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Case Study

Primary mucosa-associated lymphoid tissue lymphoma of the thyroid with concomitant papillary carcinoma

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Abstract

Objective: Papillary thyroid carcinoma combined with thyroid mucosa-associated lymphoid tissue (MALT) lymphoma is exceedingly rare and there is no standard management.

Case report: We report a rare association of MALT lymphoma of thyroid in a 60-year-old woman with Hashimoto's thyroiditis along with an incidental focus of papillary carcinoma.

Conclusion: Patients with Hashimoto's thyroiditis are prone to develop other thyroid pathology, including rare tumours such as MALT lymphoma. The differential diagnosis for a neoplasm in such patients should be wide.

Keywords: MALT; papillary carcinoma; radiation therapy; thyroid lymphoma

INTRODUCTION

The incidence of non-Hodgkin's lymphoma (NHL) in the thyroid gland is <0·5 per 100,000,[1] and the mucosa-associated lymphoid tissue (MALT) type makes up <1% of thyroid malignancies.[2] MALT lymphoma is an indolent variant of NHL, and the recommended treatment and prognosis in the thyroid gland is similar to the standard of care in other sites. Although involved field radiation is the treatment of choice, surgery is also a viable option.[3] Although patients with Hashimoto's thyroiditis are at increased risk of acquiring thyroid lymphoma,[4] no study has reported thyroid lymphoma with other malignancies of the gland. We present the case of a Hashimoto's patient whose concomitant thyroid MALT lymphoma and papillary carcinoma provided several diagnostic and treatment challenges.

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CASE REPORT

A 60-year-old woman with a long-standing history of Hashimoto's thyroiditis presented with a painless lump on her right throat of 3 months' duration. She denied any dysphagia or hoarseness, or B symptoms. A 2 cm right thyroid mass was palpated on examination, without any palpable neck lymph nodes bilaterally. A mildly enlarged (right lobe $3{\cdot}5 \times 2{\cdot}5$ cm) and heterogeneous

thyroid without any focal masses or adenopathy was noted on ultrasonography (USG) and confirmed by computed tomography (CT)scan. Fine needle aspiration (FNAC) was non-diagnostic, with possible lymphoproliferative disease or thyroiditis. She underwent a right thyroid lobectomy. Her pathology showed MALT lymphoma, Hashimoto's thyroiditis and 4 mm papillary carcinoma, follicular variant, confined to the lobe. On immunohistochemical staining, cells were positive for CD20, focally positive for CD10 (cluster designation), Bcl 6 (B-cell lymphoma) and negative for Bcl 2. Atypical lymphoplasmacytic infiltrate with kappa light-chain restriction consistent with extranodal marginal zone MALT lymphoma was noted. The rest of the workup for lymphoma including CT scan of the neck, chest, abdomen and pelvis, peripheral blood flow cytometry, complete blood count, comprehensive panel and lactic acid dehydrogenase was negative. Serum thyroid stimulating hormone (TSH) confirmed existing thyroiditis and staged as IEA. A hypertrophic scar that developed 3 months later was excised. USG demonstrated residual thyroid tissue in the right lobe and an intact left lobe.

A multidisciplinary team discussion involved treating her with radiation therapy alone. She received 3,000 cGy in 20 fractions over 27 days, 150 cGy per fraction. Target area included right thyroid bed as well as left lobe and isthmus of the thyroid gland, and excluded neck lymph nodes. Dose was prescribed to 95% isodose line and antero–posterior portals were used. Mild erythema was noted over the involved field in addition to mild dysphagia secondary to esophagitis. Serial CT scans done every 6 months showed no evidence of disease. Predictably, thyroiditis progressively worsened with elevated TSH and dose of levothyroxine was increased. She was disease free at the last follow-up of 3 years.

DISCUSSION

NHL is diagnosed in 30 per 100,000 people per year in America.[5] Prognosis is contingent upon the histological subtype and stage. MALT lymphoma is typically a less aggressive subtype, and comprises 5% of all NHLs[6] and over 90% present in gastric mucosa or orbit.[7,8] The clinical presentation of MALT lymphoma varies with the site affected. In cases like ours, thyroid lymphoma may appear as a goiter and is either asymptomatic or can be incidentally discovered at workup for thyroiditis. Treatment of MALT lymphomas is also contingent upon the site involved. Definitive radiation to 25–30 Gy is recommended for early-stage (stage I and II) MALT lymphoma with 90% response rates[9,10] and late-stage disease with Rituximab and/or chemotherapy.

The thyroid gland is an uncommon site for MALT lymphoma. The higher incidence of lymphoma in patients with Hashimoto's is likely owing to the lymphocyte-mediated response of the auto-immune disease[11] and prognosis is generally very good. In a retrospective review of 171 thyroid lymphomas, 5-year overall survival for MALT lymphoma was 89%[12], with most cases of these treated with radiation therapy alone, although the treatment paradigm is not overwhelmingly clear. Proponents of surgery argue that an indolent MALT lymphoma may return in a more aggressive form, and thyroidectomy offers the best local control.[13] Other studies have shown that combined chemoradiation results in superior outcome for more aggressive lymphomas of the thyroid but makes no difference for indolent ones.[11]

This patient was offered a particularly challenging workup because her goiter presented in the setting of an already established Hashimoto's thyroiditis. Thyroid lobectomy was needed to diagnose her lymphoma as initial FNAC was non-diagnostic. Although the incidental papillary carcinoma was contained within the right lobe, there was debate about completion of thyroidectomy and neck dissection. The literature[14–16] does not dictate a treatment recommendation for combined papillary carcinoma and MALT lymphoma; however, Cheng et al.[14] profiled a successful treatment in a similar case in which the papillary carcinoma and lymphoma were treated independently. In our case, papillary carcinoma was already treated via wide margin excision, and ultimately the success rate of definitive radiation for thyroid MALT lymphoma drove her post-operative treatment. The patient has

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no evidence of disease at 3 years with no late toxicities.

CONCLUSION

Although rare, lymphoma should be included on the differential for a mass on the thyroid gland, especially in a patient with Hashimoto's thyroiditis. The typical workup for a thyroid mass includes USG and FNAC; however, that may not be enough to diagnose the neoplasm. A diagnostic and staging workup for lymphoma should ensue. In the event that a MALT lymphoma is discovered in the thyroid, radiation therapy alone has proven to be an effective treatment.

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Conflicts of Interest

None.

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