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CASE REPORT

DEFINITIVE RADIOTHERAPY IN A RARE CASE OF THYROID MUCOSA ASSOCIATED LYMPHOID TISSUE LYMPHOMA

***Dr. Rashmi S., Dr. Sai Snehit,C., Dr. Bhaskar, V., Dr. Geeta, S.N.**

Department of Radiation Oncology, Vydehi institute of Medical Science and Research Centre Bangalore, India

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ABSTRACT

Primary thyroid mucosa associated lymphoid tissue(MALT) lymphoma is a rare extranodal Non Hodgkins Lymphoma (NHL)of the thyroid. It is most common in females, has an indolent course and mostly asymptomatic making the early diagnosis more challenging. There are no optimal guidelines for the management of thyroid MALT oma. We are presenting a case of unresectablethyroid MAL Toma treated with definitive radiotherapy. Patient is under complete response since 24 months of definitive radiotherapy. We emphasise the role of radiotherapy as single modality of treatment in early unresectable thyroid MALT omas with good local control.

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INTRODUCTION

Mucosa-associated lymphoid tissue (MALT) lymphoma is defined as extra nodal lymphoma composed of heterogeneous small B cells arising from marginal zone of MALT (Terada, 2014). It most frequently occurs in the gastrointestinal tract mucosa (50%), head and neck (15%), lung(14%), skin (11%), thyroid (4%), and breast (4%) (Terada, 2014; Latheef *et al*., 2013; Peppa, 2012). Primary thyroid lymphoma (PTL) is a rare form of malignancy, constituting about 2–8% of all thyroid malignancies and 1–2% of all extra nodal lymphomas (3,4). Thyroid lymphomas are most commonly diffuse large B-cell lymphomas (60–80%), and only about 30% are extra nodal marginal zone lymphomas (Peppa *et al*., 2012). Primary thyroid MALT lymphoma is rare, accounting 6–28% ofPTLs (Derringer *et al*., 2000) and usually arises in the setting of chronic lymphocytic thyroiditis, such as Hashimoto's thyroiditis (Latheef, 2013). It has female predominancewith 4:1ratio (Derringer *et al*., 2000; Isaacson, 1983; Pedersen, 1996). The coexistence of reactive and neoplastic processes in the thyroid may cause a significant difficulty in diagnosing MALToma using cytology or histology. This has led to the use of immunohistochemistry, flow cytometry, and molecular techniques (Southern blotting, PCR) to confirm or exclude the diagnosis.

Corresponding author:* **Dr. Rashmi S.,
Department of Radiation Oncology, Vydehi institute of Medical Science and Research Centre Bangalore, India.

Case description: A 50 year old female presented to OPD with complaints of history of neck swelling on the right side with six month duration. Swelling was gradually progressive with no associated pain, weight loss or pressure symptoms. There was no significant family history. Her general physical examination appeared normal without any obvious deformities or abnormalities. Local examination revealed diffuse smooth swelling of4x4 cm size of the right lobe ofthe thyroid which was firm in consistency. No features of hypo or hyperthyroidism were present. All her other systems seemed to be normal clinically.

High resolution ultrasound: Revealed enlarged right lobe of thyroid with altered echo texture measuring 45x37x33mm and multiple calcific foci.

Fine-needle aspiration: Cytology showed cytologic features are consistent with lymphoproliferative disorder suggestive of Non hodgkins lymphoma. Thyroid function tests were normal. All the haematological and biochemical investigations were within the normal range.

PET CT: Revealed 30x50x66mm FDG avid lesion in para tracheal region and extension into tracheoesophageal groove on right side with encasement of brachiocephalic trunk and right subclavian artery.

Histopathological examination: Of the specimen was suggestive ofextranodal marginal zone B-cell lymphoma.

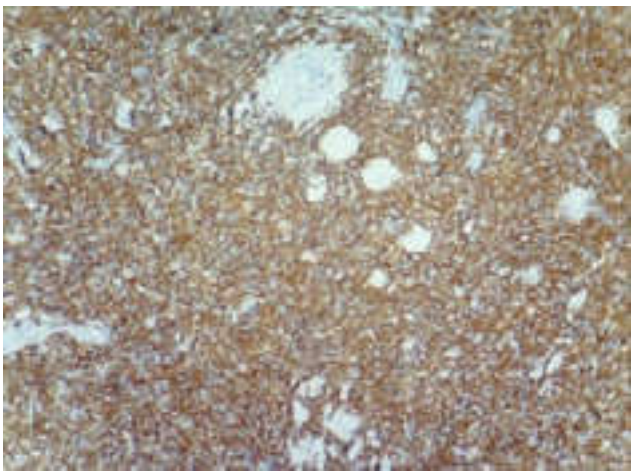

Fig. 1. IHC showing CD 45 positivity with strong CD 20 positive cells

Immunohistochemistry: Showed CD45 positivity in all atypical lymphocytes. CD20 was strongly positive in all the neoplastic B-cells colonizing the thyroid follicles and admixed with sheets of plasmacytoid cells as clusters. Bcl-2 and Bcl-6 was negative. Patient was staged as stage IAE (Ann Arbor staging) considering her disease was limited to the thyroid gland without marrow infiltration, evidence of metastatic disease, or B symptoms. Surgery was not feasible due to tumor encasing major blood vessels. The patient was planned for Definitive Radiotherapy (23.6Gy in 13 fractions) and patient has been symptom free since 24 months.

DISCUSSION

Primary thyroid MALT lymphoma follows an indolent course and presents without any overt symptoms. Although most lymphomas present with typical 'B' symptoms such as fever, nocturnal sweating and weight loss, this is usually not seen in this disease process, which make the diagnosis even more challenging (Latheef, 2013; Peppa, 2012). Approximately 10% of patients may have these symptoms and 10% may have features of hypothyroidism, usually associated with Hashimoto's thyroiditis (Isaacson, 1983). MALTomas were usually found to arise sat the sites where ongoing chronic inflammation with underlying autoimmune or infectious etiology was observed (Latheef, 2013). A similar pathophysiology is seen with chronic inflammation of thyroid gland in the setting of ongoing Hashimoto's thyroiditis, which was not seen in our patient. Nevertheless, association of H. pylori and thyroid MALT lymphoma has not been adequately addressed in the literature (Peppa, 2012). Histopathology showsatypical lymphoid cells originating within marginal zone of lymphoid follicles and extending into inter-follicular spaces and germinal centres (follicular colonization) (Latheef, 2013; Peppa, 2012; Isaacson, 1983). In our patient, FNA showed neoplastic proliferation of mostly singly dispersed small- to medium-sized cells with lymphoid cell proliferation, composed predominantly of small mature lymphocytes.

Review of 103 cases done by Tsang *et al*. with localized (stage IE/IIE) extra nodal MALT lymphoma had overall 5 years survival of 98% in 85 patients with extra nodal MALT lymphoma treated with radiation therapy alone (11). The optimal treatment regimen and follow-up for these patients remains controversial. The stage and histologic grade do play a major role in determining prognosis of the disease process. Diffuse large B-cell lymphoma has a poorer prognosis compared to localized primary MALTomas. Stages I/II respond well to localized treatment as compared to stages III/IV which are more disseminated and need combine modalities of chemo and radiation therapy (Isaacson, 1983; Mack, 2007). Although this disease has excellentsurvival rate after treatment, the optimal follow-up remains controversial at present (Derringer, 2000; Mack, 2007).

Conclusion

We emphasise the role of radiotherapy as single modality of treatment in early unresectable thyroid MALTomas with the benefit of good local control. Combined chemoradiation therapy is recommended for all diffuse B-cell or mixed lymphomas.

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