

# Primary small cell carcinoma of the larynx

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## ABSTRACT

Squamous cell carcinoma is a common histological type for laryngeal cancer and primary laryngeal small cell carcinoma (LSCC) is a rare occurrence. The prognosis of LSCC is also considered to be poor. We present here a case of 40-year-old female with localized primary small cell carcinoma of the larynx. The diagnosis of LSCC was made after histopathological examination and confirmed by immunohistochemistry study. The patient was treated by concurrent radiotherapy and chemotherapy. In our limited experience, there was a good therapeutic response to treatment of localized primary small cell carcinoma of the larynx.

**Key words:** Larynx, neuroendocrine tumor, small cell carcinoma

## INTRODUCTION

Cancers of the larynx are usually squamous cell carcinomas. Primary laryngeal small cell carcinoma (LSCC) is a rare occurrence. Neuroendocrine tumor of the larynx was first described in 1969 by Goldman *et al.*,<sup>[1]</sup> and neuroendocrine tumors in the form of atypical carcinoids are common nonsquamous malignancy that arises from the larynx. Primary LSCC accounts for <0.5% of all laryngeal neoplasms.<sup>[2]</sup> We describe here a case of localized primary LSCC that was diagnosed by histopathology supported by immunohistochemistry (IHC), its treatment by combination of external beam radiotherapy (EBRT) and chemotherapy, and response to treatment.

## CASE REPORT

A 40-year-old female presented at our outdoor department with the chief complaint of foreign body sensation in the throat of 4 months duration. There was associated history of occasional cough. The patient was a tobacco chewer

and a nonsmoker. There were no palpable cervical lymph nodes enlargements. On examination with fiber-optic endoscopy, there was a polypoidal growth involving the right aryepiglottic fold, interarytenoid region of the larynx [Figure 1]. The vocal cords mobility was intact. Multiple punch biopsies were taken from the growth, which showed features of small cell carcinoma [Figure 2]. IHC study was done for synaptophysin and cytokeratin (CK). IHC was positive for the expression of synaptophysin and CK [Figure 3a and b]. The diagnosis of primary small cell carcinoma was confirmed. The staging for the disease was T2N0M0 (Stage II) supraglottic malignancy.

The patient was treated with EBRT (35 fractions/70 Gy) and six cycles of weekly concurrent chemotherapy with carboplatin 150 mg/cycle. On the follow-up at 24 months following the completion of treatment, there was no evidence of local or regional recurrence.

## DISCUSSION

Globally the incidence of laryngeal cancers is around 2–5% of all cancers.<sup>[3]</sup> In our population, the incidence of hypopharyngeal cancer is higher than laryngeal malignancy. Neuroendocrine small cell carcinomas are usually seen in males and the median age at diagnosis ranges from 50 to 70 years.<sup>[4,5]</sup> Our patient was a female of relatively younger age. Most patients with LSCC are smokers,<sup>[2]</sup> but in our case the patient was a nonsmoker. However, she was

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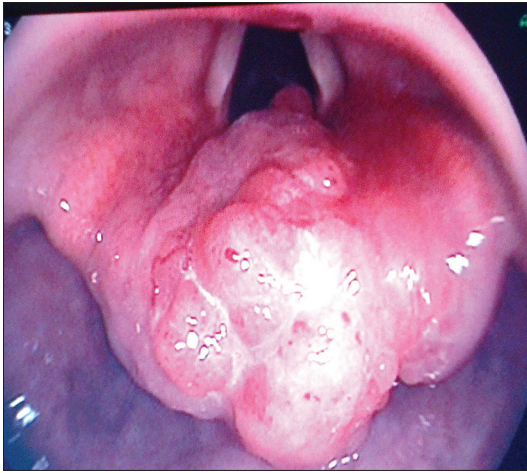
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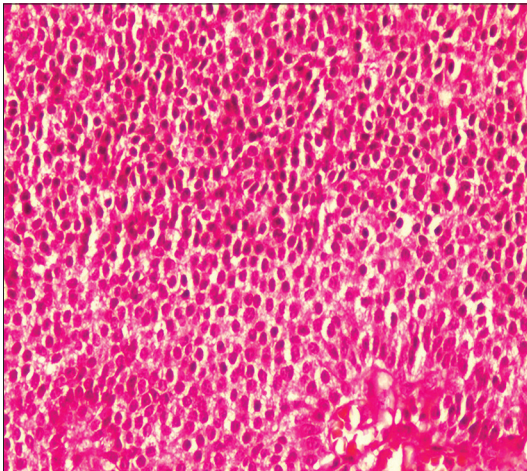
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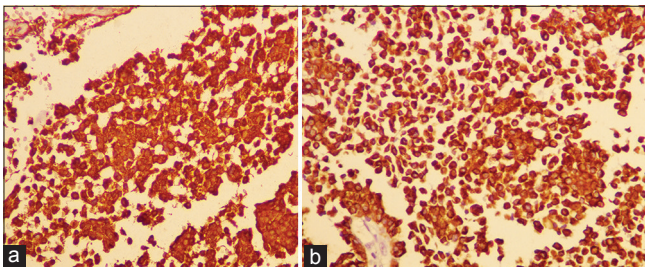
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**Figure 1:** Endoscopic image showing the localized polypoidal growth involving the supraglottic larynx



**Figure 2:** Photomicrograph with (H and E, x40) showing malignant cells with uniform nuclei, granular chromatin and inconspicuous nucleoli



**Figure 3:** (a) Photomicrograph of immunohistochemistry study showing the tumor positivity to synaptophysin, (b) Photomicrograph of immunohistochemistry study showing the tumor positivity to cytokeratin

having a history of consuming chewable forms of tobacco. Neuroendocrine carcinoid tumor of the larynx has been reported in the literature on the aryepiglottic folds, while LSCC involves whole of the larynx in a homogenous fashion.<sup>[6,7]</sup> Furthermore, primary LSCC usually presents with loco-regional disease or metastasis.<sup>[8]</sup> However, in our case the patient presented only with localized disease in the supraglottic larynx without any cervical

lymphadenopathy and metastasis. In extremely rare situation, LSCC of the larynx has been reported to occur as a second primary tumor following small cell carcinoma of the lung.<sup>[9]</sup> Our case was a primary LSCC. Neuroendocrine neoplasms of the larynx are categorized into the five tumor types: Typical carcinoid, atypical carcinoid, small cell neuroendocrine carcinoma, large cell neuroendocrine carcinoma, and paraganglioma.<sup>[10]</sup> Thus, the diagnosis of small cell neuroendocrine carcinoma should be confirmed by histopathological examination supported by IHC study. In our case, the IHC study was positive for the expression of synaptophysin and CK which confirmed the diagnosis of small cell carcinoma of the larynx. Neuroendocrine tumors of the larynx are said to be characterized by its pathological heterogeneity.<sup>[11]</sup> The classification of neuroendocrine tumors based upon its differentiation is helpful in determining its prognosis.<sup>[7]</sup> In the treatment of LSCC systemic chemotherapy should be given, as there is a possibility of occult metastasis in cases presenting with localized disease. Moreover, surgery should be avoided because of the poor prognosis and the morbidity associated with surgery of the larynx.<sup>[12]</sup> Hence, organ preservation by treatment with concurrent chemo-radiotherapy should be the prime objective in the treatment of LSCC. Primary advanced LSCC has been reported to be successfully treated only with chemotherapy agents cisplatin, and irinotecan,<sup>[13]</sup> and also by concurrent chemo-radiotherapy.<sup>[14]</sup> The prognosis of patients with primary LSCC is considered to be poor with a 5 years survival of 5%.<sup>[8]</sup> In our case, there was complete response to concurrent chemo-radiotherapy for localized LSCC, and the patient was free from disease at the 2 years follow-up. The long term survival following the treatment in this case cannot be commented upon here, as in this case, the follow-up period was for 2 years after completion of the treatment.

## CONCLUSION

Diagnosis of early stage small cell carcinoma of the larynx is extremely rare. Our case has shown that treatment of localized LSCC with chemotherapy and EBRT results in a good therapeutic response.

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