

# Advanced Cutaneous Scalp Eccrine Adenocarcinoma, Diagnosis and Treatment Challenges: A Case Report

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## Abstract

In Madagascar, a country with a tropical climate; skin cancers are often of epidermal or melanic origin. Adnexal tumors are not well known. We report the first case of an eccrine tumor in a 57-year-old woman with no personal or family history of cancer. She has no comorbidities. The patient presented with a 50 × 60 mm frontal swelling and an ulcerative lesion of the left half of the face that had been evolving for 5 months. The cerebral and cervical CT scan revealed bone lysis of the vault of the skull and left cervical lymphadenopathy. The CT chest-abdomen-pelvis showed no abnormal lesions. The mammogram was normal. The result of the pathological examination was in favor of a round-cell tumor. The AE1/AE3, CK7 and P63 markers were positive in the immunohistochemical study. The diagnosis of an advanced eccrine adenocarcinoma was retained after the exclusion of a visceral adenocarcinoma with cutaneous metastasis. The patient receives radiotherapy of 40 Gy in 20 fractions followed by 6 courses of Carboplatin-paclitaxel type chemotherapy. The lesion has decreased in size and dried up after these treatments. 3 months after stopping treatment for lack of financial means, the patient died at home. The diagnosis of an adnexal tumor requires more in-depth complementary examinations. Treatments should be multidisciplinary and adapted to each patient.

## Keywords

Adenocarcinoma, Eccrine, Chemotherapy, Radiotherapy, Scalp Lesions

## 1. Introduction

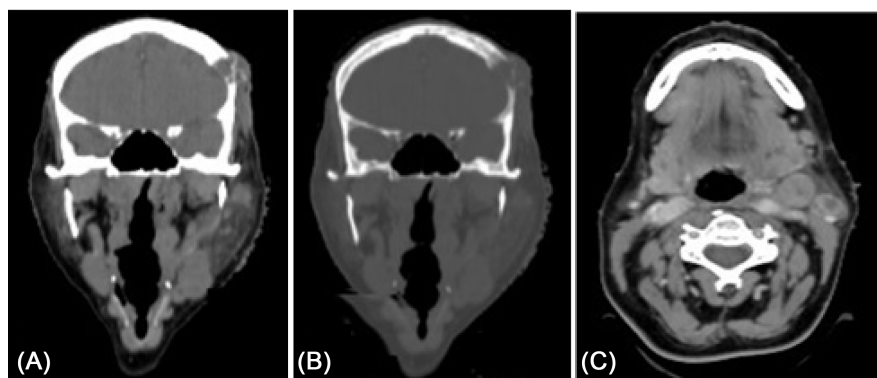
Eccrine adenocarcinoma is a rare skin tumor of the adnexal. The subset of eccrine carcinomas represents less than 0.01% of malignant skin tumors [1]. The most common location is in the lower limbs, the head and neck, and other sites include the face and scalp. Diagnosis is based on immunohistochemical examination and exclusion of visceral adenocarcinoma with cutaneous metastasis. Treatment is essentially surgical. We report a case of the advanced cutaneous scalp and facial eccrine adenocarcinoma with lymph node and bone involvement in the vault of the skull. The patient was treated with radiotherapy and chemotherapy. The objective was to describe the immunohistochemical aspects and discuss the practical management of eccrine adenocarcinoma.

## 2. Case Report

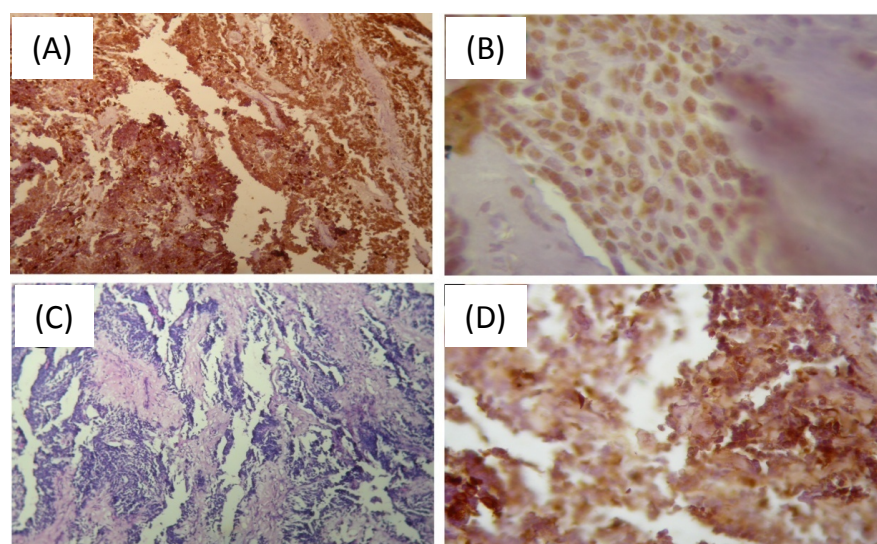
A 57-year-old woman patient was referred for frontal and left face swelling evolving for 5 months. Our patient has no personal or family history of cancer. She has no comorbidities. The physical examination showed a round tumefaction of 50 × 60 mm medio-frontal and weeping ulceration at the level of the left half of the face (Figure 1). The cerebral and cervical CT scan revealed a lesion of the scalp with bone lysis at the level of the vault of the skull, and left cervical adenopathy (Figure 2). The biopsy of a scalp lesion was performed in June 2019. Histological examination was in favor of a round-cell tumour. The immunohistochemical study revealed either a primary cutaneous adenocarcinoma or a cutaneous metastasis of a breast tumor. AE1/AE2, CK7 and P63 markers were positive (Figure 3). CK20, TTF1, CK19, CK5/6, synaptophysin and chromogranin as well as hormone receptors were negative. The mammography performed was normal. The exploration and extension assessment in search of a primary tumour, including a thoraco-abdomino-pelvic CT scan, did not show any suspicious-looking visceral mass or secondary location. Primary eccrine adenocarcinoma was retained as the diagnosis. After a multidisciplinary consultation meeting, the patient had received radiotherapy of 40 Gy delivered in 20 fractions followed by 6 courses of



**Figure 1.** Weeping ulceration of the left half of the face and at the frontal level of 50 × 60 mm.



**Figure 2.** Coronal section of a cerebral CT scan, in parenchymal window (A) and bone window (B) showing a lesion of the left frontal scalp with lysis of the vault of the skull opposite. (C) Axial section of a cervical CT scan showing cervical adenopathies at the level of the left lymph node area III.



**Figure 3.** Immunohistochemistry: (A) AE1/AE3 ×400, (B) P63 ×400, (C) Hematoxylin-eosin ×100, (D) CK7 ×400.

Carboplatin AUC 4-Paclitaxel 175 mg/m<sup>2</sup> type chemotherapy; all were well tolerated overall. The lesion has reduced in size and dried up after these treatments. Unfortunately, the chemotherapy was stopped for pecuniary problems. Three months after stopping treatment, we learned of the death of the patient from a member of her family.

### 3. Discussion

In Madagascar, the most common skin tumors were squamous cell carcinoma followed by melanoma and basal cell carcinoma; adnexal tumors are uncommon [2]. Cutaneous adenocarcinoma starts in the sweat gland. Eccrine adenocarcinoma often affects elderly male patients. The tumor is locally aggressive with a high local recurrence rate estimated at 20% after excision [3]. The clinical symptomatology is non-specific. The tumor appears as a brown nodule, papule or ul-

cerative lesion ranging from 5 to 120 mm. Given the rarity of this disease, little information is available on the place of imaging. A PET scan could help lymph node localization [4].

The diagnosis is based on the exclusion of cutaneous metastases of a visceral tumor and breast. An immunohistochemical examination is necessary to confirm the diagnosis and to differentiate from other adnexal tumors [5]. For our patient, the CK7, P63, and AE1/AE3 were positive. CK 20 negativity helps differentiate from a gastrointestinal tumor. A negative CK5/6 rules-out carcinoma of urothelial and mesothelial origin. The negativity of synaptophysin and chromogranin helps eliminate a neuroendocrine tumor. The negativity of the TTF1 eliminates the pulmonary and thyroid origin. The negativity of the hormone receptors is not in favor of an ovarian or endometrial tumour. The positivity of CK7 and the negativity of CK20 confirmed the eccrine origin of the tumor [6].

The treatment is essentially surgical either by the Mohs technique with a margin of 4 - 5 mm or surgery with a wide margin [5]. In Madagascar, the lack of a technical platform as well as the limited financial means would not allow the realization of this technique. In our case, the difficult anatomical location such as the scalp and the left face, the large extent of the lesion and the bone involvement had prevented a wide surgery.

The stability and management of eccrine carcinoma with combined radiotherapy and chemotherapy has been reported in the literature, especially when surgery is not an option [3] [7] [8]. Marcin *et al.* reported the case of a patient who received chemotherapy combined with CAV (vincristine doxorubicine cyclophosphamide) and EP (etoposide cisplatin) for the metastatic form [9]. Joseph *et al.* reported in response to taxane treatment in a patient with a metastatic eccrine tumor. The patient died after a 14-month follow-up [10]. Other authors such as Christian Larson have reported that the use of capecitabine stabilizes the disease with a progression-free survival period of 18 months and improves quality of life [5]. For our patient, she had received radiotherapy of 40 Gy delivered in 20 fractions. The lesion has decreased in size and dried up after radiotherapy. Six courses of carboplatin-paclitaxel chemotherapy were performed; all were well tolerated overall with a good tumour response. The premature termination of the treatment due to costs leads to rapid disease progression. Our patient died three months after stopping treatment. Currently, new data are encouraging concerning the use of immunotherapy for the treatment of advanced forms. Karla *et al.* reported a case of a metastatic patient having a complete clinical and radiological response to pembrolizumab [11].

#### 4. Conclusion

Primary eccrine cutaneous adenocarcinoma is a locally aggressive tumor with poor prognosis. Immunohistochemical examination and imaging are fundamental to making the diagnosis. It is important to think about aggressive scalp lesions, a quick diagnosis helps to start therapy asap. Radiotherapy and chemotherapy are

therapeutic options that can be used to control the disease in an underdeveloped country like Madagascar.

### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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