QUIZ SECTION

A Reddish Nodule on the Scalp: A Quiz

Yuelin LI[1†], Xiao TAN[2,3†], Hong REN[2,3], Guangquan JI[4*] and Renqiong CHEN[2,3*]
*[1]Graduate School, Jinzhou Medical University, Jinzhou, 121001, China, [2]Department of Dermatology, Affiliated Lianyungang Hospital of Xuzhou Medical University, Lianyungang, 222002, Lianyungang, China, [3]Department of Dermatology, Lianyungang Clinical College of Nanjing Medical University, Lianyungang, China, and [4]Department of Technology, Affiliated Lianyungang Hospital of Xuzhou Medical University, Lianyungang, 222002, Lianyungang, China. *Emails: chenrenqiong718@126.com; 530749091@qq.com*
†These authors contributed equally to this work.

A41-year-old man presented with a 7 month history of a painful nodule on his scalp. He had undergone excision of a left submandibular gland mass 5 years earlier. Clinical examination revealed a reddish, firm, infiltrative nodule with ulceration and crusting on the scalp (Fig. 1). Microscopic examination showed confluent nests of basophilic tumour cells within the dermis, accompanied by hyaline material, focal mucin deposition and prominent fibrous tissue proliferation (Fig. 2). Immunohistochemical staining was positive for EMA, CK7, CK8/18, CD117 and SMA (Fig. S1) and negative for CK20, CD34, CD56, P63 and S100. The Ki-67 proliferation index was approximately 60% (Fig. S1).

What is your diagnosis?

Differential diagnosis 1: Metastatic adenoid cystic carcinoma.
Differential diagnosis 2: Squamous cell carcinoma.
Differential diagnosis 3: Trichilemmal carcinoma.
Differential diagnosis 4: Cutaneous metastases from lung cancer.

See next page for answer.

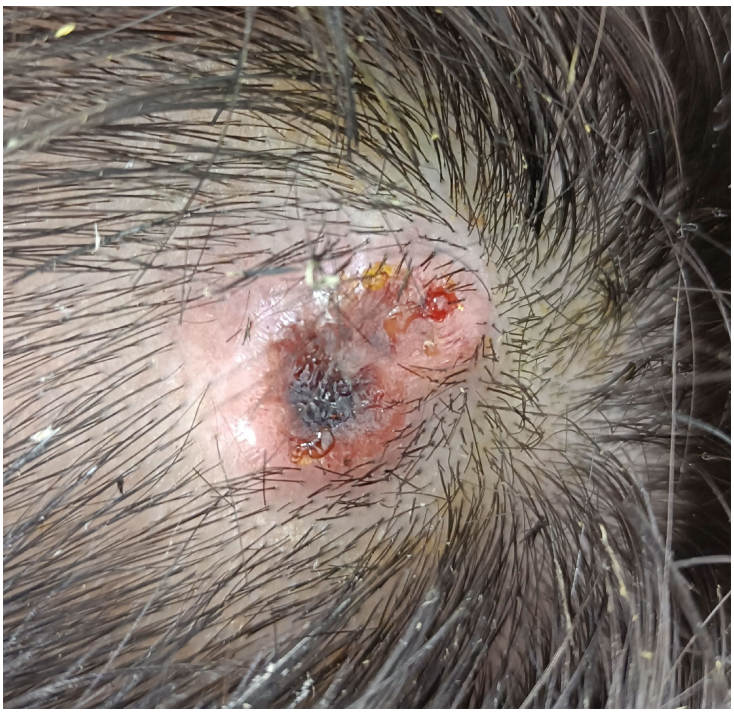

Fig. 1. Skin lesion on the scalp of the patient.

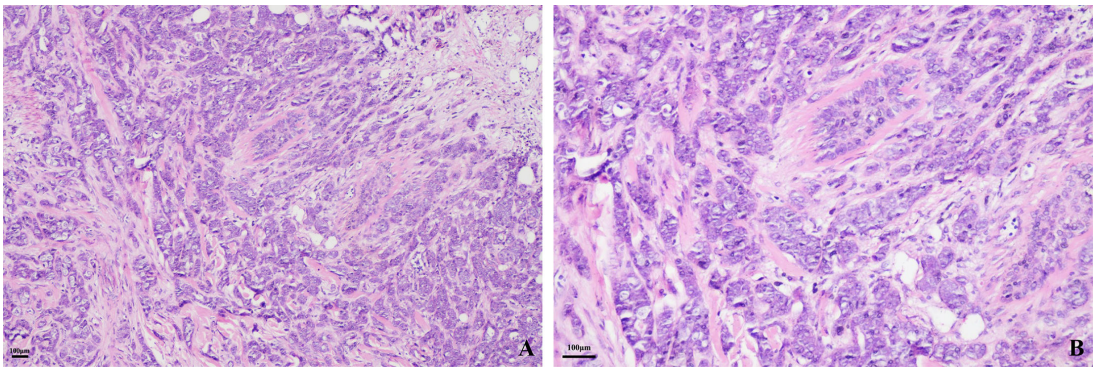

Fig. 2. The pathology showed confluent nests of basophilic tumour cells within the dermis, accompanied by hyaline material, focal mucin deposition and prominent fibrous tissue proliferation (HE staining, A:100×, B:200×).

ANSWERS TO QUIZ

A Reddish Nodule on the Scalp: A Commentary

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***Diagnosis*: Metastatic adenoid cystic carcinoma**

The patient presented with a painful lesion on the scalp 5 years after the initial diagnosis of submandibular gland adenoid cystic carcinoma (ACC). Skin biopsy confirmed metastatic ACC, characterized by a predominantly solid growth pattern. Cutaneous metastasis of ACC is exceedingly rare and generally indicates disease progression. Histologically, ACC can exhibit tubular, cribriform or solid growth patterns, with the solid pattern being associated with a more aggressive clinical course (1). In the present case, ACC showed a solid architecture, and ductal differentiation was observed under high-power magnification. Tumour cells were positive for CK7 and CK8/18. Primary cutaneous ACC and cutaneous metastasis of ACC are histologically indistinguishable, highlighting the importance of detailed clinical history and thorough examination in reaching an accurate diagnosis (2).

ACC is a slow-growing yet persistently invasive malignant neoplasm that can originate in salivary glands, the respiratory tract, breast and skin (3). It occurs most frequently in the submandibular gland and typically follows an indolent but relentless clinical course. The long-term prognosis is generally poor, largely due to a high incidence of late local recurrences and distant metastases (4). Distant metastases most commonly affect the lungs, followed by bone, liver and brain (5). Cutaneous metastasis arising from salivary gland ACC is particularly rare. Previous studies report that the median interval from initial diagnosis to metastasis is approximately 37 months, while median survival after non-pulmonary metastasis is about 21 months (6). Furthermore, it has been documented that 11 % of patients die within one year of metastasis, and one-third succumb within three years (7). Notably, our patient remains alive three years after the detection of lung metastasis and 6 months following the diagnosis of cutaneous metastatic disease.

The management of metastatic adenoid cystic carcinoma (ACC) remains clinically challenging. Current therapeutic strategies for metastatic ACC encompass surgical metastasectomy for isolated tumour deposits, localized radiotherapy, systemic chemotherapy and immunotherapy. Radiotherapy directed at isolated metastatic lesions has been demonstrated to effectively alleviate symptoms and achieve local disease control (2). Surgical resection of metastatic sites may be considered when radiotherapy fails to control local progression or alleviate patient symptoms; however, such interventions are generally palliative and do not significantly improve overall survival (8). Owing to the indolent growth kinetics characteristic of ACC, the utility of systemic chemotherapy is limited, with various regimens reporting unsatisfactory outcomes. Targeted therapy in ACC focuses on inhibiting the transmembrane tyrosine kinase receptor KIT, which is frequently overexpressed in ACC and can be immunohistochemically detected using CD117 antibodies (9). In patients with unresectable or metastatic ACC exhibiting CD117 overexpression, oral imatinib mesylate has shown modest clinical benefit (10).

ACC is characterized by marked neurotropism, exhibits a protracted clinical course and demonstrates persistent dissemination through both haematogenous and lymphatic pathways to adjacent and distant anatomical sites (10). This malignancy is associated with a high mortality rate, with the majority of patients succumbing within 5–10 years following initial treatment. Pathological evaluation in the present case confirmed ACC with extensive perineural invasion into the periglandular adipose tissue and striated muscle. Notably, despite the presence of widespread metastatic disease, the patient remains alive five years after the commencement of primary treatment.

Cutaneous metastases in ACC are exceedingly rare. This case underscores the critical importance of comprehensive cutaneous surveillance in patients with a documented history of ACC. Regular follow-up examinations are strongly advised, and the emergence of any new cutaneous nodules should raise clinical suspicion for metastatic spread, as they may represent a pivotal juncture in disease progression, thereby warranting timely adjustments in therapeutic management and prognostic evaluation.

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