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Case Report

Rare Neoplastic Condition: How to Treat a Thyroid Lymphoma?

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Abstract

Introduction: Primary Thyroid Lymphoma (PTL), although a rare malignancy, can arise in common chronic inflammatory conditions such as Hashimoto's thyroiditis. Early detection of malignancy can play a vital role in improved outcomes.

Case Report: We report the case of a 61-year-old man who was referred to the emergency unit due to compressive cervical symptoms.

Fine needle aspiration cytology of the thyroid gland was performed, proving the presence of a dedifferentiated neoplasm (Tyr5).

The patient was submitted to a total thyroidectomy and histological examination revealed a diagnosis of extra nodal marginal B-cell lymphoma (MALT lymphoma) in a background of chronic lymphocytic thyroiditis.

Results: Thyroid gland does not contain lymphoid tissue. Under pathological conditions, the appearance of lymphocytes may occur promoting the further development of the disease. This close relationship is probably due to chronic antigenic stimulation leading to malignant transformation. In this clinical case, the previous diagnosis of Hashimoto's thyroiditis was not known, but the thyroid histological examination proved the coexistence of this autoimmune disease.

Conclusion: PTL is a rare disease and the pre-operative diagnosis is not easy. To achieve a permanent resolution of symptoms or a survival improvement, active surgical intervention is mandatory.

Keywords: Thyroid; MALT lymphoma; Thyroid cancer; Thyroiditis

Introduction

Incidental finding of a thyroid nodule with chronic thyroid inflammation warrants further investigation; primary thyroid lymphoma (PTL) usually arise in a context of chronic inflammatory conditions such as Hashimoto's thyroiditis. Primary thyroid lymphoma is a rare form of malignancy: it represents 2-8% of all thyroid malignancies and 1-2% of all extra-nodal lymphomas [1-3]. Early detection of malignancy can play a vital role to achieve surgical healing or relieve specific symptoms. Thyroid lymphomas are most commonly diffuse large B-cell lymphomas (60-80%) and only about 30% are extra nodal marginal zone lymphomas [1]. Primary thyroid MALT lymphoma is a rare condition, and represents 6-28% of PTLs [4]; usually arises in the setting of chronic lymphocytic thyroiditis [5].

Mucosa-associated lymphoid tissue (MALT) lymphoma is defined as extra nodal lymphoma composed of heterogeneous small B cells arising from marginal zone of MALT [6]. MALT lymphoma most frequently occurs in the gastrointestinal tract mucosa (50%), head and neck (15%), lung (14%), skin (11%), thyroid (4%), and breast (4%) [1,6]. These are usually found to arise at sites of ongoing chronic inflammation with underlying autoimmune or infectious etiologies [7].

Case Report

A 61-year-old man was referred to our emergency unit due to compressive cervical symptoms in January 2018. The cervical ultrasound revealed an enlarged thyroid gland with heterogeneous echo structure, revealing a nodular lesion occupying almost the entire right lobe, migrating to the mediastinum, correlated with a substernal goiter. Enlarged lymph nodes were noticed.

Fine Needle Aspiration Cytology (FNAC) of the thyroid gland was performed, proving the presence of a dedifferentiated neoplasm (Tyr5). The vocal cords were evaluated by laryngoscopy revealing normal vocal folds movements and partial involvement of the glottic lumen by arytenoid hypertrophy. TSH was measured and patient was in euthyroidism.

According to the diagnosis of Tyr 5 the patient was submitted to a total thyroidectomy with intraoperative findings of a globally hardened thyroid, showing a multinodular right lobe containing an enlarged nodule, with its inferior region located on the substernal region, strongly adherent to the trachea and esophagus with no evident cleavage plane. He had no post-operative complications and was then discharged on second day

The histological examination revealed positivity for CD20, CD79A, CD43, and CD4, partial effacement of the thyroid parenchymal architecture by a population of round-to-oval lymphoid cells, with large and pleomorphic nuclei, coarse nuclear chromatin, visible nucleoli and scarse eosinophilic cytoplasm. Infiltration of the follicular epithelium, creating lymphoepithelial lesions, was noticed.

With these morphologic and immunohistochemical findings, a diagnosis of extra nodal marginal B-cell lymphoma (MALT lymphoma) in a background of chronic lymphocytic thyroiditis, was made. The patient was not subjected to further terapie and three months after patient was disease free at follow up.

Discussion

As a rule, the thyroid gland does not contain lymphoid tissue. Under pathological conditions, the appearance of lymphocytes may occur promoting the further development of the disease. The presence of

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autoimmune chronic lymphocytic thyroiditis as Hashimoto's thyroiditis is a well-established risk factor, presenting a 40-80 fold increased risk of developing PTL when compared to the general population. According to some publications, Hashimoto's disease is associated with more than 90% of the PTL. This close relationship is probably due to chronic antigenic stimulation leading to malignant transformation [7]. In this clinical case, the previous diagnosis of Hashimoto's thyroiditis was not known, but the thyroid histological examination proved the coexistence of this autoimmune disease. The optimal treatment and follow-up remains controversial. The stage and histologic grade have a major role in determining prognosis of this disease. Diffuse large B-cell lymphoma has a poorer prognosis compared to localized primary MALT lymphoma [8].

The most frequently used approach is surgical excision for the localized disease, followed by radiotherapy and chemotherapy for disseminated disease [9,10]. However, some recent studies support radiation therapy as a first approach of treatment in patients at early stages and adjuvant therapy for those who are suspected to have some residual disease even after thyroidectomy procedure [1].

Conclusion

PTL is a rare disease and the pre-operative diagnosis is not easy even though the use of immunohistochemical and molecular techniques have improved the sensitivity of the results. To achieve a permanent resolution of symptoms or a survival improvement, active surgical intervention is particularly relevant if there are no other signs that the tumour has spread. Unfortunately, due to rare occurrence of PTL and lack of larger randomized controlled studies, definitive guidelines for treatment and follow-up on these patient groups remain undeterminate.

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