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Neck and Neck: To Treat or Not to Treat - That is the Question

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Primary thyroid lymphoma (PTL) is a very rare thyroid malignancy that accounts for only 1% - 5% of all thyroid malignancies. PTL occurs in less than 3% of all non-Hodgkin's extra nodal lymphomas. Because of its rarity, PTL is a challenge to recognize pre-operatively and controversy exists regarding its management. Appropriate management is quite different from the treatment of other neoplasms of the thyroid gland. Management of PTL is based on the aim of trying to achieve the best therapeutic results while diminishing treatment-related morbidity rates. Primary thyroid lymphoma (PTL) is a very rare thyroid malignancy that accounts for only 1% - 5% of all thyroid malignancies. Because of its rarity, PTL is a challenge to recognize pre-operatively and controversy exists regarding its management. Appropriate management is quite different from the treatment of other neoplasms of the thyroid gland. Management of PTL is based on the aim of trying to achieve the best therapeutic results while diminishing treatment-related morbidity rates.

A 68-year-old female initially presented with dysphagia, hoarseness, and globus sensation in her throat. She underwent an ultrasound of her thyroid which showed a 5.1 cm solid hypoechoic right thyroid nodule. Excisional biopsy was suspicious for papillary carcinoma but the cellularity of the aspirated material was low making interpretation difficult. Excisional biopsy was recommended. Patient underwent total thyroidectomy with pathology demonstrating aggressive, and mature diffuse large B-cell lymphoma (DLBCL) of the thyroid with proliferation index (Ki-67) of 100% and extensive necrosis, focal EBER positivity, FISH negative for BCL6, MYC, and BCL2 gene rearrangement; PET/CT showed a probable inclusion cyst in the posterior right neck but was otherwise negative. Patient was therefore deemed stage IE bulky, NCCN-IPI low-intermediate risk group. Post-operatively, patient received 4 cycles of R-CHOP with post-chemotherapy PET/CT demonstrating no residual disease. She was offered consolidative radiation therapy – a total dose of 30Gy/15 fractions via daily image-guided IMRT photon therapy to her tumor bed and bilateral lymph nodes. At 5-month follow-up: patient continued to experience dysphonia, moderate dysphagia and xerostomia.

Factors in consideration include poor prognostic factors include tumor necrosis and proliferation index. The patient had chemotherapy, response to chemotherapy cannot be assessed since the patient had no measurable disease after thyroidectomy. It is unclear whether this patient's total thyroidectomy obviates the need for additional local therapy. If the decision is made to treat with radiation, controversy exists on nodal volumes etc. An overview from International lymphoma Radiation Oncology Group Guidelines recommends whole organ irradiation due to the limitations of small-volume disease extension in Primary extra nodal DLBCL after systemic therapy.

Overall, our recommendation for this patient local radiation to the neck to somewhat more generous fields then ISRT, since a pre-resection scan was not obtained. Thyroid lymphomas are not common and standard care is not well established. Generally, ISRT can be considered for bulky stage I diffuse large B-cell lymphoma. Continued discussion and research are needed to guide and evidence-based decision on extent of treatment fields and nodal volumes in these cases.