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Primary thyroid lymphoma – case report

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Abstract: *Introduction. Even though, primary thyroid lymphoma (PTL) is a rare thyroid malignancy and extranodal non-Hodgkin lymphoma, this type of disease should not be disgraced in order to achieve an early diagnosis and multimodal treatment.*

Materials and method. We selected for discussions a primary thyroid lymphoma patient with absent constitutional (B-type) symptoms, who undergoes a multidisciplinary approach of the disease, chemotherapy and radiotherapy. In this clinical case report a 84 yo male patient with stage IE diffuse large B-cell histological type (Ann Arbor staging) was treated using R-CHOP chemotherapy, followed by involved field radiotherapy, with an excellent treatment response and disease control.

Results and Discussion. Usually this type of extranodal non-Hodgkin lymphoma is present in the seventh decade of life, with males being affected earlier than females. One of the most common presentations of thyroid lymphoma is an enlarging neck mass and a history of Hashimoto's thyroiditis, which is the major risk factor. The most common PTL subtypes are diffuse large B-cell lymphoma (the most aggressive histological subtype, representing 70% of all PTLs) and MALT lymphoma (30% of the PTLs). Advances in both diagnosis and treatment in the last years have improved the early diagnosis and shortened the time for treatment implementation.

Keywords: *primary thyroid lymphoma; non-Hodgkin lymphoma; thyroid cancer; radiotherapy*

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INTRODUCTION

Even though lymphoma is one of the most common hematological malignancies, primary thyroid lymphoma (PTL) is very rare and accounts for 0.5-5% of all thyroid malignancies and about 2% of extranodal lymphomas [1]. Chronic Hashimoto's thyroiditis it is one of the main risk factors known to increase incidence of this type of malignancies [2]. PTL is a rare extranodal lymphoma which is defined as lymphoma that involves just the thyroid or the thyroid and the regional lymph nodes, without metastasis to other areas at the time of diagnosis [3].

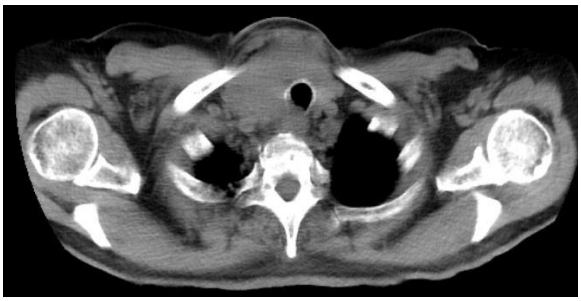
Treatment and prognosis varies according to history and clinical stage. Surgery, chemotherapy, radio-therapy or their combination are the treatment of choice in this particular type of Non-Hodgkin Lymphoma (LNH). Such a case, with PTL, is presented in this article [4].

CASE REPORT

An 84 years old man, known with Hashimoto's autoimmune thyroiditis, with no other relevant personal or family history, visits the oncology department for recent-onset gross neck mass, throat

discomfort, dysphagia and hoarseness (noticed about 1 month before presentation). The patient had no fever, night sweats or weight loss. The physical examination reveals firm, immobile and painless mass descending in the anterior chest wall. Thyroid hormones at presentation revealed high antithyreoglobulin antibodies, an increased thyrotropin level and normal thyroxin. The ultrasound showed a macronodular goiter and chronic lymphocytic cells. On the CT examination of the head and neck region, showed an enlarged thyroid gland, with a nodular structure, starting from the hyoid cartilage and ending in the upper mediastinum (Figure 1). The endotracheal biopsy and immune histological analyses led to the diagnosis of a Non-Hodkin diffuse large B-cell lymphoma (DLBCL), CD20+, Ki-67>90%. A bone biopsy was requested for disease staging, which showed no evidence of neoplazic cells on the examined tissue. Also, the CT scan of thorax, abdomen and pelvis showed no other signs of evolution.

Figure 1 (explication in text)

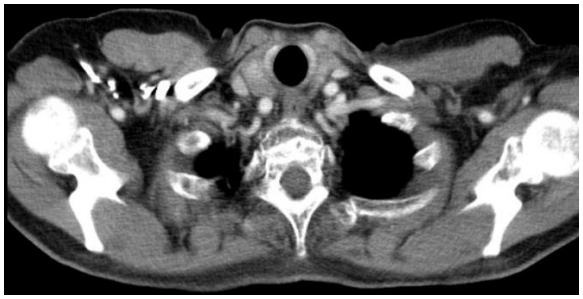


Regarding the fact that only the thyroid was involved (which is an extranodal site, on the same side of the diaphragm) and the fact that the patient had no B symptoms (fever, night sweats or weight loss), the stage of the disease, at the time of the diagnosis, was stage I AE.

After the diagnosis, the patient underwent 4 cycles of chemotherapy with R-CNOP chemotherapeutic protocol, i.e. rituximab (anti CD-20 antibody) 1400 mg, mitoxantrone 14 mg, cyclophosphamide 1350 mg, vincristine 2 mg and dexamethasone 16 mg. The treatment, after 4 cycles of chemotherapy, was stopped because of low tolerance. At the PET-CT scan, following chemotherapy was highlighted high metabolic activity in the right thyroid lobe.

Radiation therapy (RT) was recommended after chemotherapy. A total dose of 36 Gy (18 fractions, 2Gy/fraction) was administrated, using involved field technique to delineate the clinical target volume (CTV). For treatment preparation a fusion between PET-CT and planning CT-scan had to be used for accurate CTV delineation. Three months after radiotherapy, another PET-CT scan was made, to evaluate treatment response, showing low metabolic activity in the right thyroid lobe. The follow-up at 9 months post-chemoradiation was done using clinical examination and a CT scan, which showed a marked regression of the tumor mass (Figure 2).

Figure 2 (explication in text)



DISCUSSIONS

In general population this type of disease is rare, with an annual incidence rate of 2.1 persons per milion [6]. Also, it is known that PTL affects mainly women (5x times more common in women than men) between 50 and 80 years of age, whereas its incidence is low in individuals younger than 40 years of age [7].

We presented a patient with chronic lymphocytic thyroiditis (Hashimoto's thyroiditis), high levels of antithyroglobulin antibodies in different blood tests. These types of patients, with Hashimoto's disease, have a much higher risk of PTL than the general population. It has been presumed that abnormalities in immunity due to an approximately 20-year history of Hashimoto's disease increase the risk of developing PTL [8].

Clinical findings in our patient on presentation, made us consider an anaplastic carcinoma of the thyroid gland, because of the overlapping symptoms. After tumor and bone biopsy, the initial assumption was changed into primary thyroid B-cell lymphoma. The

bone biopsy was made to exclude systemic involvement [9].

The current standard of care is chemotherapy followed by radiotherapy. The role of surgery has been diminished, because an aggressive debulking surgery would not have a more successful response over chemo-radiotherapy [10]. The recommended treatment was 6 cycles of R-CHOP followed by adjuvant RT [5]. In our case it was used R-CNOP chemotherapy because of mitoxantrones lower cardiotoxicity than doxorubicin, but with the same therapeutic outcome as the R-CHOP scheme [11]. RT treatment plan was realized by applying involved field radiotherapy technique, which included in the CTV the whole thyroid gland. A total dose of 36 Gy (2 Gy/fraction, 18 fractions) was administered at the end [7, 12].

An excellent prognosis for early stage PTL (stage IE and stage IIE) is achieved with proper modern treatment management and a multidisciplinary approach. A 5-year survival rate of 87.5% has been observed in patients with DLBCL type. Using modern treatment (combination of chemotherapy plus rituximab agent and radiotherapy), a good response is observed in this type of patients [7].

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