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Case Report

Neuroendocrine Carcinoma of Oral Cavity: A Case Report of an Unusual Presentation

Imane Mbarki[1]*, Salma El Baz[1], Hamdan Asmae[1], Oumaima Eddarif[1], Sanaa Elmajjaoui[1], Hanan Elkacemi[1], Tayeb Kebdani[1] and Noureddine Benjaafar[1]

[1]Department of Radiation Oncology, National Institute of Oncology, Mohammed V University, Rabat, Morocco

[*]**Corresponding Author:** Imane Mbarki, Department of Radiation Oncology, National Institute of Oncology, Mohammed V University, Rabat, Morocco.

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Abstract

Neuroendocrine carcinoma (NEC) are tumors affecting lungs in the first line. Extra-pulmonary locations are rare, and involvement of the oral cavity is uncommon. The therapeutic strategy of this clinically aggressive entity is not codified. Nevertheless, multimodal treatment combining surgery and radio-chemotherapy is associated with the best results in terms of local control and overall survival. We report a case of a 67-year-old patient, diagnosed with a NEC, the computed tomography (CT) of the facial mass objectified a jugal mass of 2x2.4 cm with two lymph nodes. The patient benefited from an excisional biopsy of the mass, and whose histological examination and immunohistochemical testing returned in favor of a small cell neuroendocrine carcinoma.

Keywords: Neuroendocrine carcinoma; Extra-pulmonary; Oral cavity; Histology; Surgery; Prognosis.

Introduction

Neuroendocrine tumors are a heterogeneous tumor entity, which can occur in any location in the body. Pulmonary and digestive locations are the most frequent. In the head and neck region, NECs most often originate from the larynx, then the salivary glands and the sinonasal region [1]. The oral cavity rarely represents the primary site of NECs [2].

This present report is an exceptional case of NECs of the oral cavity in a patient of 67-years-old, having a neoadjuvant chemotherapy, and radiotherapy.

Case presentation

This is a 67-year-old female patient, with no notable history, who has a left jugal mass

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evolving for five months, without any other associated sign. A CT of the facial massif showed a jugular mass of 2x2.4 cm in contact with the master muscle, with two lymph nodes of eight and nine mm. The patient underwent an excisional biopsy of the mass. The anatomopathological examination objectified a muscular tissue largely infiltrated by a proliferation made up of sheets of basophilic cells with invisible cytoplasm, and hyperchromatic nucleus of neuroendocrine appearance. Immunohistochemical complement showed that tumor cells are positive for CD56 and synaptophysin, and negative for chromogranin A, cytokeratin (AE1/AE3), and CK-20. Therefore, the diagnosis of a small cell neuroendocrine carcinoma was retained. The patient was lost to follow-up for six months.

Faced with the reappearance of the left jugal mass, the patient consulted again. The initial examination showed a left jugal mass, of elastic and mobile consistency, the mucous membrane opposite is of normal appearance. Cervical palpation revealed two left latero-cervical lymphadenopathy of approximately one cm.

The CT of the facial mass revealed a mass of three cm at the level of the tumor bed, with two lymphadenopathies of nine and ten mm. No secondary localization on a thoraco-abdomino-pelvic CT scan.

Carcinological surgery was proposed, which the patient refused.

The patient received three courses of chemotherapy with etoposide-cisplatin. An evaluation CT scan does not show a tumor residue with a complete response.

The patient received radiotherapy using the VMAT technique, with Simultaneous-integrated boost on the tumor bed at a dose of 60 Gy (2Gy/fraction), and the ipsilateral lymph node areas at a dose of 54 Gy (1.8Gy/fraction). The treatment was daily in five fractions per week. The treatment passed without incidence with good tolerance.

After a follow-up of four months, the clinical examination did not show signs of loco-regional recurrence or distant metastasis.

Discussion

NECs are a heterogeneous group of tumors characterized a wide range of clinical manifestations, tissue origin, and clinical aggressiveness. According to the 2017 World Health Organization classification of Head and Neck tumors, NECs are subclassified into three categories: well-differentiated (WD-NEC), moderately differentiated (MD-NEC), and poorly differentiated (PD-NEC) with small cell and large cell types [3]. This classification incorporates elements of differentiation and grading, and the most closely correlates to 5-years disease specific survival of 100, 52.8, 19.3 and 15.3% for each diagnostic category [3].

WD-NECs are generally unrelated to alcohol-smoking intoxication [4], paraneoplastic syndrome is rarely observed, these tumors are slowly progressive [5], most often asymptomatic. In case of MD-NECs, most patients have a history of alcohol and tobacco poisoning, paraneoplastic syndrome is rare [6], these tumors are more locally aggressive, potentially affecting the uvula, the gingiva or the floor of the mouth. PD-NECs are observed in alcohol-smoking patients, taking on the

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appearance of a rapidly progressive, painful tumor, lymph node metastases are frequent [4].

The diagnosis confirmation is histological and immunohistochemical. The tumor is formed by anaplasic cells arranged in cords, sheets or irregular nests. NECs of the oral cavity are very rare tumors, not seen routinely, and therefore microscopic examination is insufficient to establish the diagnosis. The immunohistochemical complement highlight the neuroendocrine origin of the tumor. Neuroendocrine markers usually used are Neuron-specific enolase (NSE), chromogranin A, synaptophysin, S100, CD56, and CD57 [7]. NECs expresses at least one neuroendocrine marker.

Positron emission tomography (PET), computed tomography imaging (CT), and magnetic resonance imaging (MRI) are necessary to evaluate the locoregional extension of the tumor, the invasion of the adjacent organs, and the presence of distant metastases [8].

As for NECs in other locations, surgery of primary tumor in oral cavity is the angular piece of treatment, which significantly improves overall survival [9,10]. Surgical excision of the tumor with cancer-free margins is the standard. The surgery must be complete, conservative and not mutilating.

Due to the low risk of lymph node metastases and favorable biological behavior of WD-NECs, lymph node dissection is not indicated [6], and no adjuvant treatment is recommended. For MD-NECs and PD-NECs, lymph node dissection is recommended, even in case of clinical No necks.

MD-NECs are radio-resistant and chemo-resistant tumors, and therefore the indication of adjuvant radiation chemotherapy is not retained [11,12], it did not improve significantly survival rate.

PD-NECs are aggressive tumors, with a high risk of locoregional recurrence and metastases, approximately 60% to 90% of patients have possible distant metastasis to lung, liver, and bones at the time of diagnosis [13,14], and therefore the therapeutic strategy combining in addition to surgery, radiotherapy and chemotherapy. The chemotherapy protocol is modeled on that of extra-pulmonary localization including etoposide and cisplatin [15,16]. However, many patients are in the disseminated stage at the time of diagnosis, and thus surgery cannot be considered, only radiotherapy and chemotherapy are advocated [17].

For patients in poor general condition, and for whom surgery may be adverse, or refusing surgery, the therapeutic strategy only includes radiotherapy with/without chemotherapy. In their article, yoshida et al, treated their patient with radiotherapy alone [18], while khurana et al, treated their patient with combined radiotherapy [19]. All three patients had complete tumor remission in both arms, but with severe toxicities. The most important factor of poor prognosis is the poor response to chemotherapy [20]. Tumor localization can also influence the prognosis. Hautom et al, suggested in a small series that salivary gland NECs have a better overall median survival (30 months) compared to other locations of the head and neck including the oral cavity (15.2 months) [21].

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Conclusion

NECs of the oral cavity are exceptional tumors. The definitive diagnosis is histological with an immunohistochemical complement, which highlights the neuroendocrine markers. An exhaustive assessment of extension is necessary to exclude distant metastases. The therapeutic strategy is not clear due to the rarity of this entity. Surgery is the optimal treatment. Radiotherapy and chemotherapy may be indicated as an adjuvant treatment or in patients not fit for major surgery. Prognostic factors are not well established, NECs of the head and neck continue to have a poor prognosis.

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Conflicts of interest

The authors declare that they have no conflicts of interest in relation to this article.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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