CASE REPORT PEER REVIEWED | OPEN ACCESS

Syringoid eccrine carcinoma: A case report and literature review of therapeutic options

Munaf AL-Kadhimi, Tamarah AL-Dawoodi, Julio Peguero, Luis T. Campos

ABSTRACT

Introduction: Syringoid eccrine carcinoma is exceedingly infiltrative neoplasm, focally destructive, slowly growing adnexal tumor, derived from eccrine sweat glands. It is frequently misdiagnosed, both clinically and histologically, with other benign and malignant neoplasms. Owing to perineural invasion and destruction, the tumor recurrence is high. However, the regional or distant metastases are very rare. The local aggressive nature of the tumor and the high recurrence rate may necessitate further sophisticated procedures. Case Report: We report a case of a 65-year-old Siberian female who has initially missed diagnosed with benign lesions and subsequently found to have syringocarcinoma. The patient had multiple recurrences treated with surgical resection, cisplatin and docetaxel, radiation therapy and laser therapy. Computed tomography scan obtained and revealed metastatic disease involving the bone. Due to the rarity of this type of skin cancer, next generation sequencing was obtained. Based on literature review, we decided to treat the patient with capecitabine. After three cycles of capecitabine, the patient had clinical and radiological response. Conclusion: Owing to the scarcity of this tumor presentation, more therapeutic options remain to be defined. Oral 5FU seems a reasonable option in this setting. Although the tumor mutation burden is low in this patient, immunotherapy may be a reasonable therapeutic niche.

Keywords: Capecitabine, Multiple recurrence, Next generation sequencing, Skin appendageal carcinoma, Syringoid eccrine carcinoma

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Munaf AL-Kadhimi[1], Tamarah AL-Dawoodi[1], Julio Peguero[1], Luis T. Campos[1]

Affiliations: [1]Oncology Consultants, Research Department, 2130 West Holcombe Blvd. 10th Floor, Houston, Texas.

Corresponding Author: Dr. Julio Peguero, MD, 2130 West Holcombe Blvd. 10th Floor, Houston, Texas 77030; E-mail: jpeguero@oncologyconsultants.com

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INTRODUCTION

Carcinomas of the eccrine sweat gland are unique type of neoplasms which having the capacity to destruct locally and metastasize. They have high recurrence rates after conventional surgical excision. Owing to the rarity of these types of neoplasms, the specific classification of eccrine carcinomas is complex and unclear. In addition, the histologic resemblance of these types of cancers makes it even more difficult to assemble differentiation. The histogenetic association is based primarily on histochemical, immunochemical, next generation sequencing and ultrastructural features.

Eccrine carcinomas of sweat glands can be divided into low grade tumor, intermediate grade tumor and high grade tumor: those histologically similar to certain benign appendage tumors (e.g., sclerosing sweat duct carcinoma, porocarcinoma, malignant chondroid syringoma,

malignant nodular hidradenoma, and malignant eccrine spiradenoma) and those showing diverse histologic features and have no benign counterpart [1].

CASE REPORT

A 65-year-old Siberian female was presented to our facility owing to recurrent spiroadenocarcinoma (Figure 1). The patient had a known history of temporal spiroadenocarcinoma, initially misdiagnosed as a benign tumor in late 2008, when she had undergone resection. The patient had multiple recurrences at the surgical site; status post re-excision followed by three cycles of docetaxel and cisplatin, and enjoyed a disease free period until March 2013. At that time, a second recurrence occurred in the right parotid area and radiation therapy (a total of 60 Gy) was delivered to the surgical site; this was initiated in April 2013 and completed in May 2013. The patient enjoyed a disease-free interval until December 2013 when she underwent re-excision, right parotidectomy, and right neck dissection for again local recurrence.

In October 2014, computed tomography (CT) scan showed bone metastases with recurrent metastatic disease in the parietal scalp area. She was treated with surgical resection. The last recurrence was noted in February 2015, when CT revealed metastatic disease in the right temple, near the right ear, the processus coracoideus of the left shoulder blade, and its lower part.

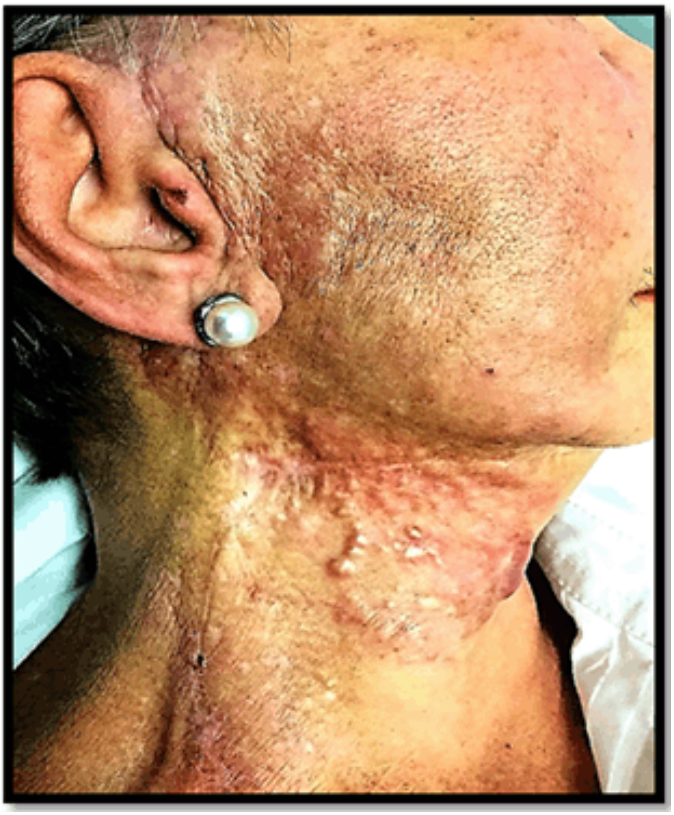

Figure 1: Before initiating capecitabine.

In March 2015, she was treated with surgical resolution, followed by two sessions of photodynamic therapy of the forehead skin. She then underwent positron emission computed tomography scan (PET) scan which demonstrated soft tissue attenuation in the subcutaneous right temporal scalp. There was increased FDG activity in the thyroid diffusely and at the subcutaneous base of the right auricle. A sub-centimeter focus of increased FDG activity was noted at the medial subcutaneous site at the lower right anterior lateral neck. The scan also revealed 1.3x1.6 cm right sub-pectoral node and bilateral sub-centimeter axillary nodes with faint increased FDG activity. Additionally, lytic defects at C7, T4, T5, T7, and T9, and a destructive lesion at the left scapula coracoid process were observed. A core biopsy of the lesion in the soft tissue of the right temple indicated a spiradenocarcinoma.

We discussed the treatment options with the patient, as spiradenocarcinoma is very rare and the number of reported cases in literature is limited. We decided to initiate treatment with capecitabine (three 500 mg tablets, twice daily every 21 days), as it had been reported to be effective in some cases [2]. After three rounds of capecitabine, the patient showed a good clinical (Figure 2) and radiological response.

DISCUSSION

Syringomatous carcinoma a rare malignant cutaneous appendageal tumor deriving from the sweat glands. It was first described by Goldstein et al. in 1982 as a microcystic adnexal carcinoma [1, 3]. The current consensus is that these are different names for the same neoplastic process with different degrees of differentiation.

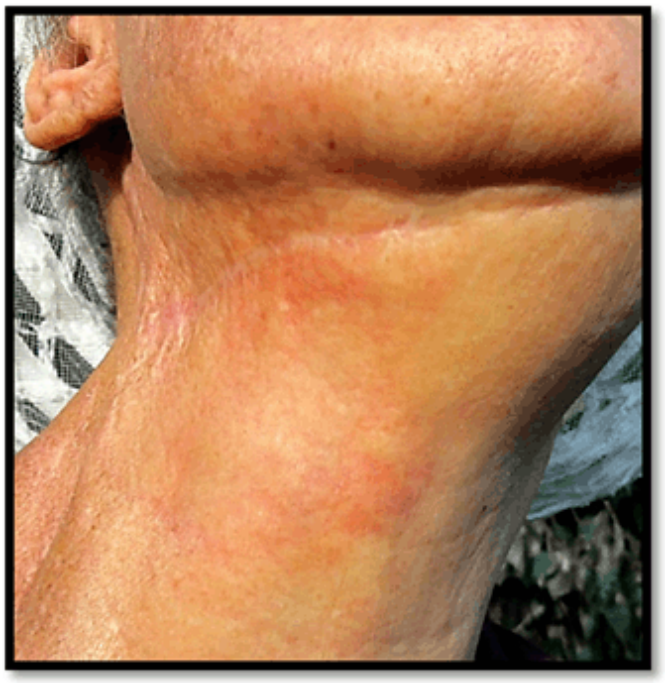

Figure 2: After three cycles of capecitabine.

The median age at the time of presentation is generally between 40 and 50 years (range 20–80 years). Moreover, the frequency in males and females is similar [4]. The most common sites involved are the head and neck with the centrofacial region being most frequently affected site (approximately 85% of cases). The axilla, trunk, and extremities are sites that are less frequently affected [4].

The invasive tumor is locally destructive but lymph node and distant metastases are rare. There are only some publications regarding the characteristics, treatment options, and outcomes of patients with syringomatous carcinoma. Treatment of recurrent metastatic syringomatous carcinoma with capecitabine is one of these publications [2]. In an effort to expand treatment options, the tumor samples were evaluated through next generation sequencing. The genetic profile revealed GATA3 mutation as well as variants of unknown significance, consistent with AURKB, FLT4 GLI1, IDH2, MLL2, NF1, NTRK3, PMS2, TSC1, ZNF217. In addition, the mutation tumor burden was low. However, the patient did not undergo targetable gene therapy.

CONCLUSION

Syringomatous carcinoma is unique invasive, slowly growing, locally destructive adnexal tumor. Owing to locally aggressiveness nature of the tumor and the high recurrence rate may necessitate further sophisticated managements. The best management is to ensure a complete microscopically controlled surgical excision with clear surgical margins. We introduced capecitabine and the patient showed a good clinical response. Owing to the scarcity of this tumor presentation, more therapeutic options remain to be defined. Oral 5FU seems a reasonable option in this setting. Although the tumor mutation burden is low in this patient, immunotherapy may be a reasonable therapeutic niche.

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Author Contributions
Munaf AL-Kadhimi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Tamarah AL-Dawoodi – Substantial contribution to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Julio Peguero – Substantial contribution to conception and design, Analysis and interpretation of data, Final approval of the version to be published
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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Dr. Peguero serves in the speaker bureau for Foundation Medicine, Bristol Myers Squibb and Bayer. He directs phase 1 and 2 clinical trials that are funded by the Pharmaceutical Industry at the Oncology Consultants Research Department.

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