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Primary cutaneous mucinous carcinoma of the cheek: a case report

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Abstract

Background Primary cutaneous mucinous carcinoma is a rare neoplasia of the sweat gland. The age-adjusted incidence was 0.024 tumors per 100,000 person-years. It is possible that the actual number of tumors may be slightly higher than previously estimated as some cases of primary cutaneous mucinous carcinoma may have been mistaken for benign tumors and removed by laser therapy without histologic examination.

Case presentation We report a 58-year-old Chinese man with primary cutaneous mucinous carcinoma. The patient presented to our care with an indolent nodule on the left cheek, which was proven to be a mucinous adenocarcinoma by excisional biopsy and immunohistochemical staining. Following a comprehensive evaluation, including whole-body computed tomography and positron emission tomography, metastases from other sites were ruled out and the patient was diagnosed with primary cutaneous mucinous carcinoma. The patient underwent an additional wide resection surgery to ensure a safe margin and was then recommended to undergo regular follow-up.

Conclusion This case is one of the few published Chinese cases in literature of primary cutaneous mucinous carcinoma. Diagnosis of primary cutaneous mucinous carcinoma is challenging, and treatment options are limited. Collaboration between clinicians and pathologists is crucial for optimal outcomes. Further studies with longer follow-up periods are necessary to provide evidence for the management of this disease.

Keywords Primary cutaneous mucinous carcinoma (PCMC), Case report, Histopathologic examination

Introduction

Primary cutaneous mucinous carcinoma (PCMC) is a rare neoplasia of the sweat gland. The age-adjusted incidence was 0.024 tumors per 100,000 person-years [1]. It is possible that the actual number of tumors may be slightly higher than previously estimated, as some cases of PCMC may have been mistaken for benign tumors and removed by laser therapy without histologic examination [2]. PCMC is a low-grade carcinoma with few metastases and a favorable prognosis, despite the possibility of local relapse [3]. Aggressive biological behavior occurs only rarely [4]. The 5-year and 10-year overall survival rates were 85.0% and 78.0%, respectively, with a mean overall survival (OS) of 11.4 years [5]. Here, we report the case of a 58-year-old Chinese man with a slow-growing nodule on the left cheek, which was diagnosed as PCMC for the first time in China in English literature.

Case presentation

In 2022, a 58-year-old Asian male presented to our facility with a painless subcutaneous nodule on his left cheek (Fig. 1). The patient reported the discovery of a sesame-like black superficial nodule on the left cheek in 2017 and underwent laser therapy at a local clinic without a pathologic biopsy. The lesion recurred in 2019 and grew

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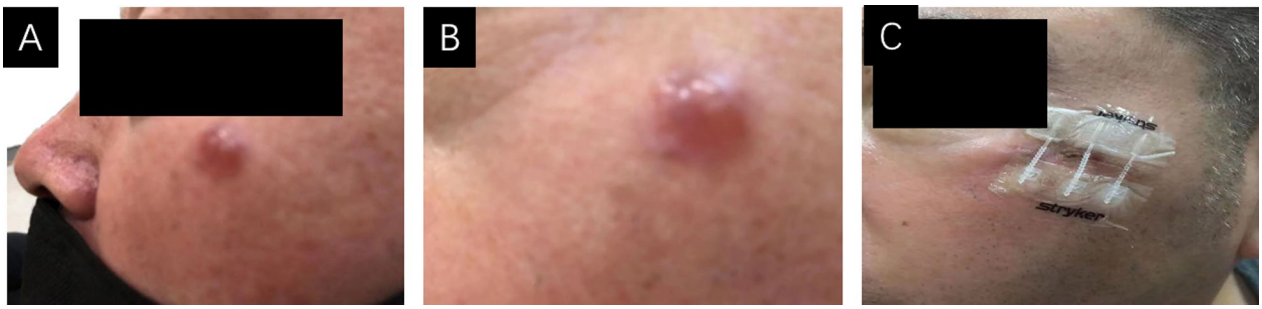

Fig. 1 Lesion photographs. **A** External photos of the lesion on the left cheek which measures 0.6 cm × 0.6 cm. **B** The nodule exhibited a medium texture, with clear borders and an intact smooth surface. **C** Left cheek after the excisional biopsy

gradually over the next 3 years. Upon examination, the nodule was observed to be a round shape measuring 0.6 cm × 0.6 cm, with a reddish coloration. It exhibited a medium texture, with clear borders and an intact smooth surface. The regional lymph nodes were not palpable. No other obvious abnormalities were observed on physical examination, including the nervous system. The heart rate of the patient was 80 beats per minute, blood pressure was 116/72 mmHg, and body temperature was 36.5 ℃. There was no history of previous disease, radiation, or toxic substance exposure, allergy, or trauma. The patient did not smoke or drink alcohol. Family history of cancer or similar lesions was also denied.

An excisional biopsy was performed on 8 June 2022. A histopathologic examination of the mass revealed heterogeneous epithelial cells floating in mucinous lakes among subdermal fibrous tissue. The neoplastic cells stained positive for cytokeratin 7 (CK7), while a few cells were positive for synaptophysin (Syn) and chromogranin-A (CgA). The staining for C-kit (CD117), p63, cytokeratin 20 (CK20), p40, mucin-5 subtype AC (MUC5AC), and homeobox transcription factor (CDX2) was negative (Fig. 2). The morphologic and immunohistochemical features indicated mucinous adenocarcinoma. Investigations for a primary visceral malignancy, including a total body positron emission tomography-computed tomography (PET-CT) scan, yielded unremarkable results (Fig. 3). The slides were sent to the oral pathology department of the Ninth People's Hospital for consultation and the results were in accordance with the primary pathological diagnosis. Immunohistochemistry showed the following staining pattern: CK7 (+), CK19 (+), CEA (small amount+), CAM5.2 (+), p63 (−), p40 (−), and CK5/6 (−). An additional wide resection of the tumor was carried out at the Ninth People's Hospital on 8 July 2022, to ensure safe margins. Postoperative histopathologic examination revealed focal areas of adenoepithelium within subcutaneous tissue on a mucin-rich background. Combined with the consulting slides, the features agreed with postoperative mucinous adenocarcinoma residue. The surgical margins were negative for infiltration. The patient was suggested to undergo regular surveillance. The last follow-up was in May, 2024 and no recurrence or metastasis was observed.

Discussion

In this case, we report a 58-year-old man with an indolent nodule on the left cheek that was diagnosed to be PCMC, a very uncommon entity which was rarely reported in the Chinese population. After ruling out metastases from other sites, the patient underwent an additional wide resection surgery and then underwent regular follow-up.

Primary cutaneous mucinous carcinoma (PCMC) is an uncommon entity. It was first described by Lennox *et al.* in 1952 [6] and formally reviewed by Mendoza and Helwing in 1971 [7]. The majority of observations have been derived from case reports and limited cohort evaluations owing to the rarity of this disease. Until recently, analyses based on large samples from databases have been conducted, overturning some inconsistent conclusions because of sampling errors and publication bias, these include the predominance of certain sexes or races [1, 5].

PCMC typically manifests as indolent, erythematous, asymptomatic nodules, papules, cysts, or ulcers [8, 9]. The highest incidence of PCMC was in individuals in their 60s or 70s [1, 5]. The head and neck was the most common site of involvement, particularly the eyelids [1, 5]. Eyelid PCMCs were more likely to present with distant disease, which may be the result of rapid tumor access to lymphatic tissue and a rich underlying vascular supply [1]. The average tumor size was 1.63 cm [1]. The tumor showed an inert biological behavior, most of patients had local disease, 10.5% had regional disease (including involvement of the regional draining lymph nodes and local extension beyond the subcutaneous tissue), and only 5.8% had distant disease [1].

The histogenesis of PCMC remains debatable [10]. Traditionally, it was believed to originate from eccrine

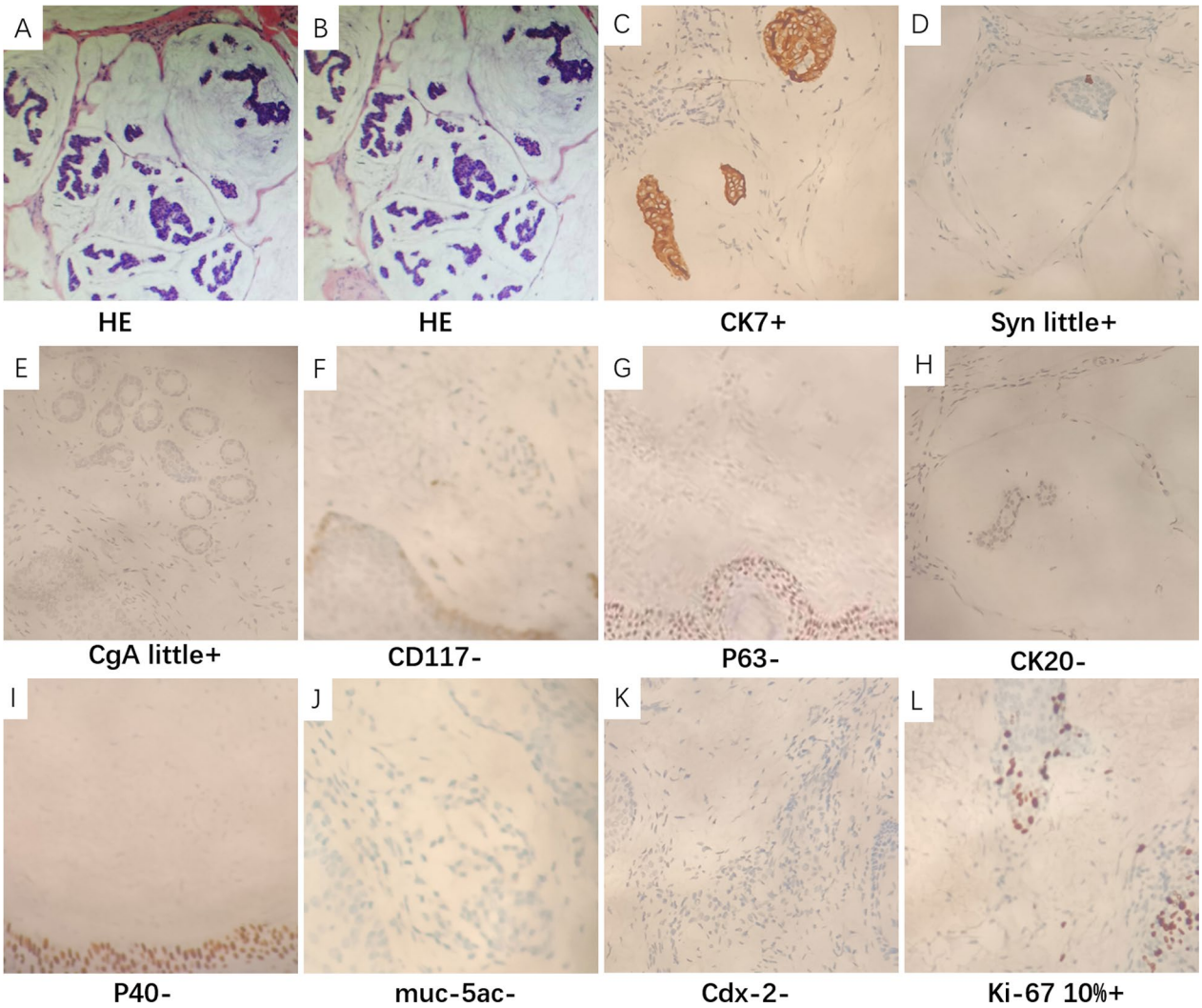

Fig. 2 Histopathology and immunohistochemistry of the left cheek lesion showing heterogeneous epithelial cells floating in mucinous lakes among subdermal fibrous tissue (hematoxylin and eosin, 40×) (**A** and **B**). Tumor cells demonstrating reactivity to CK7 (100×) (**C**). A few cells were positive for Syn (**D**) and CgA (**E**). The staining for CD117 (**F**), p63 (**G**), CK20 (**H**), p40 (**I**), MUC5AC (**J**), and CDX2 (**K**) was negative) (magnification, 100×). Ki-67 staining was 10% positive (**L**)

glands, but subsequent evidence suggests an apocrine lineage [2, 3, 11]. Mucinous carcinoma is distinguished histologically by the presence of nests and strands of epithelial cells immersed in copious pools of mucin and divided by thin fibrous septa [11]. According to 2006 World Health Organization (WHO) classification criteria, exhibits an immunoprofile that is positive for low molecular weight cytokeratins, carcinogenic embryonic antigen (CEA), epithelial membrane antigen (EMA), gross cystic disease fluid protein 15 (GCDFP-15), α-lactalbumin, salivary amylase, and β2-microglobulin [9]. Additionally, nuclear expression of estrogen receptors (ER) and variable expression of progesterone receptors (PR) have been described [9].

The most commonly reported differential diagnoses include epidermoid cyst, sebaceous carcinoma, cystic basal cell carcinoma, squamous cell carcinoma, neuroma, lacrimal sac tumor, and pilomatric carcinoma [9]. Differential diagnosis between primary and metastatic mucinous eccrine carcinoma is critical because the latter has a greater invasive capacity, metastatic potential, and much worse prognosis. A number of features may suggest a primary cutaneous origin, including the presence of an *in situ* component, the presence of myoepithelial cells around tumor lobules and immunohistochemical positivity of tumor cells for p63, podoplanin, and CK15 [11]. CK 20 is negative in PCMC, allowing differentiation from metastatic mucinous gastrointestinal carcinoma.

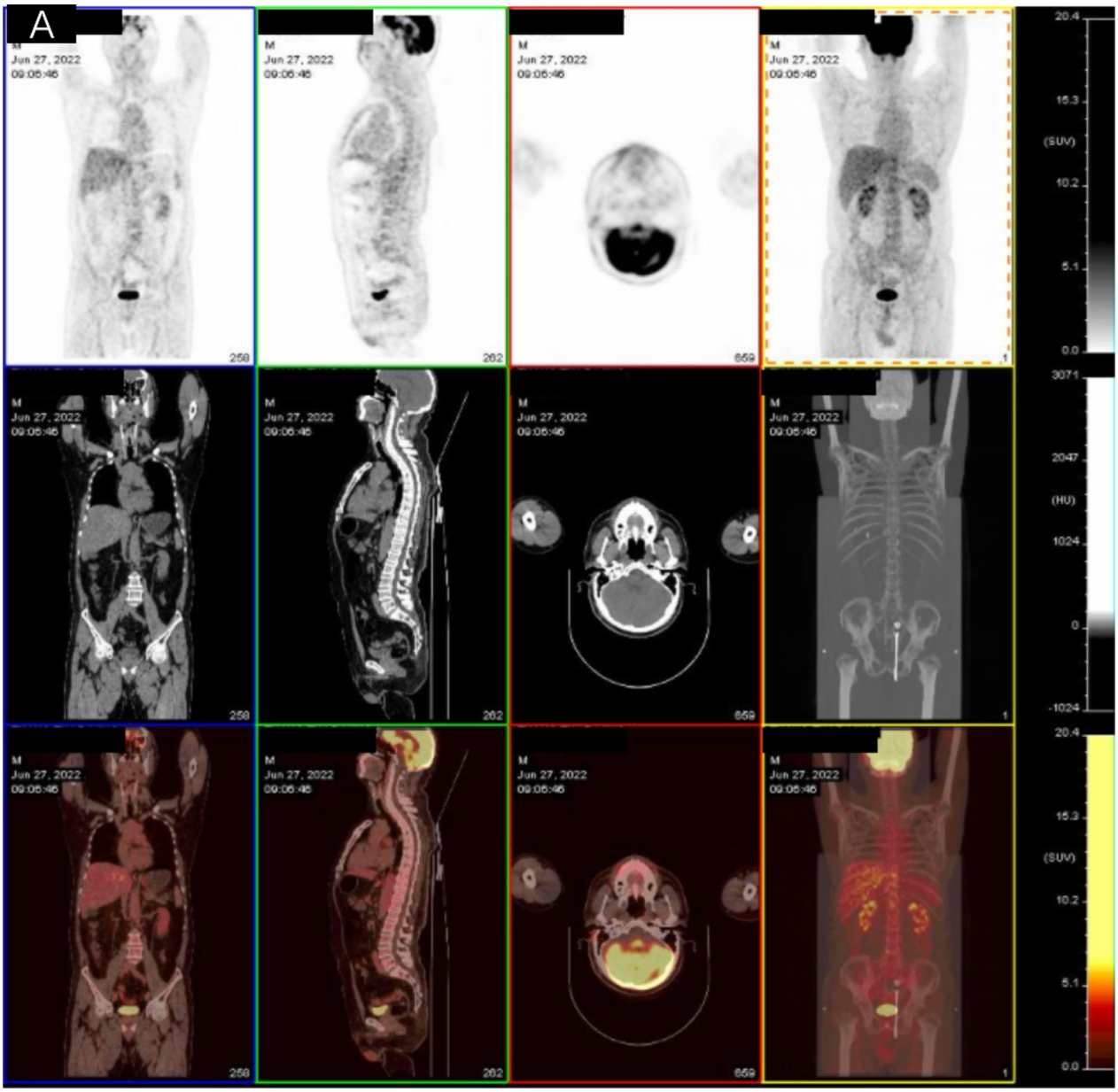

Fig. 3 A positron emission tomography-computed tomography ruled out other primary malignancies

Recent studies also suggest that the expression of CK 7 expression may assist in the diagnosis of PCMC, as these markers are rarely found in metastatic mucinous adenocarcinoma [9]. However, it should be noted that none of these features are absolutely discriminatory, and thus a thorough work-up to exclude systemic metastases is recommended [11].

Complete surgical resection is the gold standard treatment for this tumor [12]. Mohs surgery has been advocated as a first-line therapy because it can provide complete margin control while sparing uninvolved tissues resulting in a satisfactory cosmetic outcome [2]. The application of Mohs surgery has been demonstrated to bring a higher rate of disease control [8] and a better 5-year OS [5]. Recurrence is often associated with specific tumor location and incomplete resection [3]. As recurrent tumors are often demonstrate resistance to radiotherapy or chemotherapy, they may be result in substantial morbidity [8]. Long term treatment response has been difficult to evaluate, as most case reports only have a few years of follow-up [2]. Given the absence of a standardized guideline for the treatment of PCMC, patients

are typically advised to undergo close surveillance following surgery. CEA and CA15.3 may help to detect early recurrence or disseminated disease [13]. The risk of subsequent primary tumor was increased in the first year after PCMC diagnosis. Within this time window, the most prevalent malignancy identified was lung cancer [1].

Conclusion

PCMC, in contrast to other sweat gland adenocarcinomas, is a low-grade malignant tumor with a proclivity for local recurrence. This case is one of the few published Chinese cases in PCMC literature. Diagnosis of PCMC is challenging and treatment options are limited. Collaboration between clinicians and pathologists is crucial for optimal outcomes. Further studies with longer follow-up periods are necessary to provide evidence for the management of this disease.

Abbreviations

PCMC	Primary cutaneous mucinous carcinoma
PET	Positron emission tomography
CK	Cytokeratin
Syn	Synaptophysin
CgA	Chromogranin-A
MUC5AC	Mucin-5 subtype AC
HE	Hematoxylin and eosin

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Author contributions

CBZ designed, performed the study, and carried out literature search; YD helped with the literature search and wrote the paper; YD, YFZ, HXL, and CBZ contributed to the acquisition and analysis of data; CBZ made critical revisions and supersized the study. All authors have read and approved the manuscript.

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Availability of data and materials

The datasets used and/or analyzed during the current study available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate

The present study was approved by ethics committee of Anhui Provincial Cancer Hospital.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests

Consent was signed by the patient for all the images, other personal and clinical details.

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