A Middle-Aged Woman With a Thyroid Cyst: A Case of Squamous Cell Carcinoma of the Thyroid
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INTRODUCTION

• Primary squamous cell carcinoma of the thyroid (PSCCT) is a very rare malignant neoplasm of the thyroid with distinct squamous differentiation of tumor cells.

• Only 84 cases of PSCCT were reported worldwide up until 2012. This number has now increased to around 314. The incidence of PSCCT is less than 1% out of all thyroid malignancies, but it has one of the fastest increasing incidences of cancer worldwide.

• With no standard consensus to guide the management plan and a 3-year survival rate of 43%, PSCCT is challenging to treat.

• Here we describe a case of primary squamous cell carcinoma of the thyroid.

CASE SUMMARY

• A 65-year-old white female with a medical history of longstanding Hashimoto's thyroiditis on Levothyroxine was initially evaluated for left neck enlargement and compressive symptoms. There were no significant risk factors for malignancy.

• CT neck showed a left thyroid cyst measuring 4.4 x 7.0 cm.

• Two thyroid biopsies revealed cystic material and a cyst aspiration was not tolerated thus proceeded to left thyroidectomy.

• Surgical pathology showed moderately differentiated squamous cell carcinoma stage pT3aN0a, 4.5 cm without lymph node involvement. The cells stained positive for p40, Ck5/6, p63 and TTF1, but negative for thyroglobulin and calcitonin. Subsequently, completion thyroidectomy with central lymph node dissection resulted in benign pathology.

• Postoperative imaging was without evidence of residual neoplasm or pathologic cervical adenopathy.

• Levothyroxine was continued to maintain a TSH in the normal range.

• She completed 6 weeks of XRT.

• Oncology recommended supportive care due to the aggressive nature of this tumor subtype.

RESULTS

CT soft tissue neck with IV contrast

Surgical Pathology

Tumor demonstrates nests and trabeculae of pleomorphic eosinophilic cells with keratin pearls and a background of chronic lymphocytic inflammation (H&E, 50X) (A). H&E stain demonstrating Orphan Annie nuclei, characteristic of papillary thyroid cancer (B).

FOLLOW UP

• One year after surgery, a follow up CT scan of the neck showed a new enhancing lesion in the left thyroid bed concerning for local recurrence. Patient underwent excision of the neck lesion.

• Pathology showed a malignant sarcomatoid neoplasm. Immunostain positive for SMA with weak and focal positivity for PAX8. Ki-67 labeling is greater than 50%. This is thought to be related to the previous squamous cell carcinoma of the thyroid but needs genomic assessment to confirm the diagnosis.

DISCUSSION

• Typical presentation includes a rapidly increasing neck mass invading the adjacent structures with accompanying cervical lymphadenopathy. Obstructive symptoms related to the mass effect of the cancer may be present. There may be a longstanding history of goiter or thyroid diseases.

• PSCCT rarely stains for thyroglobulin or TTF-1 (thyroid transcription factor 1). There are no conclusions on the molecular profiles of the carcinoma due to the limitation of molecular studies.

• PSCCT is poorly responsive to radiotherapy and relatively resistant to chemotherapy. There is no role for radioactive iodine ablation or thyroid suppression as this carcinoma does not uptake iodine.

• Age at diagnosis, tumor grade, tumor size and presence of distant metastases are independent predictors for survival.

• Studies shows Lenvatinib (tyrosine kinase inhibitors) may show promise to extend survival.

• Immunotherapy has been approved in many cancers, including head and neck squamous cell carcinomas. Although no clinical trials have been done, immunotherapy should be studied in primary thyroid squamous cell carcinoma with residual or recurrent diseases.

CONCLUSION

• Primary squamous cell carcinoma of the thyroid is a very rare cancer of the thyroid with a mortality rate of 57% at 3 years.

• Overall survival rate, although poor, is dependent on the extent of the tumor resection and adjuvant radiotherapy/chemotherapy.

• Experts predict a 6 months average life expectancy with PSCCT.

• There are no official guidelines on the diagnosis and treatment of squamous cell carcinoma of the thyroid. Further research is needed to better manage this rare cancer.