

CASE REPORT

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# A rare localization of small-cell carcinoma in head and neck area: case report

Lorna Ting Kang Ni<sup>1</sup>, Michael Wong Sze Liang<sup>1</sup> and Hardip Singh Gendeh<sup>2\*</sup>

## Abstract

**Background:** Small-cell carcinoma (SMCC) of hypopharynx is an extreme rare entity in the head and neck region. It is commonly arising from pulmonary in origin. We report a rare case of small-cell carcinoma of hypopharynx that caused upper airway obstruction in an elderly gentleman.

**Case presentation:** A 67-year-old man is presented to otorhinolaryngology clinic with a 4-month history of worsening shortness of breath and stridor. He was heavy tobacco smoker for 60 pack-years. Flexible nasopharyngolaryngoscopy revealed a large mass at the right pyriform fossa extending superiorly and obstructing the laryngeal inlet. He underwent tracheostomy, examination under general anesthesia with tissue biopsy. Histology from tissue biopsy revealed small-cell carcinoma. The positron emission tomography-CT showed exophytic mass from right pyriform sinus with regional and lung pleural metastasis as well. There was the absence of formal staging system for SMCC due to its limited and rare incidence. He was treated with combination of carboplatin and etoposide, similar to the regime for cell carcinoma of lung. The tumor regressed loco-regionally, and his pleural metastasis had shrunk to sub-centimeter in size.

**Conclusions:** To date, there is very limited data available on head and neck small cell carcinomas to guide treatment recommendations as well as staging system. Paucity of clinical data and outcome about this rare malignancy makes this report a valuable piece of experience to be shared together with existing literature.

**Keywords:** Extrapulmonary small-cell carcinoma, Hypopharynx, Carboplatin, Etoposide

## Background

Small-cell carcinoma is known to be a rare, distinct form of malignant tumor that is most commonly of bronchogenic in origin. It is found to comprise 10 to 36% of all bronchogenic carcinomas. Extrapulmonary small-cell carcinomas constitute only 2.5 to 5% of all other small-cell carcinomas. It has been reported in multiple sites in head and neck region, most commonly at the larynx, followed by paranasal sinuses, pharynx, and oral cavity [1, 2]. To date, there is very limited data available on head and neck small-cell carcinomas to guide treatment

recommendations. Herein, we report our experience in managing a rare hypopharynx SMCC, which could strengthen the currently existing literature.

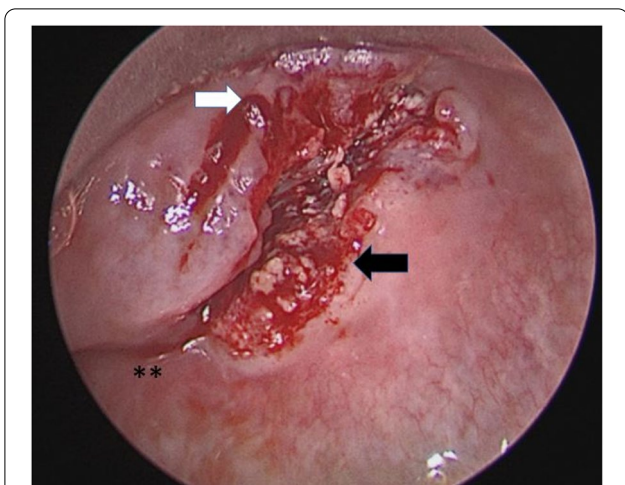
## Case presentation

A 67-year-old man presented to the otorhinolaryngology outpatients with a 4-month history of dyspnea and noisy breathing. The symptoms worsened with exertion. There was associated dysphagia, foreign body sensation, right referred otalgia, and right cervical swelling. He was heavy tobacco smoker for 60 pack-years. On examination, he was in respiratory distress with loud inspiratory stridor. He had a right cervical lymphadenopathy at level 4 that measured 4 cm by 5 cm. Flexible nasopharyngolaryngoscopy revealed a large mass from right pyriform fossa which was partially obstructing the laryngeal inlet; visualized vocal folds were mobile bilaterally.

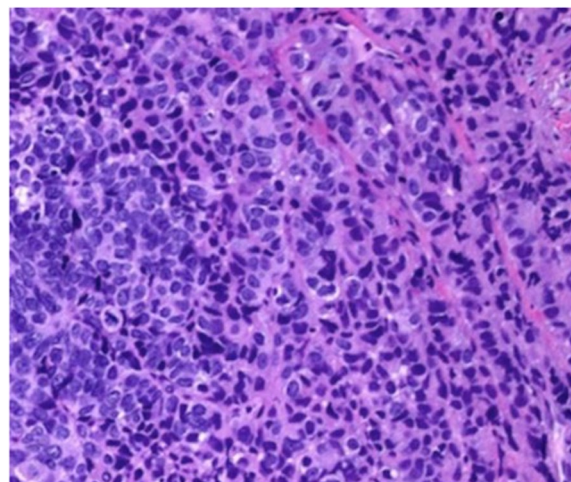
\*Correspondence: [hardip88@hotmail.com](mailto:hardip88@hotmail.com)

<sup>2</sup> Department of Otorhinolaryngology, Head & Neck Surgery, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, 56000 Kuala Lumpur, Malaysia

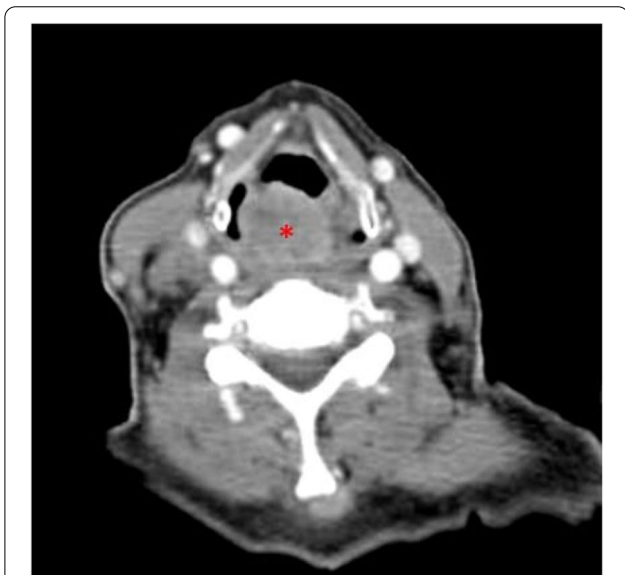
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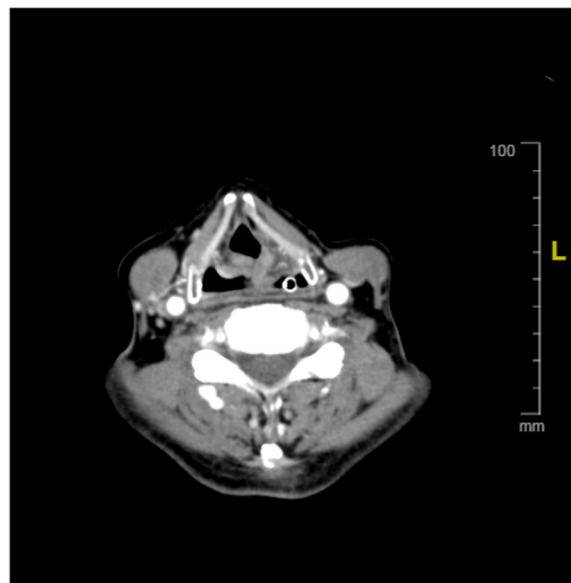
**Fig. 1** Endoscopic picture showed ulcerative mass from right pyriform sinus (black arrow) with extension into post-cricoid (\*\*) and right laryngeal inlet (white arrow)



**Fig. 3** HPE is similar to counterpart of pulmonary small cell carcinoma. There was the presence of loose aggregates of uniform small cells with indistinct cell borders, scant cytoplasm, hyperchromatic nuclei with fine granular chromatin, and nuclear molding



**Fig. 2** CT component of PET-CT scan. Axial CT neck showed an enhancing lesion from the right side of pyriform sinus\*, without a clear plane from laryngeal inlet and post-cricoid area



**Fig. 4** CT scan of the neck. Post completion of 6 cycles of chemotherapy showed a resolution of hypopharyngeal mass

He underwent tracheostomy, examination under general anesthesia with tissue biopsy. The tumor arise from the right pyriform sinus with extension to post-cricoid area and right laryngeal inlet (Fig. 1). The positron emission tomography-CT showed exophytic mass from right pyriform sinus correlated well with physical findings (Fig. 2). Regional (right cervical level 4 nodes) and distant metastasis (lung pleural) were present. The histology from the tissue showed small-cell carcinoma (Fig. 3). There was the absence of formal staging system

for SMCC due to its limited and rare incidence. He then completed 6 cycles of chemotherapy with carboplatin and etoposide and showed good response. The tumor regressed locoregionally (Fig. 4), and his pleural metastasis had shrunk to subcentimeter in size. Surveillance scan did not showed progression of the disease upon reporting of this case.

## Discussion

Small-cell carcinoma in head and neck region is known to be extremely unusual, if occurring outside the pulmonary region, and is called extrapulmonary SMCC which comprises 2.5–5% of all SMCC. According to an analysis of the National Cancer Database in 2017, only 11–16% is head and neck SMCC out of all the extrapulmonary SMCC [1–3]. It is notorious of its aggressive behavior with tendency of early hematogenous metastasis. It carries poor prognosis, and 5-year survival is 5% [4]. Haider et al. reported that 20% of extrapulmonary SMCC cases were from gastrointestinal origin, 18% from genitourinary tract, 11% from gynecological system, 10% from the head and neck, and 31% were from the unknown primary site [4]. Most common site of occurrence in head and neck and gastrointestinal region is larynx and esophagus, respectively, whereby hypopharynx made up the least incidence among all other head and neck SMCC. Hypopharynx SMCC only take up 4% out of the 11–16% head and neck SMCC [2–4]. Due to its rarity, the exact incidence and associated risk factors are difficult to be determined. There were only 12 cases reported to date regarding hypopharynx SMCC particularly pyriform sinus in the literature by which half of them were having combined histopathological picture with squamous cell carcinoma [5]. Many authors observed relationship between the disease with alcohol and smoking. In general, this type of tumors occurs at 6th and 7th decade of life with male predominance [6].

Extrapulmonary SMCC is defined as histopathologically proven small-cell carcinoma biopsied from non-pulmonary primary site. By definition, imaging from chest radiography or computed tomography scan should reveal no evidence of lesion in the lung. The bronchoscopy and/or sputum cytology was negative for malignant cells [2, 3]. However, our reported case had both hypopharynx and multiple peripheral scattered lungs mass. Primary pulmonary small-cell carcinoma will present centrally for 90 to 95% of cases [7]. Therefore, primary hypopharynx small-cell carcinoma with pulmonary metastasis was more likely rather than the reverse. Initial theory described that SMCC derives from amine-precursor uptake and decarboxylase cells (APUD) that are normally distributed in the walls of tracheobronchial tree. This possibly leads to greatest frequency of SMCC in larynx in head and neck region. However, there is a controversy against this theory that small cell and epithelial cells are found in same combined tumor [8].

Therefore, current opinion is that extrapulmonary SMCC derives from multipotential stem cells that have capacity for divergent differentiation. This explains possible incidence of combined SMCC and squamous cell carcinoma within same tumor in some reported cases [8].

To date, there is still no established staging system for extrapulmonary SMCC. Two-stage classification used for small-cell lung carcinoma is used in extrapulmonary SMCC as well. The limited-stage illustrates SMCC restricted to the primary site and surrounding organs which can be encompassed within a tolerable radiation therapy port. SMCC that is beyond the extent of limited stage is considered extensive disease [9].

Treatment of SMCC depends on staging of the disease, and it is indistinguishable from small-cell carcinoma of the lung. There is no standard established guideline or the extrapulmonary SMCC due to limited experience. In general, for limited staged tumors, combined modality treatment has better survival, such as radiotherapy and surgery, radiotherapy and chemotherapy, or multidrug chemotherapy (carboplatin and etoposide). Multidrug chemotherapy is the mainstay of treatment for extensive disease, with the aim of palliative intent [10]. Due to the anatomical complexity of pyriform sinus with close proximity to the adjacent structures, surgical resection of the tumor require extensive debulking which would require prolonged postsurgical rehabilitation. With this, quality of life would be drastically impaired in view of loss of speech and deglutition post laryngopharyngectomy. Our reported case had a tracheostomy done to bypass upper airway obstruction, while chemotherapy was given for disease eradication. According to Lee J. H. et al. in 2016, a patient was successfully treated with concurrent chemoradiotherapy for primary pyriform sinus small cell carcinoma with cisplatin- and etoposide-based chemotherapy regime. Platinum-based chemotherapy may be preferable over surgical resection because of the high propensity of metastatic dissemination even in patient with clinically localized lesions [11, 12]. Primary prophylactic cranial irradiation is also advocated due to its risk of isolated cranial metastasis. Patients with untreated extrapulmonary SMCC have a dismal outcome with only weeks to 3-month median survival. With treatment, median survival is slightly prolonged to 8–16 months [13].

## Conclusion

Small-cell carcinoma in hypopharynx is a rare entity in head and neck region. Due to its distensibility of the pyriform fossa, it usually presents late. It responds well to combined multidrug chemotherapy such as etoposide- and platinum-based chemotherapeutic agents.

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## Authors' contributions

LTKN was involved in methodology and writing of original draft including literature review preparation. MWSL was involved in reviewing and editing the original draft. HSG was involved in overall supervision, reviewing, and

final editing of the manuscript. The authors read and approved the final manuscript.

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##### Ethics approval and consent to participate

Not applicable

##### Consent for publication

Written informed consent was obtained from the participant which include details, age, gender, diagnosis, images relating to the individual participant.

##### Competing interests

The authors declare that they have no competing interests.

##### Author details

<sup>1</sup>Department of Otorhinolaryngology, Head & Neck Surgery, Queen Elizabeth Hospital, Kota Kinabalu, Sabah, Malaysia. <sup>2</sup>Department of Otorhinolaryngology, Head & Neck Surgery, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Centre, 56000 Kuala Lumpur, Malaysia.

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