

Microcystic adnexal carcinoma in a non-Caucasian patient: A case report and review of the literature

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Received December 4, 2014; Accepted January 11, 2016

DOI: 10.3892/ol.2016.4242

Abstract. Microcystic adnexal carcinoma (MAC) is extremely rare among Asians, with the majority of cases presenting in Caucasian individuals. The current study describes the case of a 38-year-old Chinese woman who presented with a 10 year history of a mass in the upper lip. A biopsy resulted in a diagnosis of MAC. The patient underwent complete surgical resection and the tumor was successfully excised. During 6 months of follow-up, there was no evidence of recurrence. To the best of our knowledge, the present case is the first Chinese case to be reported in the English literature, and is presented with the aim of increasing the awareness and aiding in the management of MAC in non-Caucasian populations.

Introduction

Microcystic adnexal carcinoma (MAC) was originally defined in 1982 (1), and is now also termed sclerosing sweat duct carcinoma (2). MAC is a rare, slow-growing, locally aggressive tumor of the eccrine sweat glands, and is characterized by pilar and eccrine differentiation (3). Sweat gland malignancies, combining all types, are particularly rare, accounting for 0.005% of all malignant epithelial neoplasms (4), with a total of ~350 cases currently described in the English literature. It has been reported that the preferential location for the development of this neoplasm is the head and neck region (5).

When referring to the English literature, it is evident that MAC primarily affects the Caucasian population, with only two cases occurring in non-Caucasian patients (6,7). Fang *et al* (8) described two cases involving Chinese

individuals in the Chinese literature; however, to the best of our knowledge, there are no cases of Chinese patients with MAC currently reported in the English literature. The present study reviewed all patients that were referred to the Hainan Province People's Hospital (Haikou, China) between January 1994 and January 2014, and identified only one patient with a MAC diagnosis. This case is described herein along with a review of the relevant literature.

Case report

A 38-year-old woman was referred to the Department of Oral and Maxillofacial Surgery at the Hainan Province People's Hospital with a mass on the left upper-lip that had been present for >10 years (Fig. 1). During physical examination, it was noted that the tumor was positioned in the left-upper lip and subnasal region. The lesion was a firm, solitary mass, and measured 2x1 cm in size. No submandibular or cervical lymph nodes were palpable during the physical examination. Abdominal ultrasonography and computed tomography (CT; Brilliance iCT; Philips Medical Systems, Amsterdam, Holland) of the head, neck and thorax demonstrated no evidence of systemic or local metastasis. As presented in Fig. 2, the head and neck CT scans revealed the lesion located in the subcutaneous tissue of the left-upper lip, with a wide base inseparable from overlying skin.

An incisional biopsy was performed with the patient under local anesthesia. The diagnosis of the specimen was confirmed as carcinoma. The patient underwent complete surgical resection of the lesion, with 10 mm tumor-free margins, which resulted in a subtotal defect of the philtrum and left-upper lip. Reconstruction of the tissue defects was performed immediately, utilizing an adjacent tissue flap of skin and mucous membranes from the cheek (Fig. 3).

Hematoxylin and eosin staining (ZSGB-BIO, Beijing, China) was performed on 4- μ m thick formalin-fixed and paraffin-embedded resected tumor tissue. Staining was visualized using a BX53 microscope (Olympus Corporation, Tokyo, Japan). Histopathological analysis demonstrated that MAC had infiltrated a number of muscle tissues and peripheral nerve fibers (Fig. 4). Surgical margins were negative. No chemotherapy or radiotherapy regimens were administered,

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Key words: microcystic adnexal carcinoma, upper lip



Figure 1. Firm dermal nodule on the left of the upper lip and subnasal region.

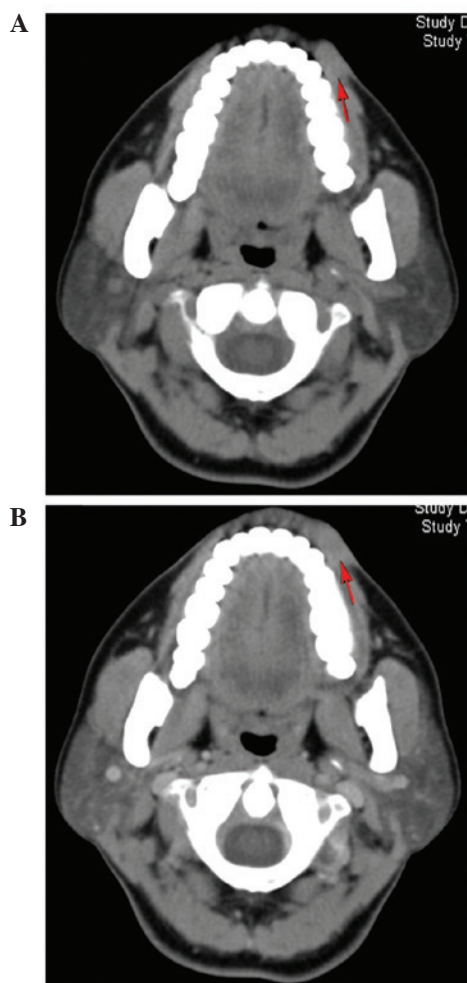


Figure 2. Pre-operative CT of the present case. (A) Contrast axial CT demonstrating the position of the lesion in the subcutaneous tissue of the left upper-lip (arrow), with a wide base that was inseparable from the overlying skin. (B) Axial contrast-enhanced CT demonstrating a slight enhancement of the lesion (arrow). CT, computed tomography.

and no recurrence was observed during 6 months of follow-up. Written informed consent was obtained from the patient for the publication of this study.

Discussion

The clinical presentation of MAC is characteristically a smooth-surfaced, slow-growing, firm, non-ulcerated,

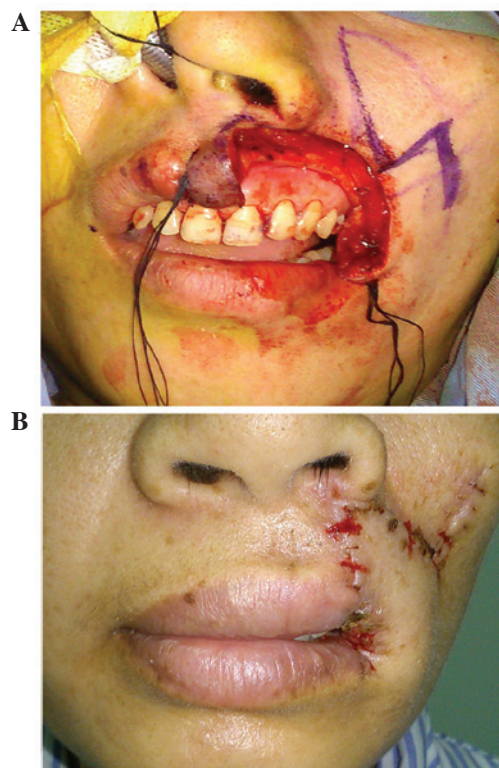


Figure 3. (A) Incision lines for reconstructive surgery marked with blue ink with a rotation flap from the cheek. (B) Post-operative appearance subsequent to reconstruction.

asymptomatic papule, plaque or nodule (9). Nonetheless, a small number of patients are symptomatic at presentation, experiencing burning, paresthesia and numbness as a result of recurrent perineural invasion (9).

It has been reported in the literature that certain MAC cases have presented with tumor histories as long as 17 and 27 years (3,10). Notably, due to the asymptomatic presentation and extremely slow-growth of the tumors, all reported patients have been diagnosed at least 1 year after the initial appearance of the lesions. In the present case, the lesion was present for >10 years prior to diagnosis.

Despite the pathogenesis of MAC not yet being fully understood, it is believed to originate from pluripotent keratinocytes, which are capable of differentiation into hair follicles or sweat glands (11). Clinically, the disease presents in the fourth to seventh decades of life, with no evident gender predilection (11,12). In 85% of cases, the tumors develop in the head and neck area (13), demonstrating a preference for the periorbital skin and the centrafacial region.

MAC is commonly misdiagnosed clinically and histologically, primarily due to its inconspicuous features (14). Rustemeyer *et al* (15) described a case of the disease that was misdiagnosed as desmoplastic trichoepithelioma for a period of 4 years.

The large majority of reported MAC cases have occurred in Caucasians; however, a small number of reports in the literature have described disease occurrence in non-Caucasians (6,7). To the best of our knowledge, there are currently no reports in the English literature regarding Chinese patients with MAC. However, due to the high prevalence of misdiagnosis and

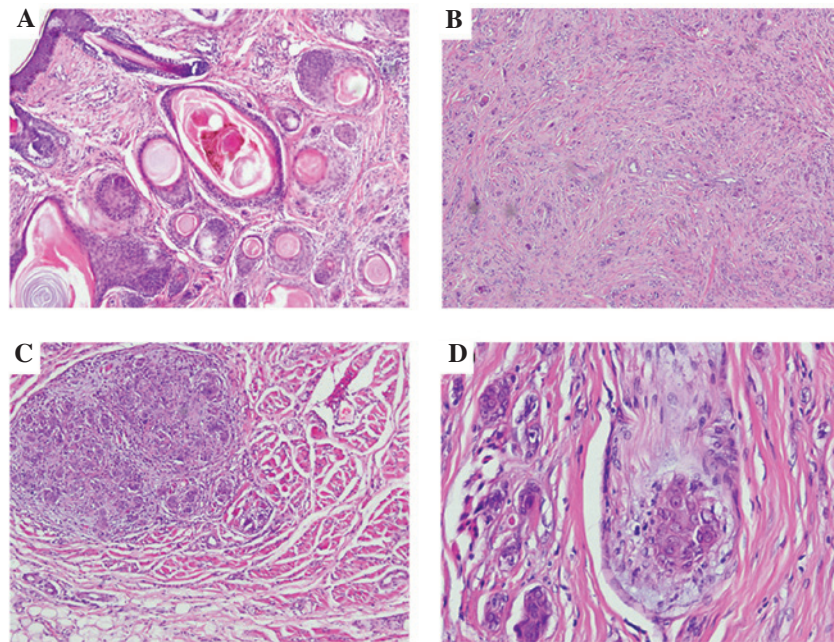


Figure 4. Histological examination of the biopsy tissue. (A) The upper dermis of the tumor exhibiting micronests, cords of cells in dense desmoplastic stroma and keratinous cysts (magnification, x10); (B) Squamoid and glandular clusters are presented in a desmoplastic stroma in thicker sections. Cellular atypia and mitoses are scarce (magnification, x10); (C) MAC-infiltrated muscle tissue (magnification, x10); (D) MAC-infiltrated the peripheral nerve fibers (magnification, x40). Staining, hematoxylin and eosin. MAC, microcystic adnexal carcinoma.

possible under-recognition of the disease within this population, a diagnosis of MAC may be significantly overlooked in non-Caucasian individuals as it most commonly occurs in Caucasians. The present case demonstrates the requirement to include MAC in the differential diagnosis in non-Caucasian patients.

The disease presents histologically as a deeply-infiltrative and poorly-circumscribed tumor, consisting of solid aggregates of tumor cells in the mid dermis, superficial keratinous cysts and elongated tubular structures in the deep aspect of the lesion. These cell nests are enveloped by dense, fibrous stroma (16).

MAC is resistant to radiotherapy and chemotherapy, which may be predicted from its slow biological activity (9). The preferred treatment option is complete surgical excision; however, the true tumor-free margins are typically far beyond the intraoperative macroscopically established margins (17,18). Mohs micrographic surgery may be regarded as the gold standard treatment option, with adjuvant radiotherapy and wide local excision offering comparable efficacy (19). When performing adjuvant radiotherapy, a dose of >50 Gy should be administered with wide margins (3-5 cm), owing to the tendency of the lesion for perineural and deep invasion (20). The current patient was not administered any chemotherapy or radiotherapy.

The aggressive and locally invasive nature of the disease results in high recurrence rates, ranging from 15-60% depending on the treatment modality, with lower rates of recurrence observed following Mohs micrographic surgery (21-24). However, such data may be an overestimate, with misdiagnosis often occurring at the time of excision, and positive margins can occasionally be misread.

It has been reported that tumor recurrence may occur up to 30 years after the initial surgical excision (12), highlighting the importance of regular, long-term follow-up.

Regional and distant metastasis have been described in the literature; however, such cases are particularly rare, with only five cases of local recurrence and three cases of distant metastasis reported (25,26). According to the relevant literature, only one mortality has occurred as a result of the disease (27).

MAC is extremely rare among non-Caucasians, with the majority of cases presenting in Caucasian individuals. The present case describes the occurrence of MAC of the lip in a Chinese woman. To the best of our knowledge, the current case is the first of its kind to be reported in the English literature. The case highlights the importance of the inclusion of MAC within the differential diagnosis in non-Caucasian individuals, and additionally aims to increase the awareness and aid the management of MAC in non-Caucasian populations.

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