Thyroid Lymphoma

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Citation

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Abstract

Background: Thyroid lymphoma and anaplastic carcinoma, diseases have differing management approaches and prognosis, and are at times difficult to differentiate. Case report: We report a man in the seventh decade presenting with a rapidly enlarging neck swelling. Both anaplastic carcinoma and lymphoma were considered. Fine-needle aspiration reported lymphocytic thyroiditis and suggested further evaluation to rule out lymphoma of the thyroid. Open surgical biopsy with on-table frozen section biopsy was done which was unable to rule out the possibility of anaplastic carcinoma. Histopathology with immunocytochemical techniques confirmed the diagnosis of Non Hodgkin's Lymphoma of the thyroid, suggestive of MALT-(Mucosa Associated Lymphoid Tissue)-oma with Hashimoto's disease as background.Conclusion: Differentiation of thyroid lymphoma from anaplastic carcinoma, clinically as well as histopathologically, may be difficult. Though fine-needle aspiration cytology is the investigative modality of choice, surgical biopsy is still important and relevant. We stress the usefulness of immunocytochemistry in diagnosis.

CASE REPORT

A sixty-two-year-old male presented with a diffuse rapidly enlarging neck swelling of two months duration and hoarseness of voice for a fortnight. Thyroid profile revealed subclinical hypothyroidism. Neck ultrasonography showed a 6.9x5.8cm, predominantly hypo-echoic mass lesion involving the right lobe and isthmus of the thyroid gland with retrosternal extension. The carotids were displaced laterally. Fine needle aspiration cytology (FNAC) reported florid lymphocytic thyroiditis with atypical lymphoid cells. Follow-up and further evaluation advised to rule out lymphoma.

In view of recent detection, rapid enlargement and FNAC report, open surgical biopsy with intraoperative frozen section biopsy was carried out which reported an atypical round-cell infiltrate suggestive of a lymphoma, though anaplastic carcinoma could not be excluded. Hence, we proceeded with total thyroidectomy. Histopathology of the biopsy specimen was reported as Non-Hodgkin's Lymphoma (NHL) of the thyroid, suggestive of MALT-(Mucosa Associated Lymphoid Tissue)-oma with Hashimoto's disease as background. Immunohistochemistry (IHC) of the tissue showed CD20 to be positive (4+), and CD3, CD5, CD10, BCL2 and CDK to be negative.

During further postoperative evaluation, blood counts, upper GI endoscopy, echocardiography and computed tomography of chest and abdomen were normal. Serum LDH levels were normal. The bone marrow showed a single nodular collection of lymphoid cells suggestive of infiltration by NHL Cd 20-95 %. The patient was started on chemotherapy with a CHOP + rituximab regime.

DISCUSSION

Primary thyroid lymphoma represents approximately 1-5% of thyroid malignancies and less than 2% of extranodal lymphomas. It continues to produce diagnostic and therapeutic dilemmas.[1]

In a rapidly enlarging thyroid gland, difficulty in distinguishing thyroid lymphoma from anaplastic thyroid carcinoma has reduced due to new immunocytochemical staining techniques.[2]

Thyroid lymphoma is more common in women than in men (3-4:1). Most patients present in the seventh decade of life. Approximately 30%-50% of patients have symptoms and signs of compression at presentation.[1] Hypothyroidism has been observed in 30-40% of patients. Hashimoto's thyroiditis appears to be a risk factor though the association is not completely understood.[3]

Thyroid lymphomas can be divided into non-Hodgkin's

lymphomas (NHL) of the B- and T-cell types and Hodgkin's lymphomas. Mucosa-associated lymphoid tissue (MALT) lymphomas are a subset of B-cell NHL. According to the revised European-American lymphoma classification and the World Health Organization classification of hematopoietic and lymphoid tissue neoplasms, they are marginal zone B-cell lymphomas. Pathologically, most thyroid lymphomas are non-Hodgkin's lymphomas of B-cell origin. Thyroid lymphomas can be divided into two distinct clinicopathologic scenarios - pure MALT lymphomas and diffuse large B-cell types or mixed histological subtypes. They may also be divided into one of two groups: low- or high-grade.[1,4]

About 90% of cases are Ann Arbor stage IE or IIE at presentation.[5] Many investigators believe that only the early Ann Arbor stages (i.e., I-II) can be considered as primary thyroid in origin because no histologic marker can separate primary and metastatic thyroid lymphomas. However, in the Workshop of the European Haematopathology Association (EAHP) and the Society for Hematopathology (SH) Update on Extranodal Lymphomas 2006, arguments for a primary designation of metastatic thyroid lymphoma included a markedly enlarged thyroid, apparent thyroiditis and simultaneous presentation of the extrathyroidal disease.[6]

Open surgical biopsy is required in several cases and still plays a role despite advances in Fine-Needle Aspiration Cytology (FNAC) and its adjuncts.[5]

Immunohistochemical studies are useful in diagnosis and excluding anaplastic carcinoma, B-cell chronic lymphocytic leukemia (B-CLL), mantle-cell lymphoma and follicle-center lymphoma. MALT lymphomas express B-cell-associated surface antigens CD20, CD22 and CD79a, and are CD5-, CD10-, and CD23-negative.[2]

The prognosis of patients with thyroid lymphoma is excellent.[7] Surgery that was once the mainstay of treatment for this disease now plays a minimal role. Current treatment regimens consist of chemotherapy and external beam radiation. The National Comprehensive Cancer Network clinical practice guidelines on oncology support the use of radiation or surgery alone in early-stage (IE) MALT lymphoma. Patients with intermediate- or high-grade lymphoma arising from MALT lymphoma need aggressive treatment with combined-modality therapy, chemotherapy followed by radiotherapy. If the lymphoma has spread or comes back after initial treatment, it is often treated with chemotherapy (CHOP regimen recommended). The addition of Rituximab was shown to provide further benefit in survival and disease-free survival in thyroid lymphoma.[4,8]

In conclusion, thyroid lymphoma is a rare disease and difficult to differentiate from anaplastic carcinoma. FNAC and IHC aid in the diagnosis though open surgical biopsy is still important and relevant. The treatment is mainly radiotherapy and chemotherapy at present, the role of surgery being minimal. Prognosis is generally excellent, especially for MALTomas and lymphomas confined to the thyroid.

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