Primary lymphoma of thyroid: A case report

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The clinical and pathologic spectrum of lymphoproliferative disorders affecting the thyroid is diverse. Primary lymphoma is an uncommon malignancy of the thyroid, comprising between 0.6 and 5 per cent of thyroid cancers. The majority of thyroid lymphomas are diffuse large cell of B-cell origin. In this case, a 43 year old woman has been diagnosed to have thyroidal lymphoma, marginal zone type, originating from the underlying Hashimoto’s thyroiditis. [Turk J Cancer 2001;31(1):39-43]

Key words: Primary thyroid lymphoma, MALT, Hashimoto’s thyroiditis

Most cases of primary malignant lymphoma of the thyroid are seen in middle-aged or elderly females. Malignant lymphomas are more common in female than males (3:1) (1). The thyroid enlargement is often rapid and can lead to symptoms of tracheal or laryngeal compression. The clinical presentations include an enlarging neck mass but patients may also present with symptoms of dysphagia, hoarseness and choking (2,3). Most patients are euthyroid, and the tumor presents as one or more cold nodules on thyroid scan. Grossly, the tumors show a solid white cut surface with a fish-flesh appearance. It has been suggested that most low-grade lymphomas of the thyroid belong to the MALT (mucosa associated lymphoid tissue) category; their sometimes follicular pattern of growth is said to be result of colonization of the follicles by the tumor cells. Low-grade lymphomas composed of small or "intermediate" lymphocytes, having either a diffuse or a nodular (follicular) pattern of growth (2). Immunoblastic sarcoma of the B cell type is the second most common type. These tumors can also occur in both the diffuse or nodular form (1).

Case Report

A 43 year-old woman (7691/99) who is euthyroid and has bilaterally enlarged thyroid lobes has been found to have focal hyperactivity and diffuse hypoactive reactivity in both of the thyroid lobes after thyroid scintigraphy. She underwent total thyroidectomy. Grossly, it was a 140 gram and 14x8x3 cm. specimen. The cut surface was grayish-white and has a smooth and fish-flesh
surface. The H&E stained sections show chronic lymphocytic thyroiditis with large portions of the thyroid having effaced architecture (Figure 1). The thyroiditis was present at one end of the slide with also maintenance of the thyroid follicle architecture. However, most of the specimen showed a diffuse lymphocytic infiltrate composed of monocytoid lymphocytes with prominent plasma cell differentiation (Figure 2). There was epithelial thyroid follicle destruction. These findings are consistent with lymphoma originating from underlying Hashimoto’s thyroiditis.

Fig 1. Microscopic appearance: light microscopy demonstrated chronic lymphocytic thyroiditis and large portions of the thyroid with effaced architecture (H&E x200)

Fig 2. Microscopic appearance: thyroid gland showed a diffuse lymphocytic infiltrate composed of monocytoid lymphocytes with prominent plasma cell differentiation (H&E x400)
Immunohistochemical stains for LCA, CD3, CD43, CD20, calcitonin, thyroglobulin, cytokeratin, lambda and kappa light chains have been applied. Thyroid follicle epithelial cells stained for thyroglobulin (Figure 3) and the majority of the lymphocytes stained for LCA and CD20 (Figure 4).

Combination chemotherapy consisted of cyclophosphamide, vincristine and prednisone (CVP, 6 cycles) was administered after the surgery. As total thyroidectomy was performed, the patient did not receive radiation therapy. The patient stayed in remission within 6 months follow-up.

Fig 3. Microscopic appearance: epithelial cells of thyroid follicle with thyroglobulin exhibit consistent positivity (Thyroglobulin x100)

Fig 4. Microscopic appearance: the majority of the lymphocytes with LCA exhibit consistent positivity (LCA x400)
Discussion

The great majority of primary thyroid lymphomas are B cell lymphomas which are associated with Epstein-Barr Virus (EBV) and Hashimoto’s thyroiditis (4,5). Extranodal low-grade B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) type occurs in the gastrointestinal tract, salivary gland, thyroid, orbit, lung, and breast (6). MALT type lymphoma is a distinct subgroup of non-Hodgkin’s lymphoma with particular clinicopathologic behaviors. This type of lymphoma tends to appear in patients with a history of autoimmune disease or chronic inflammatory disorders. The preceding lymphoid disorders are usually the Sjögren’s syndrome, Hashimoto’s thyroiditis, and interstitial lymphoid pneumonia in the salivary glands, the thyroid or lung respectively. Lymphoma initially arise from the marginal zone localized around the reactive follicles and secondarily invade epithelial tissue to form the characteristic lymphoepithelial lesion (7). Patients with gastrointestinal MALT lymphoma generally present with localized disease. These patients have long survival rates. Recurrences may rarely appear in the same organs or in other extranodal sites. Nongastrointestinal MALT lymphoma patients have also a similar disease characteristics.

The role of surgery in the treatment of stage I non-Hodgkin’s lymphoma is not well defined (8). Patients may be treated with surgery or radiotherapy if the disease is localized, or with chemotherapy if it is disseminated (7). The treatment of thyroid lymphoma usually includes thyroidectomy followed by adjuvant therapy in cases with extrathyroid involvement. Primary thyroid lymphoma should be distinguished from systemic lymphoma with thyroid involvement, a somewhat unusual event (2).

An important diagnostic finding is the packing of follicular lumen by lymphoid cells, a feature usually not present in thyroiditis. The differential diagnosis includes the small cell variant of medullary carcinoma and insular carcinoma, which may require the use of immunohistochemistry. A very high proportion of the primary thyroid lymphomas arise in a background of lymphocytic or Hashimoto’s thyroiditis, this association representing the pathogenetic link between immunoproliferative and autoimmune diseases. The tumor is usually restricted to the thyroid, but also may spread to the soft tissues by direct extension, or may involve the regional nodes. The prognosis is better for intrathyroid tumors than for those that have extended beyond the thyroid capsule and for large cell lymphomas than for immunoblastic lymphomas. It is also not uncommon for the recurrence to be located in the gastrointestinal tract.

As a conclusion, lymphoma of thyroid is uncommon. The prognosis for localized thyroid lymphoma is excellent. Chemotherapy/radiotherapy regimens have proven to be very effective for most thyroid lymphomas. In the present case there was no relapse observed after a 6-month follow-up.
References