Case Report

Rare Case of Non-Hodgkin’s Lymphoma of the Thyroid

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Abstract: Thyroid lymphoma is a rare cancer constituting 1% to 2% of all thyroid cancers and less than 2% of lymphomas. Thyroid Lymphomas are usually all Non-Hodgkin’s Lymphoma and most are B cell in origin. Non-Hodgkin’s lymphoma (NHL) of the thyroid gland is a rare disease with an incidence of 0.5 per 100,000 population. We present the case of a middle aged male patient with Non-Hodgkin’s Lymphoma of thyroid with Hashimoto’s Thyroiditis with no cervical lymph nodes.

Key Words: Non-Hodgkin’s Lymphoma, Thyroid Lymphoma

Background
Primary thyroid lymphoma is a rare diagnosis,(1) being recognized more frequently in a goiter, grown significantly over a short period. It is four times more frequently seen in women and approximately half occur in a setting of pre-existing Hashimoto Thyroiditis.(1) Initial symptoms include hoarseness, pressure symptoms, fever and diffuse pain. Physical examination usually reveals firm, slightly tender, fixed mass frequently with substernal extension. Ultrasound demonstrates a classic pseudocystic pattern. FNA, core needle/open biopsy may be considered as a diagnostic modality. In highly suspicious cases additional evaluation includes use of CT/MRI for extrathyroidal disease. Approximately 50% of the disease is confined to thyroid, 5% have disease on both sides of diaphragm or diffuse organ involvement and remaining with locoregional nodal disease. Combined modality therapy is the most common approach for the initial treatment of thyroid lymphomas.(2) The CHOP regimen has been shown to be highly effective for many types of thyroid lymphomas.(3)

Case Presentation:
We report a case of a 39-year-old male, who presented to Government Wenlock Hospital, Mangalore with complaints of swelling on anterior aspect of neck since 4 months, insidious in onset and progressive in nature. It was associated with dyspnea and weight loss with no features of hype/ hyperthyroidism. Examination revealed a 14x 13 cm thyroid swelling with the right lobe enlarged more than the left lobe along with the isthmus. It was firm in consistency with smooth surface and a palpable lower border with no palpable cervical lymph nodes. Patient had normal routine investigations including thyroid function tests. Ultrasound examination was suggestive of multinodular goiter with thyroiditis and increased vascularity with no lymph node involvement. Fine needle aspiration cytology showed thyroid follicular cells arranged in monolayered sheet papillaroid fragments, macrofollicles and singles, and lymphocytic impingement on thyroid follicular cells. Background showed scant colloid, polymorphous lymphoid cells, lymphocytes, cyst macrophages, bare nuclei and RBCs. Features were suggestive of Hashimoto Thyroiditis with Hurthle cell changes BSRTC Category II.

Patient underwent total thyroidectomy and histopathology reported thyroid tissue to be replaced by lobules of nonmonomorphous atypical lymphoid cells of small-medium size with round nucleus and scant cytoplasm. Hurthle cell changes were seen. Dense lymphocytic infiltrate and lymphoid follicle with secondary germinal center formation were noted, suggestive of Non-Hodgkin’s Lymphoma with Hashimoto Thyroiditis. IHC markers came positive for CD20, CD3, CD5 and negative for CD10, BCL 6, MUM-1, CD30 and a diagnosis of Non-Hodgkin Lymphoma- Diffuse Large B-cell was confirmed.
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Fig 1: Intraoperative findings during Total Thyroidectomy

Fig 2: Intraoperative dissection during Total Thyroidectomy

Fig 3: Front view of Total Thyroidectomy specimen

Fig 4: Total Thyroidectomy gross specimen with marked poles

Fig 5: Histopathology evaluation in 40x show round small to medium atypical lymphoid cells with round vesicular nuclei, prominent nucleoli and scanty cytoplasm. Occasional foci show bizarre cells.

Fig 6: Histopathological evaluation in 10x show section with destroyed thyroid follicular architecture with diffuse lymphoid cell infiltrate arranged in lobular pattern accompanied by dense lymphocytic infiltrate. Foci shows preserved colloid with Hurthle cell changes.
PTL is a rare condition. It is responsible for less than 5% of thyroid malignancies and no more than 2.5% of all lymphomas. Most PTL are NHL with majority being 50-80% DLBCL and 20-30% cases being of MALT lymphoma, mostly extranodal marginal type.(4) PTL staging is based on Ann Arbor staging criteria.

<table>
<thead>
<tr>
<th>Ann Arbor Stage</th>
<th>Localization</th>
<th>Initial Stage</th>
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<tbody>
<tr>
<td>IE</td>
<td>Thyroid gland</td>
<td>56%</td>
</tr>
<tr>
<td>IIE</td>
<td>Thyroid gland and lymph node regions on same side of diaphragm</td>
<td>32%</td>
</tr>
<tr>
<td>IIIE</td>
<td>Thyroid gland and lymph node regions on both sides of the diaphragm or spleen</td>
<td>2%</td>
</tr>
<tr>
<td>IVE</td>
<td>Disseminated disease</td>
<td>11%</td>
</tr>
</tbody>
</table>

Secondary thyroid lymphoma originates from non thyroid neoplasm that has metastasized to thyroid.(1) Both must be differentiated as the treatment modality is different with secondary cases having higher mortality and morbidity.(5) Thyroid gland does not contain lymphoid tissue. Presence of Hashimoto’s thyroiditis autoimmune chronic lymphocytic thyroiditis has a 40-80 times risk of progressing to PTL(2) when compared to normal population with some authors claiming the association to be in 90% of PTL.(6) In this case the diagnosis of Hashimoto thyroiditis was known but histological examination proved the coexistence of NHL.

PTL is more common in female 3:1 presenting mostly in 7th decade as a rapidly growing swelling and causing pressure symptoms in the form of dyspnea, dysphagia, stridor, hoarseness and choking sensation.(3) Some patients may have B symptoms- fever, night sweat, weight loss. This is in contrast to our case presentation making it all the more rare.(6,7)

FNAC has an important role in diagnosing thyroid nodular disease, however its role in PTL is limited as it is difficult to differentiate thyroid lymphoma, Hashimoto thyroiditis and anaplastic carcinoma of thyroid, making lymphoma an underdiagnosed condition.(1) It is important to differentiate these as the treatment modality varies with DLBCL requiring chemotherapy and anaplastic carcinoma requiring surgical resection if feasible. In this case a rapidly enlarging swelling raised the suspicion of malignancy and hence the need of surgery.

There are no studies on accuracy of FNAC in diagnosing PTL but there is increased accuracy with new technical advances in the form of flow cytometry, IHC studies and molecular techniques.(8) USG guided FNAC has also increased the sensitivity. If FNA is not diagnostic, core biopsy, incisional biopsy or thyroidectomy may be required.(9) The treatment of PTL is controversial since it is a rare entity with no large scale studies on the topic. Local control can be achieved by radiotherapy or surgery or both with chemotherapy being used to control disseminated/ undetected disease.

Prognosis depends on histological classification and staging. MALT lymphoma has a better prognosis then DLBCL owing to a better response to chemoradiation. The 5-year survival rate is 90% in cases of intrathyroidal disease which drops to 35% in cases of extrathyroidal extension. Factors associated with poor prognosis include tumor size >10cm, advanced stage, obstructive local symptoms, rapid growth, mediastinal involvement, age>60 and raised LDH and beta2 microglobulin levels.(13)

References: