Carcinoma neuroendocrine de célula pequeña en la amígdala. Un caso clínico

Small Cell neuroendocrine carcinoma of the tonsil. A case report

Arsénio Barbosa[1], Miguel Relvas[2], Clara Borges[3], Margarida Sá-Fernandes[4]

[1]Department of Internal Medicine, [2]Department of Nephrology, [3]Department of Oncology, [4]Department of Anatomic Pathology. Centro Hospitalar São João, Porto, Portugal

ABSTRACT

Primary small neuroendocrine carcinoma (SNEC) of the tonsil it's a rarity.

We present the case of a 64 year-old Caucasian male patient referred to our hospital due to a neck mass. A fixed non-tender rock hard supraclavicular right mass and ipsilateral cervical lymphadenopathy was palpable. Biopsy was performed and showed poor differentiated neoplasm with features of SNEC. A fludeoxyglucose positron emission tomography (FDG-PET) scan showed intense uptake in the right tonsil. The patient underwent right tonsillectomy and immunochemistry was positive for characteristic features of small cell neuroendocrine carcinoma. Right tonsil SNEC was diagnosed and the patient was proposed to chemotherapy.**Keywords:** Neuroendocrine carcinoma, neck tumor, tonsil, immunohistochemistry

INTRODUCTION

Neuroendocrine tumors are a heterogeneous group of neoplasms that differ from each other according to their biological behavior, histology and morphology and may be well or poorly differentiated, leading to different responses to treatment. Lungs are the most common sites for neuroendocrine tumors, although it has been reported cases in many organs, such as gastrointestinal and genitourinary tract, breast, head and neck and unknown primary origin. Extrapulmonary small cell neuroendocrine carcinomas (SNECS) accounts for 2,5-5% of cases[1,2,3].

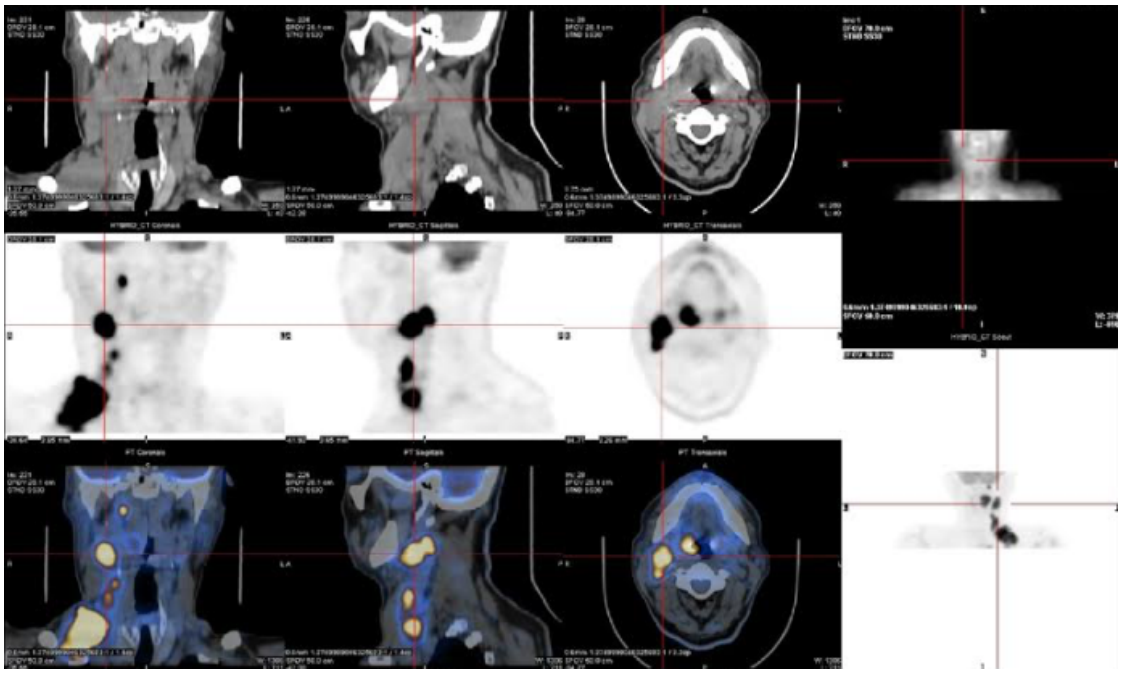
While tonsil is one of the most common site for head and neck tumor, primary SNEC of the tonsil it's a rarity[4]. Squamous cell carcinoma, minor salivary gland tumors, lymphomas, melanoma and sarcomas are also reported[2].

CASE REPORT

A 64 year-old caucasian male was admitted in the emergency department due to a neck mass. The patient had been in his usual state of health until one month before admission, when he noticed unpainful supraclavicular right neck mass. He did not present fever, hoarseness, dysphagia, dyspnea or otalgia. His medial history included hypertension, type 2 diabetes using insulin without target organ damage, atrial fibrillation on anticoagulation and a 30 pack-year history of smoking.

The physical exam reveals temperature of 36.5ºC, blood pressure 117/76 mmHg, pulse 95 beats per minute and respiratory rate 16 breaths per minute. The oxygen saturation was 96%.

Figure 1. FDG-PET scan with radiopharmaceutical uptake in the right tonsil suggesting primary tumor

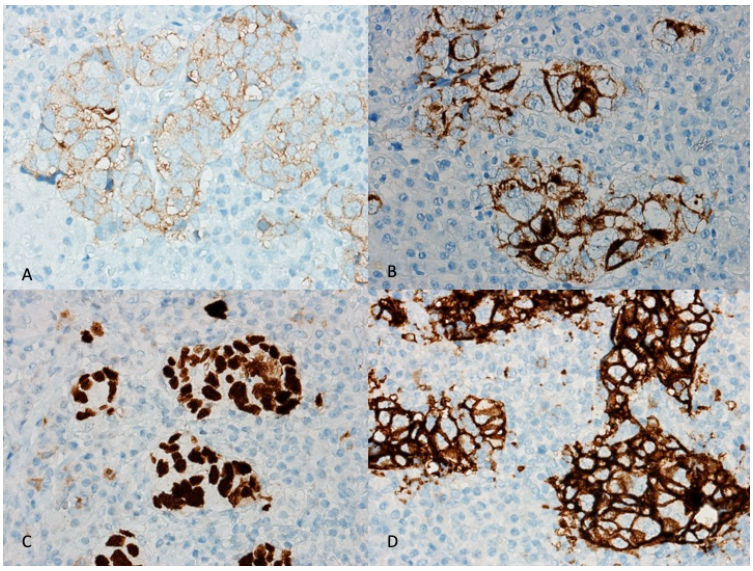


Correspondencia: arseniomiguelbarbosa@gmail.com

Cómo citar este artículo: Barbosa A, Relvas M, Borges C, Sá-Fernandes M
Small Cell neuroendocrine carcinoma of the tonsil. A case report. Galicia Clin 2021; 82-3: 171-173

Recibido: 13/07/2020; Aceptado: 05/03/2021 // https://doi.org/10.22546/62/2349

Figure 2. Histopathologic staining: A) positivity for synaptophysin; B) positivity for CK8/18; C) positivity for TTF-1; D) negativity for neural cell adhesion molecule (CD56)



The neck examination showed a fixed, non-tender, stony supraclavicular right mass and ipsilateral cervical lymphadenopathy. Thyroid gland palpation was normal. Otolaryngologist for head and neck examination evaluated the patient, which was normal besides the cervical mass.

Blood test that was obtained:

- Cell blood count showed only mild normochromic normocytic anemia (hemoglobin was 12,1 g/dL);
- Acute phase reactants, proteinogram, lactate dehydrogenase, serum calcium, renal and thyroid function and liver panel tests were normal. Human immunodeficiency virus (HIV) serology was negative.

Posteroanterior radiograph of the chest was normal. An electrocardiogram showed the knowed atrial fibrillation.

Biopsy of the neck mass reveled an invasive poorly differentiated neoplasm; the tumor cells had scant cytoplasm and irregular nuclei with fine granular chromatin with overexpression of synaptophysin and thyroid transcription factor 1 (TTF-1), characteristic features of small cell neuroendocrine carcinoma.

In order to clarify the primary tumor, a body-computed tomography (CT) scan revealed a right neck conglomerate lymphadenopathy (6,8 x 4 cm) and cervical and mediastinal lymphadenopathy (Figure 1); and rectal wall thickness. Rectosigmoidoscopy with biopsies was normal. Esophagogastroduodenoscopy was also normal. To identify the local of the primary tumor, a fludeoxyglucose positron emission tomography (FDG-PET) scan was performed and showed intense radiopharmaceutical uptake in the right tonsil suggesting primary tumor of this site (Figure 1). The patient underwent right tonsillectomy.

Histopathologic review of the lesion confirms the previous diagnose and immunochemistry was positive for synaptophysin (Figure 2A), cytokeratin (CK) 7, CK8/18 (Figure 2B), TTF-1 (Figure 2C) and neural cell adhesion molecule (CD56) (Figure 2D) and negative for CK20. The patient was diagnosed with neuroendocrine small cell carcinoma of the right tonsil. He underwent multidisciplinary evaluation and began palliative chemotherapy with carboplatin AUC 5 and etoposide, in an outpatient basis. There were no grade 3-4 toxicities. The patient presented a partial response in clinical and radiologic post-treatment evaluation and he is currently being evaluated to radiotherapy treatment to cervical and mediastinal lymphadenopathies. He is followed in an outpatient basis 12 months after the diagnosis.

CONCLUSIONS

SNEC is a malignant epithelial neoplasm with neuroendocrine morphology. SNECs are poorly differentiated and associated with poor prognosis due to its aggressive cell line[1,2].

SNEC of the tonsil is more common in males (2:1)[2,3,4] and occurs most often between the 5th and 7th decades[2]. Typically, the patient presents with a painful neck mass with symptoms of dysphagia, hoarseness and obstructive sleep apnea syndrome. (4) Although paraneoplastic syndromes including syndrome of inappropriate antidiuretic hormone secretion, Cushing's syndrome and Eaton-Lambert myasthenic syndrome have been associated with head and neck SNEC, there are no reports of these syndromes being associated with SNEC of the tonsil[1,2]. Besides the locoregional spread to cervical lymph nodes, the tumor metastasizes most commonly to liver, lungs, bones, brain and skin[2].

Image detection plays an important role in the diagnosis of head and neck lesions, particularly in the exclusion of regional invasion or distant metastasis[3]. Computed tomography (CT) and/or magnetic resonance imaging (MRI) are useful to evaluate tumor size and maybe metastasis[2]. PET is

gaining value not only for diagnosis, but also to planning and evaluates the effects of treatment[3].

Nuclear chromatin, scant cytoplasm and mitotic figures are hallmarks of SNECs on light microscopy[2]. Immunochemistry it's useful not only to confirm the neuroendocrine differentiation, but also to identify the site of origin[5].

The most common biomarkers used to assess neuroendocrine differentiation include chromogranin A (greater specificity) and synaptophysin (greater sensitivity) and CD56[5].

After confirmation of neuroendocrine differentiation, a panel of epithelial biomarkers is recommended to exclude paraganglioma, since it may change management and treatment. Immunohistochemical expression for CK 8/18 and negativity for CD20 sustains epithelial nature. After confirmation of neuroendocrine and epithelial differentiation, the next step is to assess tumor proliferation in order to classify as well or poorly differentiated neuroendocrine tumor, using the Ki-67 index[5].

Recommendations for management of SNECs have not been established due to its rarity[4]. The approach of tonsil SNECs is based on SNECs of larynx and lungs treatment regimens. Various modalities have been indicated such as: surgical resection, radiotherapy, chemotherapy or a combination of these modalities. Chemotherapy should be considered in all patients with SNECs of head and neck by reason of its propensity for early metastasis[2,4]. Platinum-based chemotherapy is the most common used regimen. Despite multimodality treatment, SNECs of the tonsil have an aggressive disease course and poor prognosis[2,4].

REFERENCES

1. Sehdev A, Zhao Y, Singh A, Sharma N; Primary Cell Carcinoma of the Tonsil: A Case Report and a Review of the Literature; Case Reports in Oncology. 2012;5:537-541.
2. Seen R, Bhutani N, Saini R, Jashanpreet; Primary Small Cell Carcinoma of the Tonsil: An Extreme Rarity; Annals of Medicine and Surgery. 2019; 44: 29-32.
3. Wang X, Chen Q, Meng J; Successful Cisplatin-Etoposide Chemotherapy Based Treatment of a Primary Small Cell Neuroendocrine Carcinoma of the Tonsil with Multiple Metastases: A Case Report; OncoTargets and Therapy. 2018: 11 5391-5395.
4. Wang H, Zou J, Zhou G, Yan J, Liu S; Primary Small Cell Neuroendocrine Carcinoma of the Tonsil: A Case Report and a Review of the Literature; Internation Journal of Clinical and Experimenting Pathology. 2014; 7(5):2678-2682.
5. Duan K, Mete O; Algorithmic Approach to Neuroendocrine Tumors in Targeted Biopsies: Pratical Aplications of Immunohistochemical Markers; Cancer Citopathology. 2016; 124:871-84.