Case Report

Primary Small-Cell Carcinoma of the Glottic Larynx: A Case Report from Northern Pakistan

Kanwal Awan, Humera Mahmood, Hadia Fatima*, Sarosh Arif, Mohammad Faheem
Department of Oncology, Atomic Energy Cancer Hospital, NORI, Islamabad, Pakistan

Abstract

Extrapulmonary neuroendocrine small-cell carcinoma is a rare tumor accounting for only 2.5%–4% of all small-cell carcinomas, and during the past 30 years, only 160 cases of primary small-cell carcinoma of the larynx have been reported worldwide. Most previously reported cases were metastatic small-cell carcinomas. Herein, we report the case of a 51-year-old male who was treated at the Atomic Energy Cancer Hospital, NORI, Islamabad, Pakistan for small-cell carcinoma of the larynx.

Keywords: Cisplatin, etoposide, larynx, small-cell carcinoma

INTRODUCTION

Approximately, 99% of the laryngeal tumors are squamous-cell carcinomas.[1] Laryngeal tumors are mostly found in the region of the glottis or true vocal cords. A neuroendocrine tumor of the larynx was first described in 1969 by Goldman et al.,[2] and neuroendocrine tumors in the form of atypical carcinoids have since been identified as common nonsquamous cancers that arise from the larynx. Primary Laryngeal small cell carcinoma (LSCC) accounts for <0.5% of all laryngeal neoplasms.[3] Herein, we describe a case of localized primary small-cell carcinoma (LSCC) that was diagnosed on histopathology supported by immunohistochemistry. The patient was treated with a combination of external beam radiotherapy and chemotherapy, with an excellent response to the treatment.

CASE REPORT

A 51-year-old male patient with hypertension which was controlled by medication and no family history of any malignancy presented with the complaints of hoarseness of voice for the past 8 months associated with an occasional cough but not accompanied with bloodtinged discharge. His swallowing and dietary habits were normal. The patient had smoked a hukka and cigarettes for the past 20 years. There was no palpable cervical lymphadenopathy. A fiber-optic endoscopic examination revealed a left vocal cord lesion involving almost the whole-vocal cord. Vocal cord mobility was intact. Multiple biopsies were taken...
from the growth, which showed the features of small-cell cancer, including a high nuclear-to-cytoplasmic ratio with scant cytoplasm, round-to-oval cells, and a large number of mitotic figures [Figure 1]. Immunohistochemistry was positive for cytokeratin, synaptophysin, and CAM 5.2 [Figure 2].

A computed tomography (CT) scan with contrast from the base of his skull to the clavicles showed subtle thickening at the anterior part of his left vocal cord, but the other findings were unremarkable [Figure 3]. His CT scan of the chest, abdomen, and bone was normal [Figure 4], and hence the disease was nonmetastatic. The diagnosis of primary small cell of the larynx was confirmed. The stage was T1aN0M0 (Stage 1a).

He was treated with external beam radiotherapy (55 Gy with 2.75 Gy/fraction in 20 fractions) followed by six cycles of chemotherapy (cisplatin 75 mg/m² on D1 and etoposide 100 mg/m² D1–d3). Six months after completing treatment, there was no evidence of local or regional recurrence in a postradiotherapy CT scan.

**DISCUSSION**

Laryngeal cancers are mostly squamous-cell carcinomas. Laryngeal small-cell neuroendocrine tumors are a rare malignancy with aggressive clinical behavior. Neuroendocrine small-cell carcinomas are usually observed in males (3:1 male-to-female ratio) with a median age at diagnosis ranging from 50 to 70 years.[4] Our patient was male and 51 years old.

Heavy cigarette smoking is the most important risk factor involved in the etiology of such tumors.[5] In addition to cigarette smoking, our case also had a history of hukka smoking. Common presenting symptoms include dysphonia, dysphagia, or in some cases a neck mass.[1] Our patient presented with dysphonia and hoarseness of voice.

Although this rare tumor has been reported in various locations in the larynx, the supraglottic region is the most frequently involved site.[4] In our patient, the site of origin was the left vocal cord. Primary small-cell carcinoma of the larynx usually presents as locally advanced or metastatic
Small-cell carcinoma may be immunoreactive for cytokeratins and for general neuroendocrine markers, including chromogranin, CD 56, and synaptophysin.\(^6\) In our case, immunohistochemical studies were positive for cytokeratin, synaptophysin, and CAM 5.2.

Systemic chemotherapy should be used to treat laryngeal small cell carcinoma, as there is a possibility of occult metastasis in cases presenting with localized disease. Moreover, the surgery should be avoided because of the poor prognosis and the morbidity associated with surgery of the larynx.\(^8\)\(^9\) Hence, organ preservation with radiotherapy combined with adjuvant or concurrent chemotherapy should be the prime objective in the treatment of laryngeal small cell carcinoma.\(^10\) Our patient was treated with external beam radiotherapy 55 Gy with 2.75 Gy/fraction for 20 days followed by adjuvant chemotherapy with cisplatin 75 mg/m\(^2\) D1 and etoposide 100 mg/m\(^2\) D1–D3 in 3 weekly protocols for six cycles.

The prognosis of patients with primary laryngeal small cell carcinoma is considered to be poor with a 5-year survival rate of only 5%.\(^8\) Although patients with localized early-stage disease can have prolonged survival, but there is a possibility of treatment resistance.\(^8\)\(^9\) Our patient had a complete response to the treatment, and he has been free from disease for the 6-month follow-up visit.

**Conclusion and Recommendation**

A diagnosis of early-stage primary small-cell carcinoma of the larynx is extremely rare, and histopathologists should keep in mind of this rare diagnosis when evaluating the laryngeal carcinomas. Treatment of localized laryngeal small cell carcinoma with external beam radiotherapy and chemotherapy can result in a satisfactory response and can be considered to be the standard treatment for early-stage laryngeal small cell carcinomas.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**