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CASE REPORT

Laryngeal neuroendocrine tumor

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Abstract

Generally rare, neuroendocrine tumors of the larynx are more frequent in the supraglottic region. They predominate in males and develop around 50 years of age. They are divided into typical carcinoid, atypical carcinoid, small cell neuroendocrine tumors and paraganglioma. A 52-year-old male patient, former smoker, complained of a foreign body sensation in his throat for three years, in addition to local pain. A laryngoscopy revealed a polypoid lesion in the right arytenoid. The diagnostic approach adopted was laryngeal microsurgery for excisional biopsy, and the anatomopathological analysis showed an atypical carcinoid. The patient underwent adjuvant chemotherapy; evidence of recurrence was not found after five years.

Keywords: carcinoma; neuroendocrine tumor; head and neck neoplasms; laryngeal neoplasms; larynx.

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Introduction

The first case of neuroendocrine laryngeal tumor was described in 1969[1], and more than 700 cases of this neoplasia have been reported in the literature since. Neuroendocrine tumors of the larynx represent 0.5 to 1% of laryngeal epithelial cancers, occurring more frequently in the supraglottic region. They have male predominance and develop around 50 years of age[2]. They are believed to originate from pluripotent cells of the laryngeal submucosa.

The WHO divided this type of tumor into 4 subgroups: typical carcinoid, atypical carcinoid, small cell neuroendocrine and paraganglioma. A separate entity has been identified within atypical carcinoids, the large cell carcinoma. Typical carcinoids have an indolent course, with a five-year survival rate of 50%. However, atypical cell carcinoids and other neuroendocrine tumors are responsible for aggressive or metastatic tumors, presenting a survival rate lower than 50% in 5 years. Small cell neuroendocrine tumors are particularly aggressive, with a five-year survival rate of 5-10%. Paragangliomas are rarely metastatic. These subtypes can overlap clinically and histologically, making stratification into subtypes difficult; however, it is important to conduct this analysis, as they present a wide range of biological behavior[3].

Case report

A 52-year-old male patient, ex-smoker (stopped for three years, smoked one pack a day), complained of a foreign body sensation with stabbing pain in his

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throat for three years. A direct laryngoscopy (Figure 1) found a polypoid lesion in the right arytenoid measuring about 4 mm. The initial diagnostic hypothesis was papilloma. The diagnostic approach was conducted with laryngeal microsurgery for excisional biopsy. The anatomopathological study showed atypical carcinoid (Figure 2a) with nuclear pleomorphism with evident nucleoli, typical mitoses, combined growth pattern (papillary-like, diffuse and trabecular) and amyloid changes. The immunohistochemical panel showed negativity for SMA (smooth muscle actin), CDX-2, CK5, S-100 protein, PSA (prostate specific antigen), p63 and TTF-1; and positivity for CK7 (cytokeratin 7), Chromogranin A (Figure 2b), Ki-67 in 1% of cells; whereas Synaptophysin and AE1 + AE3 were also positive. These findings confirmed the diagnosis of a well-differentiated neuroendocrine neoplasia, with a solid morphological and trabecular pattern of intermediate cells infiltrating the squamous mucosa (right arytenoid). A bone

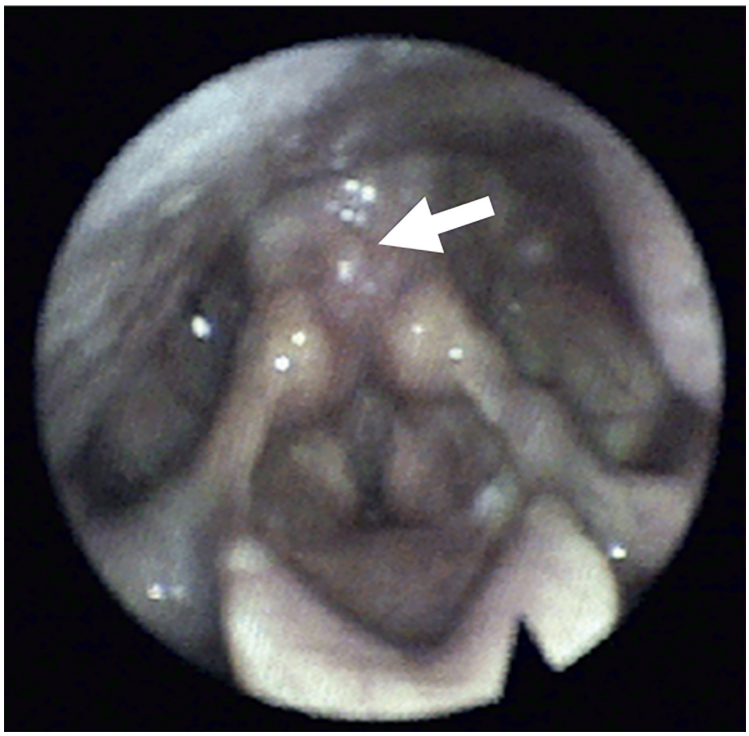

Figure 1. Laryngoscopy showing a polypoid lesion in the right arytenoid.

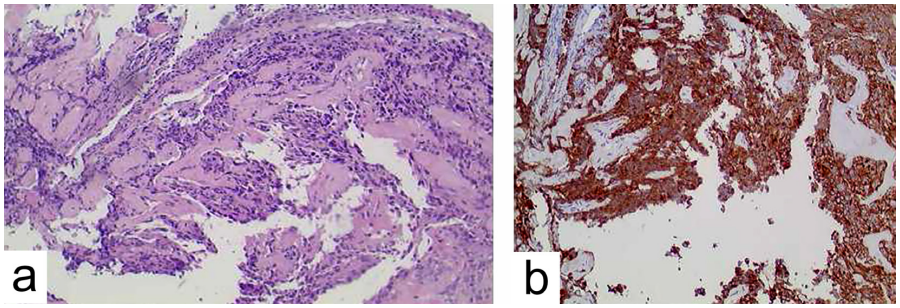

Figure 2. Photomicrographs showing: (a) papillary projections (HE, x50) and (b) immunohistochemistry strongly positive for chromogranin A (x30).

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scintigraphy showed no signs of systemic disease. The patient underwent chemotherapy with octreotide LAR 30mg IM every 30 days, in 12 cycles, and did not show evidence of recurrence after five years.

Discussion

Identifying the subtype of neuroendocrine tumor the patient presents is essential for therapeutic planning and prognosis, as different subtypes present different aggressiveness potential and clinical evolution[4]. Epithelial malignancies with neuroendocrine differentiation can occur in any organ of the body. Laryngeal tumors with neuroendocrine morphology are a distinct, unusual, heterogeneous group of neoplasms that share specific morphology, histochemistry, immunohistochemistry and ultrastructural characteristics, whereas their prognoses depend on the type of tumor. Neuroendocrine carcinomas have an immunohistochemical profile marked by some combinations of positivity for cytokeratin, chromogranin, synaptophysin, CD56, CD57, enolase, serotonin, somatostatin and bombesin[2]. Typical carcinoids can be polypoid, pedicled, or nodular. Tumors are positive for more sensitive and specific neuroendocrine markers (especially synaptophysin, chromogranin and CD56) and neuropeptide markers (in particular calcitonin and somatostatin).

Surgical excision is the treatment of choice for typical laryngeal carcinoid[5]. In addition, radiotherapy and chemotherapy were found to be ineffective for this type of tumor[2]. Partial or total laryngectomy can be performed depending on the location and extent of the primary tumor.

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