

Primary Small Cell Carcinoma of Subglottis: A Rare Case Report

Ruchika Jain*, D.P. Gupta, Beni Prasad

Department of ENT, SMS Medical College, Jaipur, Rajasthan, India

*Corresponding Author:

Name: Dr. Ruchika Jain

Email: jainr15@yahoo.co.in

Abstract: Neuroendocrine neoplasms of the larynx are rare and are most commonly supraglottic in origin. Subglottic location for small cell carcinoma is extremely rare with patient presenting with dyspnea, hoarseness and dysphagia. Small-cell neuroendocrine carcinomas are very aggressive neoplasms. We present here a case report of this rare tumor in a 50 year old female.

Keywords: Subglottis, Neuroendocrine cancer, Female, Immunohistochemistry, Radiotherapy, Prognosis

INTRODUCTION

Laryngeal carcinomas comprise approximately 2% to 5% of all malignancies worldwide, out of which approximately 99% are primary squamous cell carcinomas [1]. Neuroendocrine tumors are the most common nonsquamous neoplasms arising in larynx and account for <1% of primary laryngeal carcinomas [2]. The primary small cell neuroendocrine carcinoma of larynx is a rare, highly lethal tumor of larynx associated with frequent and early widespread metastases [3]. Most of the patients are males, with history of smoking and between 50 to 70 years of age at presentation. Symptoms are similar to other laryngeal carcinomas or rarely, may present with paraneoplastic syndrome. The diagnosis of small cell carcinoma of the larynx is essentially based on the light microscopic examination aided by electron microscopy or immunohistochemical staining [4]. The case is described here for its rare presentation in a female with subglottic origin.

CASE REPORT

A 50 year old female, chronic cigarette smoker, presented with difficulty in swallowing for last 1 month with and respiratory distress progressively getting worse for the last 10 days. An indirect laryngoscopy was performed which revealed a large subglottic mass just below the vocal cords. An urgent tracheostomy was done to relieve the stridor. Following this, the patient was evaluated further and a CECT neck and chest was done which showed an enhancing mass of size 23x16x21 mm involving posterior and lateral walls of subglottis and extending to postcricoid region suggesting a malignant pathology [Figure 1]. Multiple subcentimeter sized nodes were noted along bilateral upper internal jugular veins. Then the patient underwent direct laryngoscopy and a biopsy was taken. The histopathological evaluation revealed small round cells

with scant amount of cytoplasm and hyperchromatic nuclei suggestive of an undifferentiated malignant round cell tumor. Immunohistochemistry was done which was positive for EMA (epithelial membrane antigen), synaptophysin, cytokeratin, chromogranin-A and TTF1 (thyroid transcription factor 1). Based on IHC a diagnosis of subglottic small cell carcinoma was made. On further workup no distant metastases were identified by radiology.

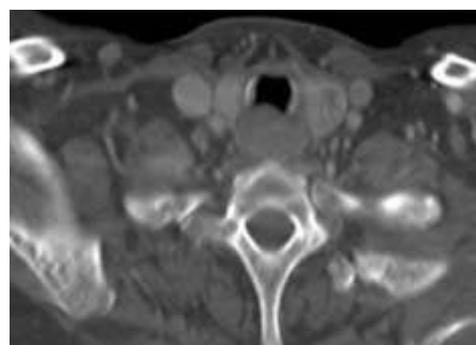


Fig. 1: CECT neck showing subglottic tumor

The patient was given palliative external radiotherapy in a dose of 20 Gy in five fractions over one week. After the completion of radiotherapy the patient was given combination chemotherapy with cisplatin and etoposide. The patient was offered best supportive care but she succumbed to the disease 8 months after diagnosis.

DISCUSSION

Small cell neuroendocrine carcinoma (SCNC) is a highly aggressive malignancy, occurring most commonly in lungs [5]. Extrapulmonary SCNCs are relatively uncommon, being most common in esophagus. In head and neck region, SCNC mostly

involves the larynx with the supraglottic region being the most common site.

The WHO has classified neuroendocrine tumor of the larynx into four types: typical carcinoid tumor (well differentiated, grade I), atypical carcinoid (moderately differentiated, grade II), small-cell (poorly differentiated, grade III) and paragangliomas [6].

An accurate histologic diagnosis is essential because the treatment and prognosis depends on the type of neuroendocrine tumor [6]. Small cell neuroendocrine neoplasm of the larynx has the worst prognosis, with five-year survival rates of 5% [5]. Small-cell neuroendocrine carcinoids are further categorized as oat-cell, intermediate-cell, or combined-cell types. Cervical and distant metastases are common at the time of diagnosis [7]. The treatment of small cell carcinoma of the larynx is controversial as only few cases have been reported in the literature and there is lack of controlled studies. Reports of improved survival with systemic chemotherapy combined with radiation therapy had been suggested as a primary treatment modality for laryngeal small cell carcinoma [8]. Cisplatin, etoposide, cyclophosphamide, doxorubicin, vincristine, and methotrexate have been the most commonly used chemotherapeutic agents [5, 8]. In our case, the patient was given radiotherapy for palliation of local symptoms followed by systemic chemotherapy with cisplatin and etoposide combination. Most authors generally agree that surgery alone or in combination with radiotherapy does not cure local tumor. Therefore, radical surgical procedure is not recommended as the initial treatment of choice. Surgery for this tumor should be reserved for persistent and recurrent disease at the primary site and neck (9). The disease extent and treatment modality used is the significant prognostic factors for survival [7].

CONCLUSION

Small cell neuroendocrine carcinomas of larynx are a rare occurrence having an aggressive course with poor prognosis. There are no specific treatment guidelines for these tumors and further studies are required to define the best treatment modalities.

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