Synchronous primary small cell carcinomas of the larynx and hypopharynx

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Abstract

Problem addressed
Poorly differentiated small cell neuroendocrine carcinoma of the larynx is the most common neuroendocrine tumor in the head and neck. However, small cell cancers of the hypopharynx have less commonly been described in the literature. Treatment methods rely upon chemotherapy and radiation, but regardless of primary site, prognosis for patients with these cancers remains very poor.

Methods and measures
We present a rare case and review the associated literature. Our patient had a remote history of left neck squamous cell carcinoma of unknown primary treated with left radical neck dissection and chemotherapy. He now presented with a right level II neck mass. PET scan revealed hypermetabolic activity in both the region of the mass and the right parapharyngeal space at the thoracic inlet. The patient underwent direct laryngoscopy which revealed synchronous primary tumors in the left subglottis and right posterolateral hypopharyngeal wall. Pathologic analysis revealed poorly differentiated small cell carcinoma at both sites. Chest CT showed no lesions.

Results
The patient was treated with four cycles of cisplatin-based chemotherapy. He had a strong initial response to treatment, with no clinical or radiographic evidence of neck disease two months following completion of therapy. However, at subsequent follow-up visits he developed a new right neck mass, and fine needle aspiration showed recurrent neuroendocrine carcinoma. He then underwent reirradiation to the neck and systemic chemotherapy.

Conclusions
This is the first report describing synchronous primary small cell carcinomas of the larynx and hypopharynx. Further efforts are necessary to better characterize these rare tumors with the hope of developing more effective treatment modalities.

Case Presentation

- CC: A 52 year old male presented with a right neck mass.

- HP: The patient denied dyspnea, dysphagia, voice change, or pain. He did note a 10-12 pound weight loss, and an extensive tooth extraction procedure completed three months prior to presentation. He had no history of smoking or excessive alcohol use.

- PMH: History was significant for left neck TxN2bM0 squamous cell carcinoma of unknown primary, treated with radiation therapy and left radical neck dissection in 1996.

- PE: A palpable right neck mass measuring 1-2 centimeters was present in the level II region. Flexible fiberoptic laryngoscopy showed mild post-radiation changes in the aerodigestive tract mucosa. No discrete masses or lesions were visualized and vocal cord mobility was normal.

- CT: Axial CT showed enlarged lymph nodes in the right level II region, a right paratracheal soft tissue mass just inferior to the thyroid gland, and right level V lymphadenopathy posterior to the internal jugular vein.

- CT Chest: No lesions were present.

- PET scan: PET showed hypermetabolic lymph nodes in both regions of the patient's palpable right neck mass and right parapharyngeal region at the thoracic inlet. FNA: Results indicated large atypical cells of unclear etiology; repeat FNA again showed atypical cells suspicious for poorly differentiated carcinoma.

- Interventions: Direct laryngoscopy and direct esophagoscopy was performed, intraoperatively, biopsies were taken of a smooth, broad-based mass in the left subglottic region, and a firm, friable, broad-based lesion in the right hypopharynx just above the upper esophageal sphincter.

- OR pathology: Results indicated poorly differentiated small cell neuroendocrine carcinoma in the left subglottis and right hypopharynx.

- Treatment: He received four cycles of cisplatin-based chemotherapy; a strong initial response to treatment was observed, with no clinical or radiographic evidence of neck disease two months following completion of treatment.

- Follow-up: The patient developed a new right neck mass in third month following completion of chemotherapy; fine needle aspiration showed recurrent neuroendocrine carcinoma. Whole body PET-CT indicated enlarged lymph nodes in the right parapharyngeal region, along the right internal jugular vein, and in the left supravacular area. Disease further progressed despite reirradiation to the neck and second-line chemotherapy, with additional follow-up CT scan indicating malignant encasement of the right common carotid artery and worsening of left supraclavicular lymphadenopathy. Further deterioration in clinical course necessitated tracheotomy for bilateral vocal cord paralysis, gastrostomy placement for severe dysphagia, and palliative esophageal stent placement for a tracheoesophageal fistula.

Discussion

Neuroendocrine carcinomas encompass a broad range of tumors occurring throughout the body. They may be classified by degree of differentiation, ranging from well-differentiated (carcinoïd tumor) to poorly differentiated small and large cell subtypes. In the head and neck, most neuroendocrine carcinomas arise from the larynx, and the majority of these cancers are of small cell histology. While chemotherapy may produce significant initial responses and slightly improved overall survival, recurrence is typical, and overall prognosis is poor. Median survival rates for laryngeal small cell cancer is in the range of 1 year, and overall 5 year survival rates are approximately 5%. Death is most commonly the result of distant metastases to liver, bone, lung, and brain.

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Conclusions

This report is the first to describe synchronous primary small cell carcinomas of the larynx and hypopharynx. Further efforts are necessary to better characterize these rare but aggressive cancers with the hope of developing more effective treatment modalities and improving patient survival.

References


