

Primary Small Cell Carcinoma of Hypopharynx: A Case Report of a Rare Tumor

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Abstract

Primary hypopharynx involvement of small cell carcinoma is very rare and very few cases have been reported in the texts. Here, we report a case of primary small cell carcinoma of hypopharynx with metastases to left humerus in a female patient of north india.

Case Report. A 54 year old woman presented with a 4month history of shortness of breath and neck swelling in the left side. Fibreoptic laryngoscopy and biopsy revealed small cell carcinoma of the hypopharynx.

Discussion. Small cell carcinoma of the hypopharynx still does not have any clear modality of treatment due to being a rare entity. Systemic chemotherapy and radiotherapy should be preferred over other treatment modalities.

1. INTRODUCTION

Small cell carcinoma (SMCC) is a subset of the neuro-endocrine tumors , mostly occurring in the lungs. Extra-pulmonary SMCC comprises 2.5- 5% of all SMCC's [1]. SMCC may arise in extra-pulmonary sites, throughout the body , commonly in oesophagus, large bowels, bladder and the larynx [2]. This form of tumor have aggressive nature with rapid local progression and early regional and distant spread [2]. The incidence of extra-pulmonary SMCC is much lower than small cell lung cancer accounting for only 6% of SMCC's [3, 4]. The role of local and systemic treatment strategies for extra-pulmonary SMCC's is still not clearly defined [3, 4].

The first reported case of extra-pulmonary SMCC, was a primary tumor in the larynx, reported by Oloffson and Van Nostrand in 1972 [5]. Since then it has been reported to occur in multiple sites of head and neck region, including the larynx, paranasal sinuses and salivary glands. In India, small cell carcinoma of hypopharynx is an extremely rare entity, with one such case being reported in August, 2023 by GH Haritha et al. from JIPMER [6].

Here, we report a case of primary SMCC of the hypopharynx in a female patient.

2. CASE REPORT

A 54 year old patient from Dwarka area of Delhi presented, with history of neck swelling for last 4 months with complaints of severe shortness of breath, to the Department of E.N.T, VMMC and Safdarjung Hospital. Emergency tracheostomy was done and fibreoptic laryngoscopy in operation theatre revealed growth in bilateral pyriform sinus while bilateral true vocal cords were mobile and chink was also adequate. Fibreoptic laryngoscopy guided biopsy was taken from the growth which revealed small cell carcinoma and patient was referred to the Department of Radiation Oncology for further management. Patient had no history of co-morbidities like hypertension, diabetes, chronic

obstructive pulmonary disease, tuberculosis etc. Patient gave history of bidi smoking about 1pack/day for more than 30 years.

PET-CT revealed FDG avid mass (SUV-11.6) measuring 5.7 x 7.5 x 8.9 cm in the hypopharynx with complete obliteration of airway and also involving the left thyroid lobe. There was subcentrimetric and enlarged bilateral cervical level II-IV lymph nodes and clinically left level III-IV hard fixed node approximately 4 x 4 cm. PET-CT also revealed an intramedullary lesion (SUV-12.6) in proximal left humerus suggestive of hematogenous metastases to bone. NCCT of brain was also done in view of brain metastases, which revealed normal study.

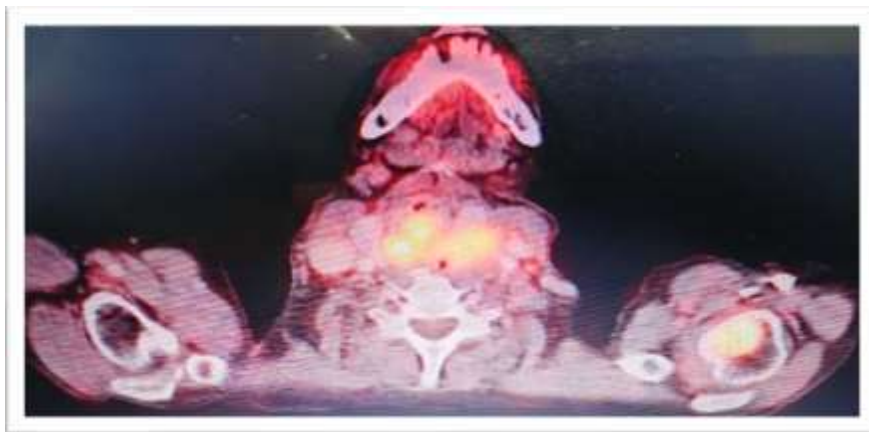


FIGURE-1: PET-CT IMAGE SECTION SHOWING FDG AVID LESION IN HYPOPHARYNX WITH ENLARGED NODES AND ANOTHER FDG AVID LESION IN LEFT HUMERUS [RED ARROW].

Histopathological analysis of the biopsy specimen revealed a tumor consisting of tightly packed cell nests and diffuse irregularly shaped sheets of cells with areas of necrosis. The tumor cells were of small size with hyperchromatic, round to oval nuclei. The tumor cells were immunoreactive for EMA, synaptophysin, chromogranin and Ki-67 index was very high (Fig). These findings were compatible with primary oat cell small cell carcinoma of hypopharynx.

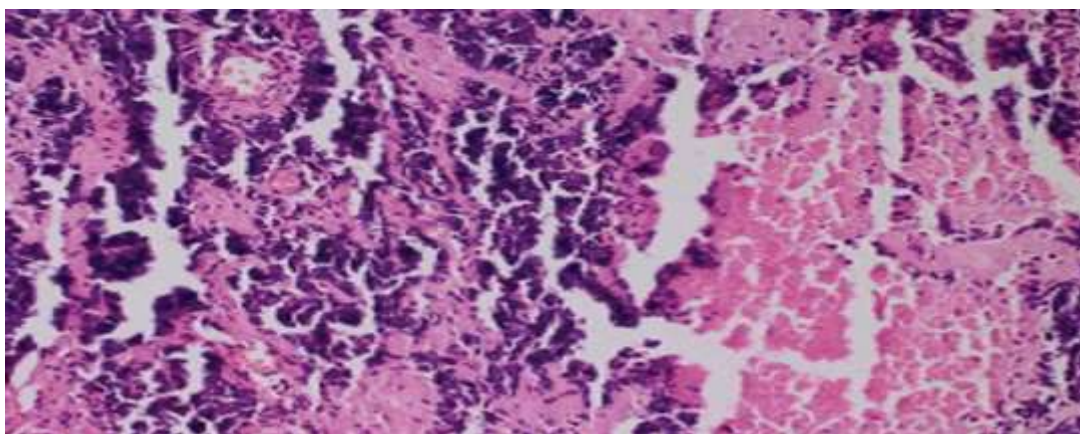


FIGURE 2: Hematoxylin and eosin staining showing small cells, round to fusiform in shape with scanty cytoplasm and absence of nucleoli.

The patient was finally planned for palliative systemic chemotherapy with Cisplatin and Etoposide with zoledronate for bone metastases and palliative radiation therapy to left humerus lesion whenever

required. Currently the patient is doing well with chemotherapy showing signs of good response clinically and subjectively.

3. DISCUSSION

The mainstay of treatment for EPSMCC is systemic chemotherapy, because these tumors are characterized by a high propensity for metastatic dissemination even in patients with clinically localized tumor. As EPSMCC does not have a proven algorithm for treatment, it has generally been treated in a similar fashion as small cell lung cancer [7]. The latest evidence based clinical guidelines for small cell lung cancer by National Comprehensive Cancer Network guidelines v.2023 [<http://www.nccn.org>] suggests maximum of four to six cycles of etoposide and platinum based therapy in limited stage disease. However we need more novel agents which can provide significant impact on patients survival like immune checkpoint inhibitors, thereby providing a robust outcome [8].

4. CONCLUSION

Small cell carcinoma of hypopharynx is an extremely rare entity of tumors of head and neck region with no clear treatment strategies or standard guidelines.

CONFLICTS OF INTEREST The authors declare that they have no conflict of interests concerning this paper.

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