Clear Cell Hidradenocarcinoma of the Skin: A Case Report and Literature Review of a Rare, Highly Malignant Lesion

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ABSTRACT

INTRODUCTION

Clear cell hidradenocarcinoma (CCH) is a rare tumor of eccrine sweat gland origin that is infrequently described in the literature. Malignant eccrine tumors occur in only one out of every 13,000 dermatopathological biopsies, with the CCH subtype approximating only 6% of these lesions. 1,2 First described in 1954 by Keasbey and Hadley3 as a highly malignant clear cell papillary tumor, several synonyms have been used to describe the lesion in the subsequent literature. These include nodular hidradenocarcinoma, clear cell eccrine carcinoma, malignant clear cell acrospiroma, solid cystic adenoacanthoma, and malignant clear cell myoepithelioma. 1,2,4,5,6

CASE REPORT

A 59-year-old Caucasian woman with no history of smoking presented to her dermatologist after noticing a new lesion of her right cheek. She denied tenderness, bleeding or any other associated symptoms. A biopsy demonstrated findings consistent with malignancy and she was subsequently referred to an otolaryngologist for surgical management.

The patient underwent a wide local excision (WLE) of the right cheek lesion, which was histopathologically diagnosed as clear cell hidradenocarcinoma of the skin. All margins were negative, with the closest margin measuring 0.5 mm to the inferior border. Extensive angioinvasion was noted. One month post WLE, patient presented to the Head and Neck Oncology clinic with a moderately enlarged, tender and swollen right neck mass following a recent upper respiratory tract infection. She was treated with a course of Augmentin for suspected lymphadenitis and instructed to return to clinic in two weeks. The right neck mass persisted and subsequent fine needle aspiration (FNA) demonstrated findings consistent with metastatic carcinoma.

Recommendations from the Multidisciplinary Tumor Board suggested repeat surgical resection and right parotidectomy with a modified radical neck dissection for concerns of regional lymph node metastasis and possible tumor recurrence. Prior to surgery, patient underwent a CT neck and PET/CT scan for full metastatic evaluation. Both failed to reveal any evidence of distant disease.

Due to its non-specific presentation, inconsistent terminology within the literature, and obscurity in clinical practice, misdiagnosis of CCH is not uncommon and diagnosis is confirmed only after histopathological evaluation. 5 Macroscopically, hidradenocarcinomas demonstrate an asymmetric, nodular shape that is typically 2 to 3 cm larger when compared to benign hidradenoma. 2,6,8 Cytology reveals vacuolated cells with abundant clear cytoplasm due to a high glycoprotein content, which is the cardinal finding of all clear cell tumors. 6 Histology demonstrates a prominent nodular or trabecular pattern, with uniform-sized nuclei. 5,6 Certain characteristics are helpful in differentiating CCH from its more common benign counterpart, nodular hidradenoma. Features favoring malignancy include increased mitotic activity, dense tissue extension, angioinflamatory or perineural invasion, and focal areas of necrosis. 5,6

These lesions most frequently occur on the head, neck, and axilla.1 Other clinical presentations and their corresponding treatment options.

DISCUSSION

Sentinel lymph node biopsy is useful in cases concerning for regional metastasis, however, no evidence currently supports a mortality benefit.1 Due to the high incidence of lymphovascular invasion some reports further recommend regional lymph node dissection at the time of surgical excision, but this remains controversial and should be reserved for patients with extensive local disease in the absence of known distant metastases. 1,2,10 Originally thought to be radio-resistant, a study by Harari et al. has shown positive outcomes with the use of adjuvant radiotherapy, describing a disease-free period in 2 patients without evidence of metastasis for 27 to 35 months post-operatively.11

The prognosis of CCH has traditionally been poor and, despite aggressive surgical management, a 5-year disease-free survival is reported in less than 30% of patients. 5,6 These tumors are associated with a 50% local recurrence and metastasis in 60% of patients within the first 24 months. 2,5,6 The average time to first recurrence is 16 months. 8 Metastasis favors regional lymphatics followed by hematogeneous dissemination to lung and bone. 5,6 Additionally, there seems to be an inversely proportional relationship to the size of the lesion and survival time.1

CONCLUSIONS

In conclusion, this case highlights the often-nonspecific presentation of CCH, a highly malignant neoplasm uncommonly encountered in clinical practice. Although no standard management protocol currently exists, most physicians recommend a multidisciplinary approach that primarily includes a margin-free resection and consistent long-term surveillance due to the high incidence of metastasis and local recurrence. Adjuvant therapies like radiation and neck dissection have proven beneficial in a number of cases, but without any clinical trials, no gold standard treatment plan exists.

REFERENCES