Original Research Paper

Volume - 10 | Issue - 9 | September - 2020 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

General Surgery

A CASE OF PRIMARY LYMPHOMA OF THYROID PRESENTING WITH RESPIRATORY DISTRESS.

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ABSTRACT Here is a case of a 50-year-old woman with hypothyroidism who presented with anterior neck mass causing compressive symptoms. The patient underwent tracheostomy with incisional biopsy of the thyroid gland and was diagnosed with Diffuse Large B Cell Lymphoma of the thyroid. The patient was later subjected to Chemotherapy.
Primary thyroid lymphomas are rare. New immunohistochemical and molecular techniques have improved the diagnostic accuracy with core-biopsy limiting the role of surgery. The treatment should first include the control of local disease with radiotherapy and/or surgery combined with chemotherapy to control obscure or disseminated disease. Palliative surgery may be needed to relieve airway compression symptoms. The prognosis of patients depends on the histological classification of the tumor and the stage of the disease.
PTL should be diagnosed at the earliest and appropriate treatment plan should be implemented to improve survival rate.

KEYWORDS :

A 50-year-old woman with hypothyroidism on medications was referred to the higher center due to compressive cervical symptoms. There was no history of cervical irradiation or family history of thyroid cancer. She presented with a gradual onset of anterior cervical mass since 2 months and rapidly progressive since 6 days with dyspnea on lying down and stridor. The indirect laryngoscopy revealed right vocal cord palsy. The Laboratory tests showed mild elevation of TSH. The x-ray revealed soft tissue mass in the cervical region causing leftward shift of trachea (fig. 1).

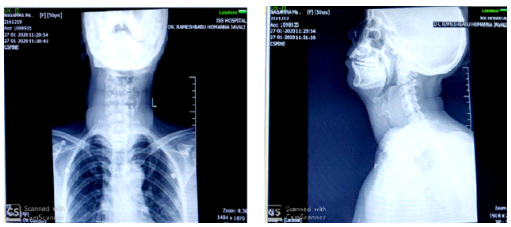

fig. 1 Fine needle aspiration cytology (FNAC) of the thyroid gland was inconclusive.
The patient underwent tracheostomy with incisional biopsy (IB) (fig. 2) of the thyroid gland in accordance with Medical oncologist opinion.

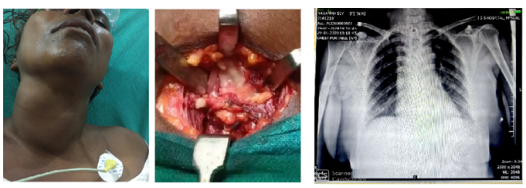

fig.2 Macroscopically, the resected thyroid gland was enlarged consisting of multiple grey and white tissue fragments and asymmetrical, while measuring 2 x 2 cm.

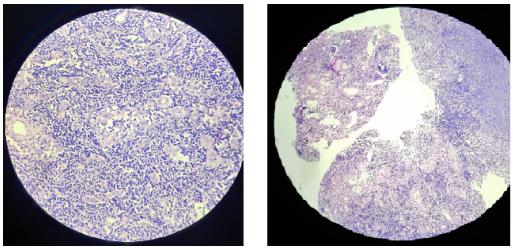

fig.3Microscopically, sections from thyroid tissue showed few residual thyroid follicles with askanazy cell change with diffuse dense infiltration by two population of lymphoid cells having coarse chromatin. Individual cells have scant cytoplasm and pleomorphic nuclei. Few residual lymphoid follicles are also seen with reactive germinal centers and the rest consisted of small reactive lymphocytes (fig. 3)

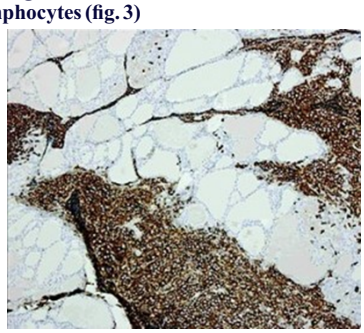

fig.4 The neoplastic cells express CD20, BCL-2, BCL-6 and MUM-1 Ki67 index is upto 70% (fig. 4).

Thyroid mass - Morphology and IHC was consistent with a diffuse large B cell lymphoma - Non Germinal center variant (Activated B cell type).

The patient was diagnosed to have Diffuse Large B-Cell Lymphoma (DLBCL) and was then subjected for 6 cycles of chemotherapy and is due to follow up.

PTL is a rare condition accounting for < 5% of all thyroid malignancies and < 3 % of all extranodal lymphomas [2] Most PTL are non-Hodgkin's lymphomas. Around 50–80% of the PTL are DLBCLs and 20–30% are mucosa-associated lymphoid tissue (MALT) lymphomas, most of these extranodal marginal type. Other rare subtypes include follicular lymphoma (12%), Hodgkin's disease (7%), small lymphocytic lymphoma (4%) and Burkitt's lymphoma (4%). While there are some reports of T cell PTL .[3]

PTL is staged based on the Ann Arbor staging criteria (Table 1[4])

Ann Arbor classification on PTL. (Table 1)

Ann Arbor stage	Localization	Initial stage
IE	Involvement of Thyroid gland	56%
IIE	Involvement of Thyroid gland + lymph node regions, on the same side of the diaphragm	32%
IIIE	Involvement of Thyroid gland + lymph node regions, on both sides of the diaphragm and/or spleen	2%
IV E	Disseminated disease	11%

The secondary thyroid lymphoma originates from a disseminated non-thyroidal neoplasia that metastasizes to the thyroid gland and is a widespread disease and the mortality rate is higher compared to PTL. There is an increased risk for certain non-Hodgkin lymphoma subtypes in association with some autoimmune disorders e.g. "Sjögren's syndrome and extra-nodal marginal zone lymphoma of the salivary gland, and thyroid MALT lymphoma in a background of Hashimoto's thyroiditis"[5]

PTL is more prevalent in female patients (F:M ratio - 3–4:1) and present more frequently in the seventh decade of life (mean age of 67 years), as a cervical mass with a quick and painless growth . 30% of patients present with symptoms and signs of compression of adjacent structures such as dyspnea, dysphagia, stridor, hoarseness, coughing or choking Most individuals are euthyroid at presentation, 10% may be hypothyroid 10–20% of patients may be diagnosed with the presence of B symptoms (fever, night sweats or weight loss) .[6]

Lymphomas of the thyroid are almost exclusively of the non-Hodgkin's, B-cell type. The most common subtype of PTL is DLBCL, more than 50% of cases, followed by MALT lymphoma, which represents about 10–23% of cases [7, 8]

DLBCL show lack of cellular cohesion, pleomorphism with many showing prominent nucleoli, numerous mitotic figures and presence of lymphoglandular bodies in the background.

Ultrasound-guided FNAC also increases the sensitivity of this technique since it is possible to prevent the biopsy of necrotic tissue regions and minimizes the risk of trauma of the adjacent structures. When FNAC is not diagnostic, a corebiopsy, IB or even a thyroidectomy may be required.

IB may be required in cases where the therapeutic strategy depends on the histological subtype, such as the large B-cell lymphoma vs MALT lymphoma as in this scenario. This method should be used only when less invasive techniques are not sufficient for diagnosis as in this clinical case.

In a study conducted by Mayo Clinic analyzing 62 patients with PTL, the combination of total thyroidectomy with adjuvant radiotherapy did not demonstrate increased survival compared to a biopsy associated to radiotherapy in stages IE or IIE. The same authors decline thyroidectomy emphasizing the possible morbidities associated and the absence of an improvement in survival rate.

Meyer-Rochow et al. suggested that patients with obstructive symptoms, radiotherapy and chemotherapy can exacerbate symptoms because of tissue edema, and therefore a palliative reducing size surgery prior to adjuvant treatment should be considered in selected patients. PTL respond rapidly to combined chemotherapy regimen, commonly the CHOP scheme with Rituximab effective for DLBCL. Combination therapy of chemotherapy and radiotherapy has more benefits over isolated locoregional therapy or systemic therapy.

MALT lymphomas have a better prognosis than DLBCL. The 5-year-survival rate in patients with intrathyroidal disease is better than patients with extrathyroidal disease. Clinical factors that predict a worse prognosis include tumor size over 10 cm, advanced stage (greater than stage IE), presence of obstructive local symptoms, rapid tumor growth, mediastinal involvement, age > 60 years and elevated LDH and β2microglobulin levels

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