Int. j. adv. multidisc. res. stud. 2024; 4(5):645-646

Received: 17-08-2024
Accepted: 27-09-2024

International Journal of Advanced Multidisciplinary Research and Studies

ISSN: 2583-049X

Primary Small Cell Neuroendocrine Carcinoma of Larynx: An Atypical Case Presentation

[1] Dr. Tania Hoq, [2] Dr. Suriana Sultana, [3] Dr. Sonya Begum, [4] Dr. Shamima Afroz Trina, [5] Dr. Kazi Manzur Kader

[1] Registrar, Department of Clinical Oncology, Delta Hospital Limited, Mirpur-1, Dhaka, Bangladesh
[2, 3] Junior Consultant, Department of Clinical Oncology, Delta Hospital Limited, Mirpur-1, Dhaka, Bangladesh
[4] Consultant, Department of Clinical Oncology, Delta Hospital Limited, Mirpur-1, Dhaka, Bangladesh
[5] Senior Consultant and Head of the Department, Department of Clinical Oncology and Medical Physics, Delta Hospital Limited, Mirpur-1, Dhaka, Bangladesh

DOI: https://doi.org/10.62225/2583049X.2024.4.5.3303
Corresponding Author: **Dr. Tania Hoq**

Abstract

Neuroendocrine tumors account for <1% of all malignant tumors that can originate from any part of the body, most frequently diagnosed in the lung. Extrapulmonary presentation is rare, common sites are genitourinary and gastrointestinal. According to the 2017 WHO proposed classification, small cell type (SCNEC) is a poorly differentiated neuroendocrine carcinoma. The most common site in the head and neck is the larynx, accounts for <0.6%. Small cell neuroendocrine carcinomas are extremely rare entity of laryngeal cancers. We report a case of small cell neuroendocrine carcinoma of larynx.

Keywords: Carcinoma, Neuroendocrine, Larynx

Introduction

Unique growth of neuroendocrine cells, commonly seen in adults, are generally converted into neuroendocrine carcinomas, which was also called carcinoid tumors. It may be spread into other parts of body which is highly heterogenous [1]. The commonest parts where this kind of carcinoma can occur are, lungs, appendix, small intestine, rectum and pancreas [2].

As it is one of the uncommonest comparing with the other cancer, and the symptoms are also non specific (cough, abdominal pain, bloating and weight loss) to physicians as well as the patients, the diagnosis is also delayed due to these reasons. There are few studies found in this rare diseases, where we can find the evidences of neuroendocrine carcinoma patients with other metastases. This is one of the consequences of frequent delayed diagnosis. The diagnosis is confirmed by pathological testing (Obtaining tissues). Chromogranin A, diagnostic biomarker, has a high sensitivity (53%–91%) but low specificity (< 50%). CT scan and MRI are another diagnostic tools of this rare disease [3].

One of the rarest, however, highly aggressive type of malignant cancers are neuroendocrine carcinoma of larynx. It can be classified into 2 divisions on tissue origins: Epithelial and neural. Typical carcinoid, atypical carcinoid, small cell neuroendocrine carcinoma, and large cell neuroendocrine carcinoma are The majority of LNECs [4].

Case Presentation

A 55 years old male came to the outdoor of Delta Medical Hospital, on June 2021, having a positive history of smoking, had been experiencing progressive hoarseness of voice for 6 months. Direct laryngoscopy showed huge polypoid mass in right vocal cord which was removed surgically. Biopsy report revealed confirmed diagnosis of small cell neuroendocrine carcinoma on December 2021. Immunohistochemistry showed positive for synaptophysin and chromogranin,, negative for CD-20, CD-3 and LCA. After that patient didn't take further treatment as voice was restored. After 3 months, horseness reappeared along with progressive dysphagia.

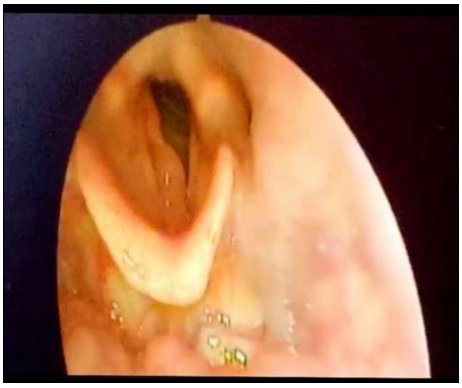

Fig 1: Direct laryngoscopy showed huge polypoid mass in right vocal cord which was removed surgically

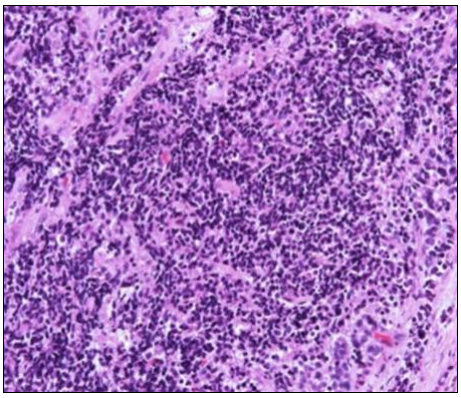

Fig 2: Biopsy report revealed small cell neuroendocrine carcinoma

On CT scan of neck, growth extended from right vocal cord to infraglottic part of right side of larynx including laryngotracheal junction. Prominent lymphnodes at right side of neck (level II, VA).

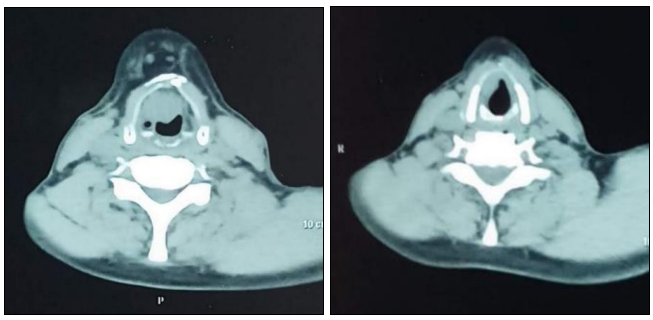

Fig 3: On CT scan of neck, growth extended from right vocal cord to infraglottic part of right side of larynx including laryngotracheal junction. Prominent lymph nodes at right side of neck (level II, VA)

Treatment

Patient was treated with adjuvant chemotherapy (Etoposide+ Cisplatin) for 6 cycles and response was satisfactory. Chemotherapy was started from July 2022 and ended on October 2022. After the chemotherapy, he received radiotherapy 60 Gray, 2 Gray/ fraction over 6 weeks (December 2022-January 2023). Patient was on regular follow up and 6 months after the end of treatment the he remained in good loco-regional control up to June 2023. 8 months after completion of treatment patient developed ill defined opacity in lower zone of left lung and multiple liver SOLs which was found on September 2023. FNAC from liver SOL suggested metastatic carcinoma. On December 2023, patient was adviced for palliative chemotherapy but couldn't received due to lack of physical fitness.

Outcome

After 1 month, in the year of 2024, at January, the patient died for metastatic complications.

Discussion

Small cell neuroendocrine carcinoma is the most lethal tumor of the larynx, which develops rapidly. More than 90% of these patients have metastatic disease in its clinical course and the most common sites of metastatic spread are the cervical lymph node, liver, lung and bone. Surgical results for this tumor have been disappointing and is reserved for cases of local relapse with no evidence of metastasis. Chemotherapy and radiotherapy currently appear to offer the least disability and most effective form of therapy. Resistance to chemotherapy represents an important indicator of poor prognosis [5, 6].

Conclusion

Small cell laryngeal cancers are extremely rare and represent 1% of all invasive laryngeal cancers. It should be recognized and treated early because of its biologic potential of rapid growth, early dissemination and responsiveness to chemotherapy. Chemotherapy and radiotherapy remain the mainstay of treatment.

References

1. Oronsky B, Ma PC, Morgensztern D, Carter CA. Nothing but NET: A review of neuroendocrine tumors and carcinomas. Neoplasia. 2017; 19(12):991-1002.
2. Ahmed M. Gastrointestinal neuroendocrine tumors in 2020. World Journal of Gastrointestinal Oncology. 2020; 12(8):791.
3. Raphael MJ, Chan DL, Law C, Singh S. Principles of diagnosis and management of neuroendocrine tumours. CMAJ. 2017; 189(10):E398-E404.
4. Zhu Y, Gao L, Meng Y, Diao W, Zhu X, Li G, *et al*. Laryngeal neuroendocrine carcinomas: A retrospective study of 14 cases. BioMed Research International. 2015; 2015(1):832194.
5. ALAMI Z, Bouziane A, Hassani W, Farhane FZ, El M. Small Cell Neuroendocrine Carcinoma of the Larynx: Case Report and Literature Review.
6. Raposo A, Marco A, García-Solano ME, Martínez-Ortiz MJ, García-Purriños F, Lajara J. Primary small cell carcinoma of the larynx. Survival time of 47 months. Case report. Annals of Medicine and Surgery. 2018; 30:46-49.