

Extranodal Marginal Zone Lymphoma of Thyroid – An Unusual Tumor at Unusual Location

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Abstract

Case Report

Primary thyroid lymphoma accounts for 5% of all the malignancies of the thyroid gland. The thyroid is very rare and unusual site for lymphoma, only few cases have been described in the literature. It is associated with chronic inflammatory lesions such as chronic lymphocytic thyroiditis. We report a rare case of Marginal Zone lymphoma in a 44 year old male patient presented with complaint of swelling in the right side of neck since 3 months. The definitive diagnosis was confirmed by histopathological examination and immunohistochemistry.

Keywords: Thyroid, Lymphoma, Histopathology, Immunohistochemistry.

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INTRODUCTION

Primary thyroid lymphoma is a type of extranodal marginal B cell lymphoma, in which the thyroid gland is affected primarily because of lymphocytic infiltration seen in immunological conditions such as lymphocytic thyroiditis [1]. In general gastrointestinal tract is considered to be the most frequent site of occurrence of marginal zone lymphoma, but it is a very rare occurrence in thyroid (2-8%) [2,3]. It shows female predominance with mean age of 65 to 75 years and the M:F ratio is 1:4 [4]. Primary thyroid lymphoma is very difficult to diagnose due to their rarity and as well as their presentation of both reactive and neoplastic processes [1]. In difficult cases techniques such as histopathology, immunochemistry, and flow cytometry are utilised for the confirmation of the diagnosis of marginal zone lymphoma [1].

CASE REPORT

44 year old male presented to ENT department, Narayana General Hospital with complaint of right sided neck swelling for 3 months, which was gradually progressive. There was no history of weight loss, pain and pressure symptoms. No significant family history. General physical examination was normal. On clinical examination, right lobe of thyroid was diffusely enlarged, firm in consistency and there was no retrosternal extension. Hypo/hyperthyroidism features

were absent. Clinical examination of other systems was normal. USG showed diffusely enlarged Right lobe of thyroid without calcification. Thyroid function tests were within normal range. Other biochemical and haematological investigations were within normal range. Cytosmears examination from thyroid swelling shows follicular cells arranged in cluster forms along with lymphocytes. Background shows hemorrhages. FNAC was reported as lymphocytic thyroiditis (Fig.1).

Right hemithyroidectomy was done. Specimen was sent to the Pathology department for histopathological examination. Grossly the specimen is diffusely enlarged with no nodularity. Sections examined reveal thyroid parenchyma with sparsely arranged thyroid follicles showing architectural effacement and diffuse infiltration of polymorphous lymphoid population. Mature small lymphocytes are seen predominantly, along with few atypical lymphocytes. Impingement of lymphocytes over thyroid follicles noted in many areas. Hurthle cell change with granular eosinophilic cytoplasm noted in some follicles. Histopathological findings were consistent with features of lymphoma in thyroid (Fig.2).

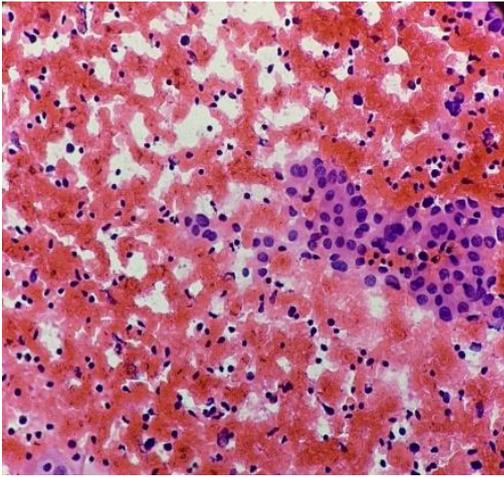


Figure 1: Cyto smears examination showing follicular cells in cluster forms along with lymphocytes (H & E; x400)

CD20 was strongly positive in all the neoplastic B-cells present in the thyroid follicles (Fig.3) and cytokeratin focally positive in thyroid follicular epithelial cells, indicating thyroid origin (Fig.4). Immunohistochemistry confirmed the diagnosis of Marginal zone lymphoma of thyroid.



Figure 2: Microscopic examination shows thyroid follicles and atypical lymphoid cells along with small lymphocytes (H & E; x 100)

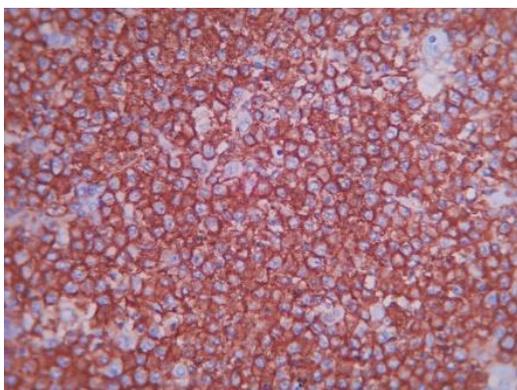


Figure 3: Neoplastic cells showing positivity for CD20 (IHC, x 400)

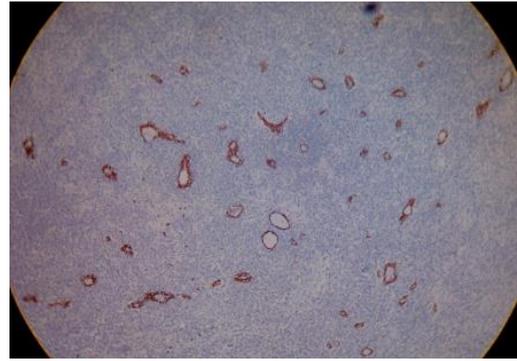


Figure 4: Thyroid follicular epithelial cells showing positivity for cytokeratin (IHC; x 100)

DISCUSSION

Among lymphomas Maltomas are slowly growing tumors which are primarily recognized at mucosal sites. Maltomas are marginal zone lymphomas belonging to B-cell tumors. They predominantly arise from the tissue having chronic inflammatory pathology due to autoimmune disorders/infectious pathology, like helicobacter gastritis associated maltoma and Hashimoto's thyroiditis associated thyroid maltoma. They lie on a spectrum between reactive hyperplasia of lymphnode and full blown B-cell lymphoma [5]. Lymphnodes, spleen and extranodal organs are the various sites for heterogenous B cell marginal zone lymphoma.

Monoclonal B-cell neoplasms will arise because of mutations and chromosomal aberrations in reactive lymphoid hyperplasia. Tissues like thyroid gland is devoid of lymphoid tissue, but in autoimmune conditions shows presence of intrathyroid lymphoid tissue, so it can develop into lymphoma [5]. In 1983, Issacon and Wright introduced the concept of MALT type lymphomas in extra nodal region [6]. In thyroid the most common histological subtype of Non Hodgkin's Lymphoma is diffuse large cell lymphoma. Lymphomas in thyroid accounts for 3% of Non-Hodgkin's lymphoma with high incidence in women and M:F ratio of 1:4.8. Most commonly seen in seventh decade [7].

B-cell antigen expressions like CD20, CD22, CD79a are observed in MALT type. CD3, CD5, CD10 are negative in MALT lymphoma. Majority of the patients are asymptomatic as the maltomas are slowly growing and remains localised for long Duration [8]. Histologically thyroid maltomas are characterized by presence of atypical lymphoid cells arising from the marginal zones of lymphoid follicles and invading into the interfollicular space and germinal centres i.e, follicular colonisation is seen in thyroid maltomas [6]. Extranodal marginal zone B-cell lymphoma was diagnosed on histopathological examination. Immunohistochemistry in our case showed CD20 positive in neoplastic B-cells colonizing thyroid follicles. Investigations like lactate dehydrogenases, β -

microglobulin also helps in diagnosis along with CT, PETCT to know the extension of disease [8].

Treatment and follow up of thyroid maltoma patients is still controversial. Disseminated diseases are treated by chemotherapy and localised lymphomas by radiotherapy. If FNAC is inconclusive or when patient presents with presence of symptoms, surgery is indicated [5]. Histopathology and immunohistochemistry are the gold standard in definitive diagnosis.

CONCLUSION

Primary thyroid maltoma is very rare in thyroid. Our case is a extranodal marginal B-cell lymphoma of thyroid. Clinical diagnosis of thyroid lymphoma can be suspected in slow growing neoplasms, when it is associated with chronic lymphocytic thyroiditis. Histopathology and immunohistochemistry is the gold standard in the diagnosis of lymphoma. Early and accurate diagnosis is very important for proper medical intervention and there by improving the prognosis of patient.

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Conflicts of interest: Nil

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