

Case Report

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Rare Localization of Lymphoma

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ABSTRACT

Objective: Primitive Thyroid Lymphomas (PTL) are rare tumors. Women in the sixth or seventh decade of life are more commonly affected. In the present study, we report a case of primitive thyroid lymphoma and we review the epidemiology, the clinical presentation, the diagnosis, and the treatment of this rare disorder.

Case Report: A 47-year-old woman presented to our department reporting a recent-onset neck mass since 3 months. Clinical examination revealed an enlargement of the thyroid gland with a 2.5 cm-firm left nodule. Cervical ultrasound was done. The patient had a thyroidectomy associated with bilateral Central Lymph Node Dissection (CLND). The diagnosis was a transformation of a Mucosa-associated lymphoid tissue (MALT) lymphoma into an aggressive Diffuse Large B-Cell Lymphoma (DLBCL).

Conclusion: The most common type of primary thyroid lymphoma (PTL) is diffuse large B-cell lymphoma, which behaves in a more aggressive manner than mucosa-associated lymphoid tissue lymphoma. Treatment and prognosis of PTL depend upon the histology and stage of the tumor at diagnosis.

KEY WORDS: Thyroid; Lymphoma; Mucosa-associated lymphoid tissue (MALT); Diffuse large B-cell lymphoma.

ABBREVIATIONS: PTL: Primitive Thyroid Lymphomas; CLND: Central Lymph Node Dissection; MALT: Mucosa-associated lymphoid tissue; DLBCL: Diffuse Large B-Cell Lymphoma; FNA: Fine Needle Aspiration; NHL: Non-Hodgkin lymphoma.

INTRODUCTION

Primary Lymphoma (PL) of the thyroid is a very rare disease. It is less than 2 to 5% of malignant neoplasms of the thyroid.¹ Involvement of the thyroid gland may occur in the context of a systemic disease or rarely be primitive. The most common histological sub-type is a diffuse large B-cell lymphoma (DLBCL) followed by Mucosa-associated lymphoid tissue (MALT) lymphoma.

CASE REPORT

A 47-year-old woman presented to our department reporting a recent-onset neck mass since 3 months. There were no local obstructive symptoms associated such as dyspnea or dysphonia. Her medical past history was unremarkable. She had no history of personal or family thyroid disease or radiation exposure. Clinical examination revealed an enlargement of the thyroid gland with a 2.5 cm-firm left nodule. No cervical nodes were palpable.

Cervical ultrasound showed multinodular goiter with hypoechogenes heterogeneous nodules containing microcalcifications. Infracentimetric lymph nodes in levels II, III, IV of the both lateral neck were detected. The patient was in biological euthyroidie. She underwent thyroidectomy associated with bilateral central lymph node dissection (CLND).

On macroscopic appearance, the thyroid gland was solid, nodular white-gray colored

with a fish-flesh. On microscopic examination, the findings showed a marked diffuse lymphocytic infiltration destroying thyroid follicles. Large lymphatic cells with a typical and large nuclei containing central nucleoli were observed.

Elsewhere, we found a typical lymphocytes with small and centrocyte-like cell. The neighboring thyroid parenchyma contained lesions of thyroiditis. The metastasis of central neck node was evident. Immunohistochemistry showed CD20 positivity and CD15, CD3, CD20 negativity. The diagnosis was a transformation of a MALT lymphoma into an aggressive diffuse large B-cell lymphoma (Figure 1).

Total body Computed Tomography (CT) scan was realized to complete staging according to Ann Arbor classification. It did not document any pathological finding. The diagnosis of primitive thyroid lymphoma was then made. The patient was at stage IIE. A combination of chemotherapy and radiotherapy treatment was adjuncted post-operatively. No tumor recurrence was observed after a mean follow-up of five years.

DISCUSSION

Primitive thyroid lymphoma (PTL) represents 2 to 7% of all extranodal primitive lymphomas. It mainly occur in middle- to older-aged patients with a predilection for females in the 6th decade of life.²

Most of the patients with PTL have a previous history of auto immune thyroid it is with or without hypothyroidism. In fact, Hashimoto’s thyroiditis co-exists in 83% of patients with PTL. Furthermore, in patients affected by chronic autoimmune thyroid it is, the probability of developing a PTL is 20 times greater than in the general population.²

Non-Hodgkin lymphoma (NHL) is the most common PLT (93%). Two sub-types are frequent: Diffuse large B-cell lymphoma is the most encountered accounting for more than 50% of cases, followed by mucosa-associated lymphoid tissue (MALT) lymphoma.³ Upto 40% of all diffuse large cell lymphomas appear to have undergone transformation from a MALT lymphoma.⁴

Clinically, general symptoms associated with lymphomas, such as fever, excessive perspiration and weight loss, are present in only 10-20% of patients.² A rapidly growing (usually within 1-3 months), painless thyroid enlargement, either in the form of goiter or discrete nodule, is the most common clinical presentation in PTL.

Diffuse large B-cell lymphoma (DLBCL) thyroid lymphoma is considered as a high grade lymphoma with a more aggressive clinical course. They present as a painless fast-growing mass causing compressive symptoms like dysphagia, hoarseness or dyspnea. These symptoms overlap with that of the anaplastic thyroid carcinoma.⁵

MALT lymphoma is considered low grade tumor with an indolent natural history and presents as a slow-growing tumor with early stage disease confined to the thyroid. DLBCL can develop from MALT lymphoma, and these two subtypes can be detected in the same gland. The mixed MALT and DLBCL sub-type shows the same clinical behavior as that of DLBCL.⁶

The diagnosis of PTL is not always evident. In fact, due to their rarity and clinical polymorphism the diagnosis is often made on definite histology after thyroid surgery as the case of our patient. Effectively, Fine Needle Aspiration (FNA) results are in-consistent due to the histopathological similarities between primary thyroid lymphoma and Hashimoto’s thyroiditis.⁷

Once a diagnosis is made, total body CT scan should be performed to complete staging, according to the Ann Arbor classification. Large series of PTL revealed that about 50% of cases is confined to the gland (stage IE), 45% involved the gland and regional lymph nodes (stage IIE). lymph node involvement above and below the diaphragm (stage IIIE) or extranodal disease (stage IV) are found in only 5% of cases.^{2,4}

Treatment depends on the histological subtype and the stage of the disease. Despite controversy regarding the optimal modality for the management of PTL, the combination of chemotherapy and locoregional radiotherapy is the standard treatment of localized aggressive lymphoma diffuse large B-cell lym-

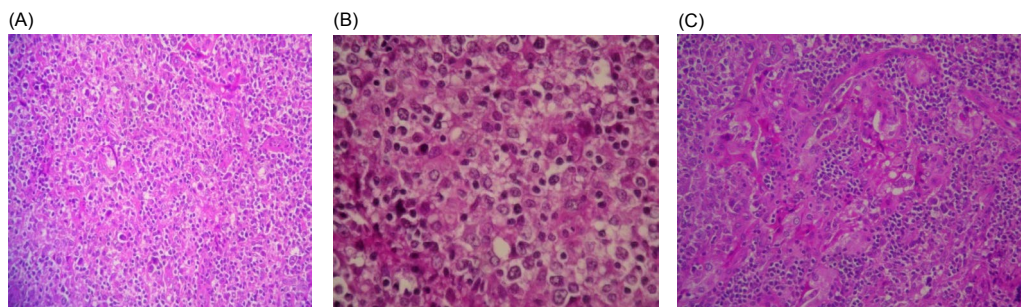


Figure 1: Microscopic finding of the thyroid mass showing. A) Marked lymphocytic infiltration of the thyroid parenchyma. B) Infiltration is made by large lymphocytes with voluminous nuclei. C) Lymphoid infiltration destroying follicular thyroid realizing lymphoepithelial lesions.

phoma.⁸ The conventional chemotherapeutic regimen consists of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP), and radiotherapy is used for local disease control.⁶

Surgery is the primary treatment of localized MALT lymphomas^{1,9} in disseminated or aggressive disease, surgery may be indicated for alleviation of compressive symptoms or protection of the airway.⁹ Surgical dissection may be more complicated than in standard cases of thyroidectomy due to the possible tight adhesions existing between the gland's capsule and the surrounding structures.²

The prognosis of PTL is affected by disease stage. In localized tumors, it is usually favorable with a survival rate at 5 years from 70% to 80%.¹⁰ However, the prognosis is very poor for lesions with extracapsular invasion (IIE) (20 to 50%). For stages IIIIE and IV, the rates are 15 to 35%.²

CONCLUSION

In summary, PTL has excellent prognosis when it is confined to the regional neck area and treated properly according to histologic sub-type and stage. The diagnosis should be early evoked in abrupt thyroid enlargement or compression symptoms. Treatment of PTL requires a multidisciplinary approach in order to choose the most appropriate therapy.⁴

CONFLICTS OF INTEREST: None.

CONSENT

The authors article did not publish any personal photo or information regarding any of the patients in his manuscript. Thus, the consent is not required for the article publication.

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