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CASE REPORT

# Adenoid cystic carcinoma in atypical site: case report

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## Abstract

Adenoid cystic carcinoma (ACC) is a rare form of malignant neoplasm of the salivary glands, with slow growth and late symptoms. One of the challenges presented by this tumor is its high rate of metastasis, with tropism to the nervous system, liver and bones. Treatment is based on surgical resection of the tumor, radiotherapy, and chemotherapy – it varies depending on the condition of each patient. In view of the low frequency of ACC, this study aims to report a case of ACC in a female patient, with anatomopathological diagnosis of basaloid cell neoplasm.

**Keywords:** adenocarcinoma; neoplasm metastasis; case reports.

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## Introduction

Adenoid cystic carcinoma (ACC) is a rare malignant neoplasm that accounts for 1% of head and neck cancers[1]. ACC is a slow-growing, commonly asymptomatic tumor diagnosed between the 5th and 7th decades of life, with higher prevalence amongst female patients[2,3]. ACC represents approximately 10% of salivary gland cancer cases, and it can occur in several anatomical regions, such as in the lacrimal glands and nasal cavities, as well as in structures outside the head and neck: the esophagus, breasts and lungs[2].

There is often a late diagnosis of ACC, since its submucosal, painless and slow-growing development is usually neglected for a long time, and only perceived when the patient begins to report discomfort due to disease onset or compression of other adjacent structures[4]. The greatest challenge thereof is the silent and aggressive invasion due to its tropism to the nervous system (mainly perineural spaces and sheaths), liver tissue, and bone marrow, where ACC dissemination occurs more easily and less evidently, since radiological findings do not always show the actual tumor progression, which contributes to its high rates of metastasis and recurrence[2]. Thus, several strategies are used to reach an accurate diagnosis and prognosis, such as biopsies and immunohistochemistry (IHC) tests[2].

Histopathologically, ACC is described as cribriform, tubular and/or solid, and its identification is essential to determine the therapeutic options and prognosis of the patient. Regarding this differentiation, the solid subtype is the

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least frequent, associated with high levels of pleomorphism, mitotic activity, metastasis, and perineural invasion, and presents the worst prognosis[1].

Treatment, in most cases, is based on surgical resection of the tumor, radiotherapy, and chemotherapy. One of the most significant difficulties found is that, even with a successful surgery and radiotherapy treatment, tumor recurrence and development of distant metastases commonly occur[3].

Therefore, this study aims to contribute to the scientific community by presenting a case of a solid ACC in an unusual site of origin in a female patient, with anatomopathological diagnosis of basaloid cell neoplasm.

## Case report

Female patient, 54 years old, alcoholic, and crack cocaine user, living on the streets, complaining of a tumor on the left cervical base, with onset approximately seven years ago, associated with moderate to severe pain. The patient reported having undergone a biopsy on 22 December 2020 at the Hospital Aristides Maltez, but she did not undergo follow-up so that the anatomopathological results could be obtained and did not attend the two surgeries scheduled at the aforementioned Institution.

In 2021, she sought care by her own means at the Irmã Dulce Social Works for medical assessment reporting worsening of pain and the desire to be operated. Upon physical examination, it was confirmed that the patient had a granular cell tumor, with an ulcerated center, areas of telangiectasia at the edges of the lesion, and areas with necrotic foci (~10x7 cm) on the left side of the neck (Figure 1).

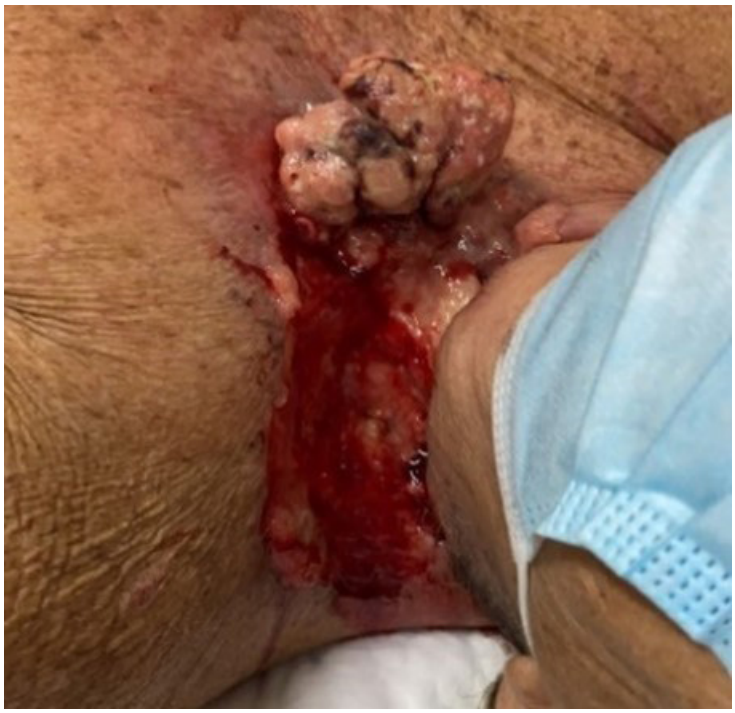

**Figure 1.** Clinical examination: tumor lesion in November 2021.

Following a chronological logic, the anatomopathological result of 24 December 2020 evidenced a basaloid cell neoplasm and the IHC test identified it as ACC (Tables 1 and 2).

**Table 1.** Anatomopathological study of December 2020 showing basaloid cells.

| ANATOMOPATHOLOGICAL STUDY |
|---|
| **MACROSCOPY**<br>Received in formalin, 03 tissue fragment (s) measuring 1.0x0.8x0.6 cm.<br>Secretions: 3/1c (all materials included) |
| **MICROSCOPY**<br>Secretions show a neoplasm consisting of basaloid cells amid fibrous and myxoid tissue. Fragment of mitosis figures can be seen. |
| **CONCLUSION**<br>Skin:<br>- BASALOID CELL NEOPLASIA |

**Table 2.** Immunohistochemical panel of January 2021 indicating a solid ACC.

| IMMUNOHISTOCHEMICAL REPORT | | |
|---|---|---|
| **Method**: Immunohistochemistry on paraffin-embedded tissue; EnVision FLEX+ System, High pH Link, manual, wet heat antigen retrieval in stages according to the manufacturer's specifications – DAKO. | | |
| **ANTIBODIES USED** | | |
| Bcl2 (124) S | CEA (mono-11) | P-53 (DO-7) |
| CD117 (YR145) | EMA (E29) | P63 (4A4) |
| Kl67 (SP6) | Actin (1A4) | BerEp4 |
| CK (AE1AE3) | S-100 | Calponin (CALP) |
| **MICROSCOPIC DESCRIPTION**<br>Morphological and immunohistochemical aspects favor the diagnosis of adenoid cystic carcinoma with expression for AE1, AE3, and BCL2. Focally positive for EMA, p63, and p53. Positive myoepithelial cell markers (calponin, actin, and S100). Kl67 is about 30%. All others were negative (CD117, BerEP4, and CEA). | | |
| **CONCLUSION**<br>Skin:<br>- MORPHOLOGICAL AND IMMUNOHISTOCHEMICAL ASPECTS FAVOR THE DIAGNOSIS OF CYSTIC ADENOID CARCINOMA. | | |

Among the previously requested medical exams, laboratory tests (27 Apr 2021) showed a reduction in the level of hemoglobin (8.3 g/dL; RV: 12.5-15.7 g/dL) and hematocrits (28.4%; RV: 36-48%), and normal levels of platelets, prothrombin time (PT), partial thrombin time (PTT), international normalized ratio (INR), urea, and creatinine. As for the imaging tests, the chest X-ray (27 Apr 2021) showed a small, calcified, residual nodule in the left the supra- and infra-clavicular regions, while the chest computed tomography (CT) (30 Apr 2022) evidenced a soft-tissue, lobulated nodule (5.0x4.2x3.6 cm) on the left side of the cervical spine, extending to the outer wall of the hemithorax on the same side, with thickening of the adjacent subcutaneous tissue.

In addition, the CT of the neck performed on 28 April 2021 showed an expansive vegetative formation with soft tissue density located to the left of the cervical-thoracic region, hypervascularized, infiltrating the subcutaneous tissue and myoadipose planes, attached to the left thyroid lobe and the sternocleidomastoid muscle, maintaining broad contact with the left jugular vein, measuring about 3.5 x 5.9 x 4.7 cm (AP x LL x CC). The CT scan also showed the coexistence of thickening of the left platysma muscle, an enlarged level IV lymph node (1.6x0.9 cm) on the left side, and salivary glands with no changes in symmetry, dimension, diffusion, and attenuation coefficients – indicating a primary ACC.

Surgical resection of the lesion was recommended, with ipsilateral radical debulking and reconstruction with a deltopectoral flap or pectoralis major myocutaneous flap, and scheduled for 29 Sep 2021, but the patient did not attend the appointment and did not continue with the follow-up despite the attempts to contact the patient made by the head and neck surgery team.

In December 2021, surgery was no longer a therapeutic possibility to treat the lesion. Due to the patient's non-adherence to treatment, it was decided to refer her to the clinical oncology and radiotherapy service; however, she has not appeared for evaluation so far.

## Discussion

Deviating from classical cases[1], the tumor described herein did not appear covered by intact mucosa, presenting granulation tissue in the ulcerated lesion, along with areas with telangiectasia and necrotic foci on the left side of the cervical-thoracic region, maintaining wide contact with adjacent structures[1]. Furthermore, there was a well-defined lobulated nodule, in a site of origin different from that usually seen clinically – in the left cervical region to the ipsilateral hemithorax.

In addition, this patient had a nodule and ulcer dissociated to the topography of the salivary glands (a region with no alterations by the complementary exams). ACC lesions can develop in a variety of anatomical regions; however, they are most frequently found in the major and minor salivary glands, lacrimal glands and, more rarely, in glands of the upper aerodigestive tract[2].

The IHC analysis identified an ACC in the referred patient, and the histopathology of the case was not obtained, the latter being an important information for the prognosis[2]. In this case report, it was possible to observe extensive infiltration into adjacent areas, wide contact with the thyroid and necrotic areas, suggesting a diagnosis of solid ACC. Other important factors may be associated with this subtype, such as the close contact with the jugular vein and the presence of enlarged lymph nodes along nearly the entire bilateral cervical extension - conditions that do facilitate and indicate the process of dissemination of neoplastic tumor cells.

Regarding the ACC-related metastases, approxiamtely 10% of patients developed cervical metastasis and 8.5% presented distant metastasis at the time of diagnosis[4]. This was also accompanied by high rates of recurrence and low survival[2]. Min et al.[5] reported that life expectancy was directly related to the presence of metastases – the overall 5-year survival rates of patients with and without lymph node metastasis were 48% and 77%, respectively.

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The initial treatment for this carcinoma is already well established and consists of surgery for resection of the adjunct lesion with ipsilateral radical debulking. This intervention was indicated by the professionals who provided care to this case report, despite not being effective on account of the patient's evasion.

It is important to emphasize that the condition this patient was already critical, with advanced symptoms. Several factors can be considered when assessing the swift evolution of this case, including the patient's living conditions: unstructured, without housing, hygiene and basic care, in conjunction with issues such as the excessive use of alcohol and crack cocaine. Likewise, the psycho-emotional factor was yet another obstacle to the treatment, since the patient reported that she abandoned the medical follow-up and neglected the of surgical treatment possibilities as a result of fear and psychosocial trauma.

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