal carcinoma, Mohs micrographic surgery

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INTRODUCTION

Microcystic adnexal carcinoma (MAC), or sclerosing sweat duct carcinoma, is a rare skin appendeal carcinoma first described by Goldstein in 1982. Most cases reported previously are Caucasian; but in the recent years there is a rising population that this tumor occurring among patients with Oriental or African background. The clinical course of MAC, like many other skin malignancies, is relatively benign and asymptomatic. However, the tumor is highly infiltrative pathologically. Mohs micrographic surgery (MMS) now is considered a superior method than wide excision in the rate of recurrences.

CASE REPORT

A 66-year-old male presented to our clinic with a slowly growing whitish indurated plaque approaching his left nasolabial area for more than 20 years (Fig. 1). The plaque was asymptomatic, measuring approximately 1.5 cm in diameter. The surface of this plaque was irregular with slight protruding margin. On physical examination there was no palpable cervical or axillary lymph node. This patient did not receive any previous radiotherapy or immunosuppressive therapy. The patient's medical history was otherwise unremarkable. An initial punch biopsy was performed under local anesthesia for the purpose of diagnosis. The specimen was taken with some clinically normal skin and the depth was reaching to the subcutaneous layer with visible fat tissue. On hematoxylin and eosin stain, the tumor showed multiple scattered tumor islands arranged in cords and strands from upper to lower reticular dermis and even deep into subcutaneous tissue (Fig. 2). Between these tumor cords and strands, moderate amount of branching ductal structures, some containing keratin, could be identified. These tumor islands were composed of basoloid cells without significant mitosis and pleomorphism. Scanty amount of mononuclear cells could be found around

Fig. 1 A slowly growing whitish indurated plaque with pearly margin on his left cheek.

Fig. 2 Deeply infiltrative cords and strands of tumor cells with eccrine differentiation among the sclerotic stroma. Cystic structures can be identified in the upper dermis. (H & E, 40X)

Fig. 3 CEA stain. Ductular structures with positive activity in the apical portion (inner cells) (arrow) and amorphous contents (arrow head) in the lumen exhibiting eccrine differentiation within the tumor nests. (CEA stain, 400X)
there was no retraction artifact around these cords or strands. The stroma showed a sclerotic change. No melanin deposition was found in any of these tumor islands. Initially, trichoblastoma and desmoplastic trichoepithelioma were impressed in the above descriptions. However, the immunohistochemical study showed these ductal lumens and glandular components revealing positive activity for carcinoembryonic antigen (CEA) (Fig. 3). The final pathologic diagnosis of this plaque was microcystic adnexal carcinoma. Consequently, Mohs micrographic surgery was performed, since the infiltration of the tumor was extensive and this procedure was of much help to catch the panorama of the tumor. The initial excisional margin of the tumor was additional 1 to 2 millimeter wider than the visible tumor margin. A tumor-free plane was achieved after one stage surgery and a rhomboid flap (2 cm x 5 cm) was applied for the reconstruction of this surgical facial defect. The wound healed smoothly and the stitches were removed a week later (Fig. 4). The patient has been followed monthly in our outpatient clinic for 5 months and there was no evidence of recurrence so far.

DISCUSSION

MAC is known to have an insidious clinical course and locally invasive behavior. It typically occurs in mid-aged women as a slowly growing asymptomatic lesion, while some patients reported to have tenderness, numbness, paresthesia, and anesthesia. The common site of MAC is centro-facial area but some unusual sites are also reported in the literatures such as scalp, breast, and genital area. Distal metastases seldom occur except for severely immunocompromised patients. Carroll et al. reported a patient with an underlying chronic lymphocytic leukemia had multiple inoculations of recurrent MACs on scalp, along with regional lymph node metastases. Patients with MAC may have a history of previous local radiotherapy for acne or other malignancy unrelated with MAC, but large series on occupational exposure to radiation and its association with MAC remained elusive. The incidence of MAC and other cutaneous epithelial malignancies, like squamous cell carcinoma and basal cell carcinoma (BCC), may arise after immuno-suppression therapies especially among the recipients of organ transplant. UV exposure was proposed to trigger the mutation of P53 and other related genes to facilitate the formation of MAC but conclusive data is still lacking to date. However, Ohstuka and Nagamatsu collected 51 Japanese patients with MAC. In this study, 25 of the cases were reported between 1987 and 1996, and the remaining 26 cases between 1997 and 2000. It was clear that among the Orientals like Japanese, the number of patients with MAC is growing. The rising of case number in the recent years may be related to the fact of the thinning of ozone layer, which increases the amount of UV reaching the earth or well-recognized gradually by dermatopathologists and pathologists.

Clinically, the differential diagnoses of MAC include various tumorous, sclerotic and cystic lesions, such as basal cell carcinoma (BCC), squamous cell carcinoma, other adnexal tumors, hypertrophic scar and epidermal inclusion cyst. Chiller et al. estimated there may be 30% of misdiagnosed cases for MAC pathologically because it shares many similar features with other tumors. It is usually misdiagnosed pathologically as other benign or malignant tumor such as syringoma, desmoplastic tri-
choepithelioma (DTE) or morpheaform BCC especially in the era lack of multiple immunohistochemical tools. In the recent investigations, MAC may exhibit positive reactivities for carcinoembryonic antigen (CEA), epithelial membrane antigen (EMA), S-100, AE13, AE14 and Leu-M1 stains. In eccrine sweat glands, CEA typically stains the apical portion (inner cells) of acrosyringium and dermal eccrine ducts, as well as the amorphous contents in the lumen, which exhibits eccrine ductal differentiation in MAC as in our case. However, CEA may not be a reliable marker to differentiate eccrine duct from apocrine duct. As to the origin of MAC, controversies still exist if these tumor cells are pilar and eccrine differentiation or pilar and apocrine differentiation. MAC may occur in some sites containing apocrine glands such as breast, axilla and face, while some MAC may show prominent sebaceous gland differentiation, which implies these tumors may be derived from folliculo-sebaceous-apocrine unit. Perineural invasion is a frequent finding of MAC, which may account for the burning or tenderness sensation on these lesions. In our case there is no obvious perineural invasion and no clinical symptom was reported by the patient. The three tier changes of MAC in the dermis was proposed by Ackerman et al., who characterized the classic pattern of MAC: (1) the upper dermis resides cornifying cystic lesions; (2) cord and other solid structures occupy the center; (3) tubules house homogeneous material in the lower part. However, these three components are not always present, but they are useful to help rule out other tumors. In our case, this tumor does not show prominent cystic structures as in classic MAC; instead, cords and strands of tumor cells in sclerotic stroma constitute the main components of this tumor and the pathologic diagnosis of solid carcinoma should be considered. Solid carcinoma is a variant of MAC but some authors regard this as a discrete entity. Syringoma should be differentiated from MAC because they both have few cellular atypia, few mitotic features and share tubular structures. MAC often penetrates into the subcutaneous layer as an infiltrative ill-circumscribed tumor while syringoma is often limited in the upper dermis as a localized well-circumscribed tumor. DTE, like MAC, has cornifying cysts but it does not often have tubular structure, perineural invasion, and subcutaneous fat layer invasion.

The application of MMS for cutaneous malignancies has been well-documented, which is considered a better method to excise these tumors. Lower recurrence, better cosmetic outcome and better functional reserve establish the superiority than traditional wide excision although this procedure is still time-consuming and costly. Randle, et al. demonstrated a large single section to provide histologic preparation, which is much time-saving and may avoid another saucerization excision. Newer and much mature techniques to modify this procedure will sprout in the recent years because the number of cutaneous malignancies is climbing. Snow et al. have analyzed 148 cases of MAC treated by MMS in which only 8 cases have local recurrence during follow-up period. The success rate with more than 2-year follow-up is 89.7% while the recurrence rate of traditional excision is 41% by Carroll et al. Although the cases treated by other methods, such as wide excision, were not included in this study to compare their efficacy with MMS, this procedure is still more predictable than traditional wide excision on deep and lateral margin of the tumors, especially MAC characterized with highly infiltrative invasion. However, long-term follow-up is still suggested, because one case recurred 30 years after treatment. It can not be stressed that MMS may be the optimal treatment for MAC to date.

In conclusion, MAC, a rare entity in Taiwan, should be kept in mind, and the importance of MMS for MAC should be imposed to achieve a lower recurrence rate and better cosmetic and functional satisfaction for the patient.

REFERENCES
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