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(CASE REPORT)

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Small cell neuroendocrine carcinoma of the larynx: A case report

F. El Gharroudi *, M.Alahyane, S. Rafi, G. El Mghari and N. El Ansari

Department of Endocrinology, Diabetology, Metabolic Diseases and Nutrition, Mohammed VI University Hospital of Marrakesh, Morocco.

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Abstract

We report a case of small cell neuroendocrine carcinoma of laryngeal localization. The involvement of the larynx still exceptional. We present in this paper the radiological, pathological and therapeutic aspects of this tumor that was diagnosed in a 48-year-old man.

Keywords: Larynx; Small cell neuroendocrine tumor; Endoscopy; Radiology; Pathology; Treatment

1. Introduction

Neuroendocrine tumor of the larynx is a rare entity still it is the second most common tumor of the larynx [1]. Extra pulmonary neuro-endocrine tumor (NET) is a relatively rare disease and larynx is the commonest site in head and neck region. NET consists of 0.5– 1% of epithelial carcinoma of larynx, arytenoid, aryepiglottic fold and epiglottis are the usual site [2,3]. Neuro-endocrine tumor mainly affect males, female to male ratio is 1:3 and smoking is the proven risk factor [2].

They constitute a group of heterogeneous tumors, numbering 5 according to the WHO 2005 classification. Among the subtypes are small cell carcinomas, which have been mainly described in pulmonary tumor pathology. In fact, the primary extra-lung location represents less than 5%, which explains the rarity of publications on similar cases. Small cell carcinoma is known to be radio- and chemo-sensitive, however, with a frequent locoregional and metastatic evolution and a low 5-year survival rate [2].

2. Case report

A 59 years old male patient, chronic smoker, with 10 packs per year; without any particular medical history. he consulted for a rapidly progressive cervical swelling since a year, associated with dysphonia. In a context of apyrexia and alteration of the general state (asthenia, weight loss considered very important not quantified with a preserved appetite). There was no dysphagia, no dyspnea, no carcinoid syndrome or other paraneoplastic syndrome. The patient consulted an ear nose and throat physician, who ordered a cervico-thoracic computed tomography (CT scan) [figure 1,2], showing thickening of the right vocal cord with two nodular lesions measuring 8*10 mm for the largest one, with multiple cervical adenopathies visible in the jugular carotid artery, measuring 23 mm on the right and 15 mm on the left, with necrotic content. Rigid laryngoscopy under sedation revealed a regular submucosal thickening of the right vocal cord extending superiorly.

* Corresponding author: F. El Gharroudi

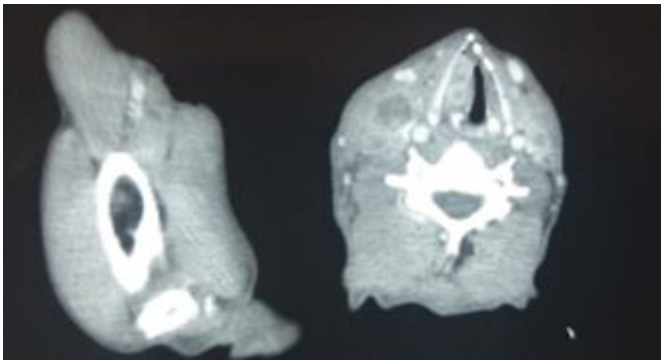

Figure 1 Cervico-thoracic CT scan axial plan

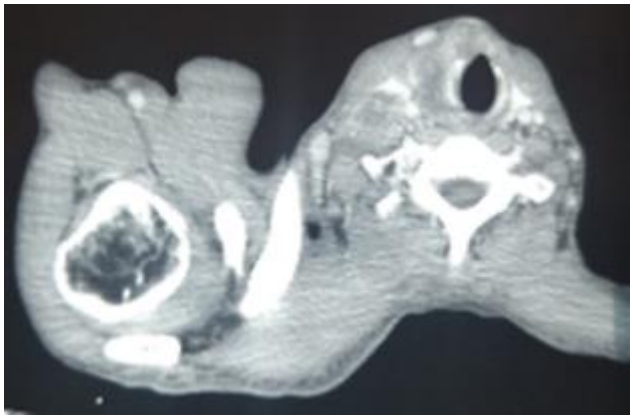

Figure 2 Cervico-thoracic CT scan axial plan

The anatomopathological study of the biopsy showed: an undifferentiated carcinoma, largely necrotic, infiltrating the fibrous tissue with vascular emboli at the level of the labelled specimen. A second specimen of the right vocal cord discreetly reworked by fibrosis without neoplastic localization at this level is associated. This morphological aspect must make us eliminate an aerodigestive primary. With immunohistochemical complement morphological aspect and immunohistochemical profile in favor of an infiltrating small cell neuroendocrine carcinoma (anti AE1/AE3: diffuse positivity / anti chromogranin A: diffuse and intense positivity / anti synaptophysin: diffuse positivity of moderate intensity / anti -KI67: estimated at 60%). Complementary tests showed a very high carcinoembryonic antigen level of 156.4ng/ml, i.e. 26 times normal. The tumor extension assessment showed secondary iliac bone lesion. The octreoscan was not performed due to lack of reagent. Given the histological nature of the tumor which contraindicates surgical treatment, the case was discussed with the oncologists with the decision to stop the investigations and to refer the patient for urgent radio-chemotherapy.

3. Discussion

Extrapulmonary neuroendocrine small cell carcinoma is a relatively rare disease, with the larynx the most frequently affected organ in the head and neck. They can occur in any region of the larynx but the supraglottic is the most commonly reported site [5]. Laryngeal neuroendocrine neoplasms (LNN) have been recognized as the most common non squamous types of neoplasms arising in this area. They account for less than 1 % of all laryngeal neoplasms. To date, more than 700 cases of LNN have been reported in the literature and approximately 500 publications deal with this relatively uncommon yet intriguing family of laryngeal tumors [6, 7]. The atypical carcinoid tumor is the most frequent of all LNN, followed by the small cell neuroendocrine carcinoma, paraganglioma, and the typical carcinoid. Neuroendocrine tumors are divided in to neural and epithelial based on the tissue of origin. Epithelial category consists of 3 types:

- typical carcinoid (well differentiated neuroendocrine carcinoma, Grade I),

- atypical carcinoid (moderately differentiated neuroendocrine carcinoma, Grade II; large cell neuroendocrine carcinoma)
- small cell neuroendocrine carcinoma (poorly differentiated neuroendocrine carcinoma, Grade III).

Neural category consists of paraganglioma [8].Small cell carcinoma represents less than 0.5% of all malignant laryngeal tumors [9]. Anatomopathological examination must always be coupled with immunohistochemistry to confirm the diagnosis. This tumor expresses some neuroendocrine markers such as neuron-specific enolase (NSE), CD56, chromogranin and synaptophysin [10]. Other markers may also be expressed by small cell carcinoma such as cytokeratin, epithelial membrane antigen (EMA), Ber-ep4, and epithelial related antigen (ERA) [11].

They are more common in male . the common age of presentation ranges is from 50 to 70 years and about two third of them are smokers [4]. No clinical symptoms are characteristics of the condition and depends upon the site and extent of the disease. The most common symptoms are hoarseness, dysphagia, odynophagia, dyspnea and hemoptysis. Neuroendocrine tumors are associated with paraneoplastic syndrome and such association should be thoroughly investigated as it changes the management plan. The possible symptoms of paraneoplastic syndrome are headache, confusion, hyponatremia, reduced hematocrit level. Syndrome of inappropriate secretion of antidiuretic hormone (SIADH), myasthenia syndrome, ectopic adrenocorticotropic hormone (ACTH) syndrome are some of the described condition [4].

The treatment of small cell neuroendocrine tumors remains controversial given the rarity of the tumors and the lack of studies. Small cell carcinoma represents a surgical contraindication given the severity of its prognosis and its metastatic potential [12]. The reference radio chemotherapy treatment includes the combination of etoposide and cisplatin as in small cell bronchial cancers.

The survival time is 16% at 2 years and 5% at 5 years. However, prophylactic brain irradiation does not seem to be necessary as in small cell carcinoma of the lung, brain metastases being rarer [13].

4. Conclusion

Small cell neuroendocrine carcinomas of the larynx are extremely rare. An early diagnosis is the most important thing, considering the aggressiveness of the tumor and its poor prognosis and the frequency of distant metastasis comparing it with squamous cell carcinomas. The treatment is based on the combination of chemo-radiotherapy.

Compliance with ethical standards

Acknowledgments

I thank all the authors of this article.

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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