

Case Report

Dysphagia Revealing Primary Large B-Cell Thyroid Lymphoma

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Abstract

Primary thyroid lymphoma is a rare and uncommon disease accounting for about 0.5-5% of all thyroid cancers and about 2% of extra-nodal lymphomas. It complicates in the majority of cases a preexisting or concomitant Hashimoto's thyroiditis, and represents often a real diagnostic challenge.

Classically the main sign of this cancer is the rapidly enlarging anterior cervical mass. Associated compressive symptoms such as dysphagia are usually seen in advanced cases with large tumors or invasion of adjacent structures. Primary thyroid lymphoma remains, however, an exceptional and unusual etiology of dysphagia, and revealing forms are uncommon. We report an original case of primary thyroid lymphoma, without underlying Hashimoto thyroiditis, and revealed by dysphagia.

Keywords: Dysphagia; Lymphoma; Thyroid; Thyroid Lymphoma

Introduction

Primary thyroid lymphoma is a rare and uncommon disease [1,2], accounting for about 0.5-5% of all thyroid cancers and about 2% of extra nodal lymphomas [3]. It complicates in the majority of cases a preexisting or concomitant Hashimoto's thyroiditis [1,4], and represents often a real diagnostic challenge [1,5]. Classically the main sign of this cancer is the rapidly enlarging anterior cervical mass [1,4]. Associated compressive symptoms such as dyspnea, dysphagia, and hoarseness can be helpful for the clinical suspicion of this lymphoma [1,4].

Primary thyroid lymphoma remains, however, an exceptional and unusual cause of dysphagia [6]. Dysphagia induced by these cancers is usually seen in advanced cases with large tumors

or invasion of adjacent structures [1,4]; however, dysphagia has been reported even in small thyroid lymphomas without any clinically palpable goiter or thyroid mass [6]. A biopsy is needed for diagnostic and to identify the subtype of lymphoma. A multidisciplinary approach including radiotherapy and/or chemotherapy can eradicate the disease [1-6]. We report an original case of primary thyroid lymphoma, without underlying Hashimoto thyroiditis, and revealed by dysphagia.

Case Report

A 79-year-old male patient with no pathological medical history was explored for recent upper dysphagia with dysphonia. There were no reports of toxic substances use, recent neck trauma, fever, or associated dyspnea.

Somatic examination revealed an enlargement of the thyroid gland and multiple bilateral cervical lymphadenopathies (Figure 1).



Figure 1: Clinical examination: enlargement of the thyroid gland and multiple bilateral cervical lymphadenopathies.

A cervical CT scan was performed showing a multinodular goiter associated with bilateral cervical lymphadenopathy (Figures 2 and 3). Basic biology, particularly thyroid tests, was within normal limits.



Figure 2: Axial cervical CT scan: heterogeneous enlargement of the thyroid gland with bilateral cervical lymphadenopathies.

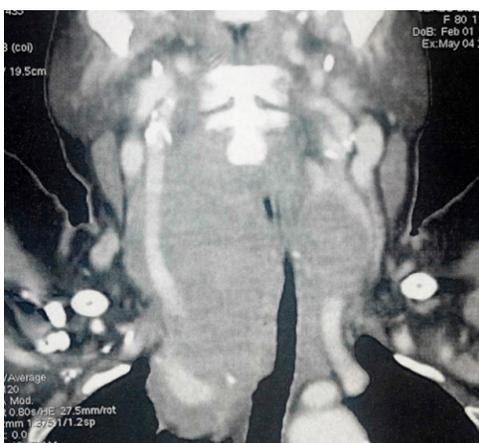


Figure 3: Coronal cervical CT scan: asymmetrical and heterogeneous enlargement of the thyroid gland.

A fine needle aspiration revealed undifferentiated cells with a carcinomatous appearance. He underwent thyroidectomy but presented intraoperatively a cardiopulmonary arrest. The patient had recovered, so a tracheostomy with the biopsy of the tumor were practiced and he was hospitalized in the intensive care unit. istopathological examination concluded to a large B-cell lymphoma. The patient died postoperatively.

Discussion

Primary thyroid lymphomas are rare among thyroid cancers [1,2,7]. Non-Hodgkin's large B cell lymphoma is the most common subtype (approximately 50-80%) followed by mucosa-associated lymphoid tissue MALT (approximately 20-30%), whereas follicular lymphomas, Hodgkin's disease, small lymphocytic lymphoma and Burkitt's lymphoma are much rare [8]. Thyroid lymphoma is more common in women and the mean age is 60 years at initial presentation [9,10]. The incidence of underlying Hashimoto's thyroiditis has been reported to be 80%, and patients may take years to develop lymphoma [11]. Sjögren's syndrome can be also associated with thyroid lymphoma, especially MALT subtype [12].

The diagnosis of thyroid lymphoma is evoked in case of a rapidly growing painless thyroid enlargement either in the form of discrete nodule or goiter; that may be associated with signs of compression, cervical lymphadenopathies and recurrent paralysis [1-6]. General symptoms such as fever, night sweats and weight loss, are less common, and signs of hypothyroidism can be present in 40% of cases [9,11].

Dysphagia, as a symptom associated with thyroid lymphoma, usually occurs in advanced or bulky forms that compress or invade the esophagus [1,2,4]. Indeed, in the Mayo Clinic's broad series of seventy-five patients with biopsy-proven thyroid lymphoma, dysphagia was noted in 45.3% of cases [1]. Revealing forms remain unusual.

Cervical ultrasound shows a heterogeneous hypoechoic mass, and allows searching for associated cervical lymphadenopathy. CT-scan of the head, neck, chest, abdomen, and pelvis is very useful in order to stage the patient according to the Ann Arbor classification. Fine needle aspiration is useful to guide the diagnosis, but a surgical biopsy is needed to confirm it and determine the subtype [11-14].

Anaplastic carcinoma of the thyroid is the main differential diagnosis that must be recognized because prognostic and management varies [15]. Appropriate treatment for patients with thyroid lymphoma depends on histological subtype and stage of the disease. Surgery is essential for diagnostic but is also indicated in the case of tracheal compression and can be also propose alone to manage a localized MALT lymphoma (stage IE and IIE) [16]. Standard treatment is based on radiotherapy, chemotherapy or the combination of both, and depends on the histological type of the lymphoma and the stage of the disease. The classical chemotherapy regimen is based on the CHOP protocol (cyclophosphamide,

doxorubicin, vincristine, prednisolone), combined with rituximab in the CD20 + forms [10,16].

The most important prognostic factors are the histological subtypes and the stage of the disease; thus, the five-year life expectancy for large B-cell lymphoma is 75% and it is 96% for MALT lymphoma. For the IE stage the survival is 86% and it is 64% for the IIIIE and IVE stages. Other prognostic factors are evoked such as **age, stage, tumor type and size, lymph node involvement, treatment selection, mediastinal involvement, and presence of B symptoms** [17].

Dysphagia is an important symptom to evaluate in patient with thyroid lymphoma because it was the only presenting symptom that was significantly associated with prognosis in the series of Penney SE & Homer JJ of 63 patients treated for thyroid lymphoma ($p = 0.001$) [2].

Conclusion

Thyroid lymphoma is a rare entity that can occur in patients with autoimmune thyroiditis and should be suspected in any patient with rapidly growing thyroid enlargement. Management is based on surgery, chemotherapy and radiotherapy. Dysphagia is a classic symptom in advanced forms of this cancer, but remains unusual as a revealing manifestation. It is also an important prognostic factor to consider in these patients.

Conflicts of Interest

None.

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