Primary Squamous Cell Carcinoma of the Thyroid

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Abstract
Primary squamous cell carcinoma of the thyroid represents less than 1% of all thyroid malignancies. Unlike other thyroid cancers, it is often fatal. We report a rare case of a male patient with primary squamous cell carcinoma of the thyroid. This paper emphasizes the importance of fine needle aspiration in diagnosis, the urgency of early diagnosis and the importance of multi-modality treatment.

INTRODUCTION
Thyroid cancer is the most common endocrine malignancy [1] and is the seventh most common cancer in women. [2] Typically, these malignancies are of papillary, follicular, or medullary histology, arising from normal tissues present in the thyroid. Patients will rarely manifest with primary squamous cell carcinoma of the thyroid even though the thyroid lacks squamous cell epithelium. Though this puzzling pathology accounts for less than 1% of thyroid malignancies, [3] it has an unusually high mortality making early diagnosis and treatment crucial for patient survival. We report a case of a male patient with primary squamous cell carcinoma of the thyroid and discuss the diagnosis, workup, epidemiology, treatment, and postulate etiologies of these unusual cancers.

DISCUSSION

At diagnosis, primary SCC is typically found invading the trachea, esophagus, and other surrounding structures. [6] At presentation, patients will most commonly complain of an anterior neck mass, dyspnea and dysphasia. [5] Diagnosis of SCC of the thyroid is typically through fine needle aspiration, [7] although a distinction between primary SCC of the thyroid and a secondary metastatic SCC cannot be made. Further investigation via PET-CT is necessary to make a further distinction. Immunohistochemistry of SCC demonstrates positive staining of p53, p63, cytokeratins 7-19, and squamous cell carcinoma antigen, in addition to negative staining for thyroid transcription factor (TTF) and calcitonin. [8-10]
Figure 1 PET-CT demonstrating increased uptake in the posterior aspect of the right lobe of the thyroid with no distant metastatic disease.

Figure 2 Radiation treatment plan for patient with right squamous cell carcinoma of the thyroid. The PTV is delineated in red and received 3700 cGy in 35 fractions. Avoidance structures included the left and right parotid glands, spinal cord, cochlea, larynx, eyes, and brainstem. The 5985 cGy isodose line is shown in light green; the 5040 isodose line in purple.
Mean survival time for patients with SCC of the thyroid is 9 months, with a 20% 3-year survival rate. [5] In comparison, the mean survival rate of patients with metastatic papillary carcinoma, the most common thyroid malignancy, is significantly higher at 76.6%. [11] The cause of death in the majority of SCC of the thyroid is airway compromise. [4]

There is currently no standard of care for treatment for primary SCC. Due to the aggressive nature of the disease, total thyroidectomy alone has led to minimal survival time. [12,13] Some studies have shown that patients have achieved greater longevity using aggressive regional surgery and post-operative radiotherapy, with reported survival up to 31 months. [14,15] However, no prospective trials have directly compared longevity using combination therapy versus thyroidectomy alone. In the present case, although the patient had gross residual disease post-operatively, he has exceeded the mean survival time with post-operative treatment consisting of high dose Cisplatin and radiation therapy with no residual or metastatic disease according to his latest PET-CT scan. This suggests that using a combination of Cisplatin-based chemotherapy, radiation and surgery would prolong patient survival, not unlike the data in the treatment of other cancers.

While the etiology of SCC of the thyroid remains uncertain, three theories have been postulated to explain the pathogenesis of this condition. The embryonic nest theory suggests that embryologic remnants, such as the thyroglossal duct, give rise to epithelial cells. [16] While the de-differentiation theory suggests that existing thyroid carcinoma of papillary, follicular, or medullary histology de-differentiates into SCC. [17] Another theory called the metaplasia theory hypothesizes that local inflammatory condition, such as Hashimoto’s thyroiditis, results in squamous metaplasia. [18] In the present case, since the patient has no past medical history of chronic inflammatory conditions, the embryonic nest theory and de-differentiation theory are more likely.

SCC of the thyroid is a rare condition that is often diagnosed late in the disease process, leading to low survival rates. Since the condition is aggressive and fatal, it is important to recognize the associated symptoms sooner in both male and female patients. In the present case, the tumor was discovered late in stage IVa, but nevertheless, the cancer is in remission. Part of this success can likely be attributed to the multi-modality treatments, suggesting that a multi-modal approach can increase survival and prevent recurrence even in late stages. Observing and documenting more patients with this rare malignancy will continue to develop our understanding of the etiology, diagnosis, and treatment of the disease in order to best care for our patients.

CONFLICT OF INTEREST

We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

We confirm that the manuscript has been read and approved by all named authors and that there are no other persons who satisfied the criteria for authorship but are not listed. We further confirm that the order of authors listed in the manuscript has been approved by all of us.

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REFERENCES


