

Typical Laryngeal Carcinoid Tumor: A Case Report

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Abstract

Neuroendocrine tumors of the larynx comprise a rare group of tumors categorized as typical carcinoid, atypical carcinoid, small-cell neuroendocrine carcinomas, and paragangliomas. Atypical carcinoid type occasionally denotes carcinoid syndrome. Typical carcinoid is extremely rare. The primary diagnosis is based on light histochemical studies that should be confirmed by immunocytological and/or ultrastructural investigation. The different biological behaviors of these tumors lead to a specific diagnosis of paramount importance.

Keywords: Carcinoid tumor; Laryngeal tumor; Neuroendocrine tumors

Introduction

More than 500 cases of neuroendocrine neoplasms of the larynx have been reported in the literature.¹ Typical carcinoids are the rarest.^{2,3} The biologic behaviors of neuroendocrine neoplasms of the larynx make a specific diagnosis of paramount importance.

Case Report

A 68-year-old man referred because of a 6-month history of a permanent and progressive hoarseness and an intermittent, right ear referral otalgia without dysphagia. He had smoked two packs of cigarettes per day for 40 years and drank alcohol occasionally. The patient denied any significant past medical or surgical problems. Indirect laryngoscopy revealed the presence of an exophytic and ulcerative mass on the surface of the right false vocal cord (FVC) with extension to ipsilateral arytenoid cartilage. Laryngeal CT revealed unilateral supraglottic mass without extra laryngeal extension. The patient was admitted and scheduled for direct laryngoscopy and biopsy that revealed right ventricular space involvement and bi-

opsy specimen as a poorly differentiated adenocarcinoma. Unfortunately, the patient underwent total laryngectomy and right thyroidectomy due to the report of biopsy specimen. Microscopically, the tumor was made up of sheets of uniform neoplastic epithelial cell with round to oval mildly pleomorphic nuclei separated by a thin fibrovascular stroma (Figur 1). Immunoperoxidase staining showed a positive cytoplasmic reaction for epithelial membrane antigen (EMA), carcinoembryonic antigen (CEA) cytokeratin and synaptophysin (Figure 2). However, a final diagnosis of well-differentiated neuroendocrine carcinoma (Typical Carcinoid) was made.

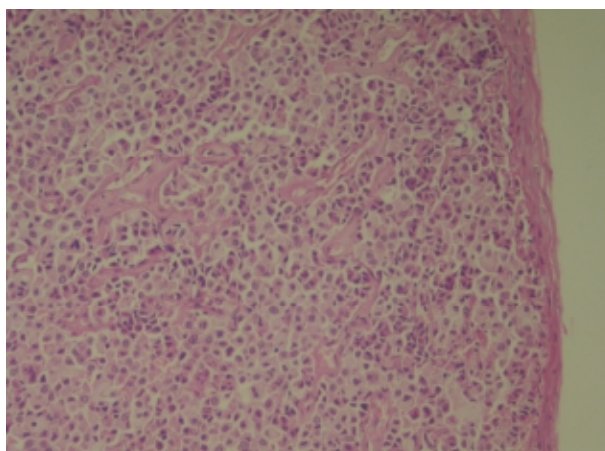


Fig. 1: Shows nests & islands of uniform epithelial cells separated by thin vascularized stroma (H&E, x100)

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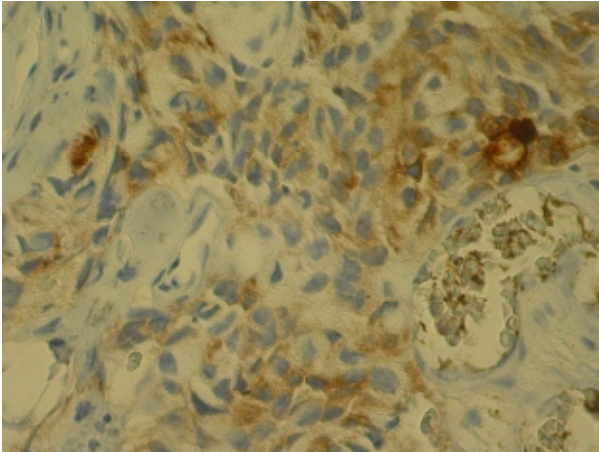


Fig. 2: Shows positive immunoperoxidase stain for synaptophysin (Immunoperoxidase stain, x400)

Discussion

Neuroendocrine neoplasms of the larynx are rare tumors that arise from the amine precursor uptake and decarboxylase (APUD) cells or the dispersed neuroendocrine cell system.⁴ However, a great deal of confusion about classification of these tumors exists. In 1991, the World Health Organization recommended a classification for neuroendocrine tumors of the larynx. Typical carcinoid tumor of the larynx is an extremely rare one; only 14 documented cases have been reported, including 3% of the entire neuroendocrine of the larynx.³ These tumors have an overwhelming male predisposition; only one of the patients was a woman.^{5,6} These tumors usually originate from un-committed stem cells in the supraglottic region. The most common complaints are pain, or lump in the throat, hoarseness, dysphagia, dysphonia and

otalgia. Histologically, these tumors are made up of sheets and nests of uniform cells with small, round or oval, centrally placed nuclei in a granular eosinophilic cytoplasm. The cells are separated by a fibrovascular or hyalinized stroma. Mitoses, pleomorphism and necrosis are lacking. The tumor usually appears as a polypoid or subepithelial nodular mass.⁷ The overlying epithelium is usually intact. Argyrophil stains are characteristically positive, and argentaffin stains often negative. Typical carcinoid should be distinguished from atypical carcinoid. Immunohistochemical, histochemical and ultrastructural investigations cannot differentiate the two types. Atypical carcinoids can stain for the same cytologic markers as typical ones, but calcitonin staining is almost universal.⁸ The diagnosis is predominantly based on light microscopy combined with positive staining for some of the general neuroendocrine markers [e.g. chromogranin, neurofilament protein (NFP), synaptophysin, neuron specific enolase (NSE), protein gene product 9.5 (PGP-9.5)] to firmly establish the lesion as a neuroendocrine tumor.⁹ None of the typical carcinoids have regional lymph node metastases at the time of clinical diagnosis, just like our case and we did not find any distance metastasis although it has been reported in four out of 14 tumors with distant metastases manifestation. Only one patient died of the disease.^{10,11} Although typical carcinoids are treated preferably conservatively unless in the case of extensiveness and invasiveness. Our patient underwent total laryngectomy upon the pathological report as poorly differentiated. However, prognosis is excellent following complete excision with clear margins.¹²

Conflict of interest: None declared.

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