Small Cell Carcinoma of the Larynx
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ABSTRACT
Educational Objective: At the conclusion of this presentation, the participants should be able to understand the clinical and pathologic features of small cell carcinoma of the larynx. Treatment options should be discussed in order to manage this poor-prognosis cancer.
Objectives: Small cell carcinoma arises commonly in the lung and rarely in the head and neck, especially in the larynx. It tends to be very aggressive and metastasize widely, leading to very poor prognosis. We describe our experience with a systematic review and discuss treatment options of this rare tumor.
Study Design: Case report and literature review.
Methods: A case report is described with fiberoscopic images, positron emission tomography - computed tomography (PET-CT) and histopathologic images. A literature review is presented and treatment options are discussed to achieve better prognosis.
Results: A 70 year old man presented with throat discomfort. Fiberoptic laryngoscopy revealed ulcerative tumor in the right false vocal fold of the larynx. PET-CT demonstrated the tumor, invading the thyroid cartilage, and metastatic lymph nodes swelling in the right neck, however there was no evidence of metastasis in other organs. The patient underwent total laryngectomy and right modified radical neck dissection. Histopathologic analysis confirmed the diagnosis of small cell carcinoma of the larynx. Five cycles of chemotherapy (Cisplatin/etoposide) were delivered, according to the therapeutic regimen for pulmonary small cell carcinoma.
Conclusions: Small cell carcinoma of the larynx is a very rare and aggressive tumor with high rate and early tendency for distant metastases. Systemic chemotherapy should be delivered as soon as possible to prevent distant metastases. Surgical resection of primary tumor for locally advanced tumor without distant metastasis might be one of therapeutic options to achieve quick local control and deliver systemic chemotherapy immediately, leading to better prognosis.

INTRODUCTION
Small cell carcinoma arises commonly in the lung and rarely in the head and neck, especially in the larynx. It tends to be very aggressive and metastasize widely, leading to very poor prognosis. We describe our experience with a systematic review and discuss treatment options of this rare tumor.

CASE PRESENTATION
Clinical History
A 70 year-old man was seen in a private clinic with many-year history of hoarseness and a month history of throat discomfort. Fiberoptic laryngoscopy revealed a tumor in the larynx. He was referred to Japanese Red Cross Wakayama Medical Center for further examinations.
Previous History
Chronic thyroiditis
Smoking Habit
20 cigarettes/day x 40 years
Clinical Examination
Fiberoptic Laryngoscopy (Figure 1).
A tumor extended from the right false vocal fold to the laryngeal side of the epiglottis. The mobility of the right vocal fold was slightly limited.
Positron Emission Tomography - Computed Tomography (PET-CT) (Figure 2)
The supraglottic tumor (red arrow) showed high uptake of FDG and invasion of the thyroid cartilage. Level III and IV lymph nodes swelling (green arrow) in the right neck showed high uptake of FDG and suspicious invasion of the right internal jugular vein. There was no evidence of metastasis in other organs.
Biopsy
Suspicious of small cell carcinoma
Clinical Diagnosis
Laryngeal Cancer (Supraglottis), cT4aN2bM0

THERAPEUTIC STRATEGY
Need for airway management because of airway narrowing by the tumor
Locally advanced tumor (T4a) without distant metastasis (M0)
Need for quick delivery of chemotherapy if pathology confirms small cell carcinoma
Total Laryngectomy + Right Neck Dissection + Postoperative Chemotherapy

THERAPEUTIC COURSE
Operation
Total Laryngectomy
Right Neck Dissection (Level I-V)
Excised Specimen (Figure 3)
The tumor (red arrow) was found on the right false vocal fold.
Histology and Immunohistochemistry (Figure 4)
High N/C ratio, naked nucleus like cells
No keratinization
Thyroid cartilage invasion (-)
Definitive Diagnosis
Small Cell Carcinoma of the Larynx (Supraglottis)
pT3N2bM0, stage IVA
Postoperative Course
Five cycles of chemotherapy were delivered from three weeks after the operation, according to the therapeutic regimen for pulmonary small cell carcinoma (Cisplatin 80mg/m2 + Etoposide 100mg/m2).

DISCUSSION
Small Cell Carcinoma of the Larynx
Firstly reported in 1972 (Olofsson et al. 1972)
0.5% of laryngeal cancer
Age
Median age; 64 years old (rarely seen in patient younger than 40 years)
Gender difference
Male:Female = 3:1
Occurrence site
Supraglottis:Glottis:Subglottis = 6:1:3
Metastases at the initial visit
Neck lymph nodes: >50%
Distant organs: 10-20%
Overall survival (OS)
2-year OS: 36%, 5-year OS: 5% (2-year OS w/o chemotherapy: w/ chemotherapy = 10%: 52%)
Cause of death
Distant metastases (70%)
Therapy
w/ distant metastasis:
- Concurrent chemoradiotherapy + adjuvant chemotherapy
- T1: Surgical resection or concurrent chemoradiotherapy + adjuvant chemotherapy
- T2: Concurrent chemoradiotherapy + adjuvant chemotherapy
- T3, T4: Not established because of few reports

CONCLUSIONS
Small cell carcinoma of the larynx is a very rare and aggressive tumor with high rate and early tendency for distant metastases. Systemic chemotherapy should be delivered as soon as possible to prevent distant metastases. Surgical resection of primary tumor for locally advanced tumor without distant metastasis might be one of therapeutic options to achieve quick local control and deliver systemic chemotherapy immediately, leading to better prognosis.