

Burkitt's Lymphoma of the Thyroid: A Case Report

Dr. Antony Prestine K.P¹, Dr. Sugeeth M T^{2*}, Dr. Priya Mary Jacob³, Dr. Manoj S¹

¹MD, Senior Resident, Department of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India

²MD, DNB, DM, DNB, Assistant Professor, Department of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India

³MD, Associate Professor, Department of Pathology, Regional Cancer Centre, Trivandrum 695011, India

DOI: [10.36347/sjmcr.2023.v11i01.017](https://doi.org/10.36347/sjmcr.2023.v11i01.017)

| Received: 12.12.2022 | Accepted: 24.01.2023 | Published: 30.01.2023

*Corresponding author: Dr. Sugeeth M T

MD, DNB, DM, DNB, Assistant Professor, Department of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India

Abstract

Case Report

Burkitt's Lymphoma arises from mature B cells and is a highly aggressive neoplasm but potentially responds to intensive combination chemotherapy. Primary thyroid lymphomas are rare and Burkitt lymphoma of thyroid is still rarer. We present the case of middle-aged lady with rapidly enlarging neck mass diagnosed as Burkitt's Lymphoma post hemithyroidectomy. She received systemic combination chemotherapy and is in remission at a follow up of 1 year.

Keywords: Neck Mass, Primary Thyroid Lymphoma, Burkitt's Lymphoma.

Copyright © 2023 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

Lymphomas of the thyroid are rare, accounts for 5% of thyroid neoplasms, 2.5% of lymphomas in general and up to 7% of extranodal lymphomas [1]. Primary thyroid lymphomas (PTL) typically occur in middle- to older-aged women with a history of autoimmune thyroiditis [2]. The most common pathological type is diffuse large B-cell lymphoma (DLBCL), accounting for 57.4% of all PTL. Second most common pathological type is MALT lymphoma which is an indolent lymphoma [3]. Primary Burkitt's Lymphoma (BL) of the thyroid is very rare and there are only very few cases that have been reported till date. Here we report a case of primary BL of thyroid in a 42-year old lady, its clinical features and management along with brief review of the literature

CASE REPORT

A forty-two-year-old lady with history of hypothyroidism on thyroxine supplementation since last 6 years was evaluated for rapidly increasing neck swelling of 2 months duration. She had difficulty in swallowing. There was no history of difficulty in breathing or hoarseness of voice. Patient had no swelling anywhere else in the body and had no B symptoms. Ultrasonogram of neck showed features of resolving lymphocytic thyroiditis with multiple

hypoechoic solid appearing nodules of 9 to 20 mm in left lobe (TIRADS 4), with no significant cervical lymphadenopathy (Figure 1). Fine needle aspiration cytology (FNAC) from the thyroid nodule was suggestive of lymphocytic thyroiditis and she presented to us post left hemithyroidectomy. Histopathological examination of the specimen showed neoplasm with tumor cells showing scanty cytoplasm, round nuclei with clumped chromatin in diffuse sheets. On immunohistochemistry these cells were positive for CD10, CD20, BCL6, c-Myc and negative for CD5, BCL2, CD34, Tdt with a high labelling index of 100% (Figure 2). The picture was compatible with BL and Fluorescent insitu hybridization for c-MYC rearrangement was positive. At presentation she had a haemoglobin of 11.8g/dl, total count 6600/mm³, platelet count 3.2lakh/mm³, Serum lactate dehydrogenase was 251U/L. Computed Tomography (CT) scan of neck, chest, abdomen and pelvis done as part of lymphoma workup revealed only postoperative changes in left lower cervical region and no significant lymphadenopathy or organ involvement elsewhere. Bone marrow study was within normal limit. She was diagnosed to have low risk BL (Stage I, non-bulky with normal LDH) of the thyroid gland was planned for R-CODOX-M x 3 cycles. She completed R-CODOX-M x 3 cycles and attained complete remission and is on regular follow up since last one year.

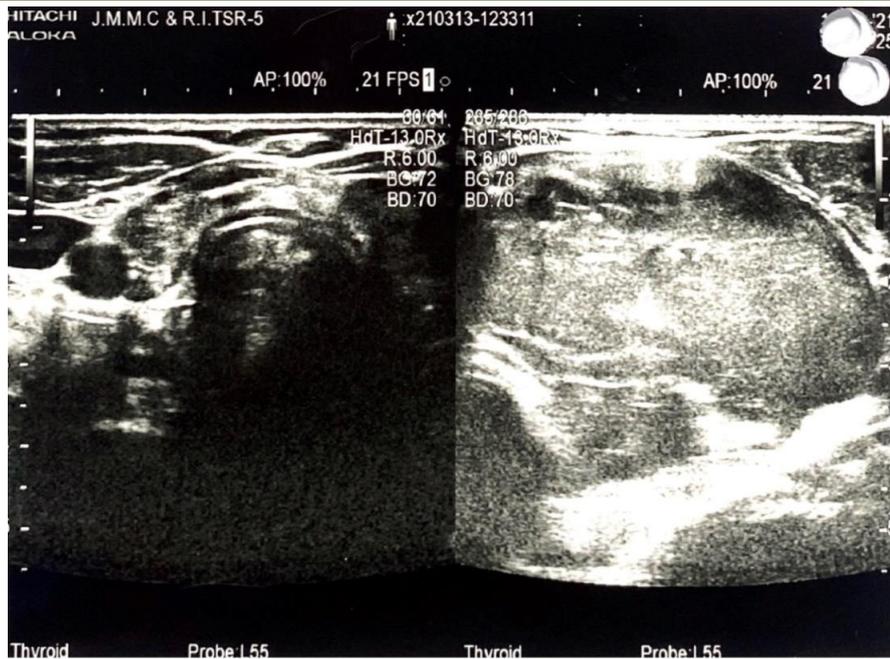


Figure 1: Ultrasonogram of neck showed features of resolving lymphocytic thyroiditis with multiple hypoechoic solid appearing nodules of 9 to 20 mm in left lobe (TIRADS 4)

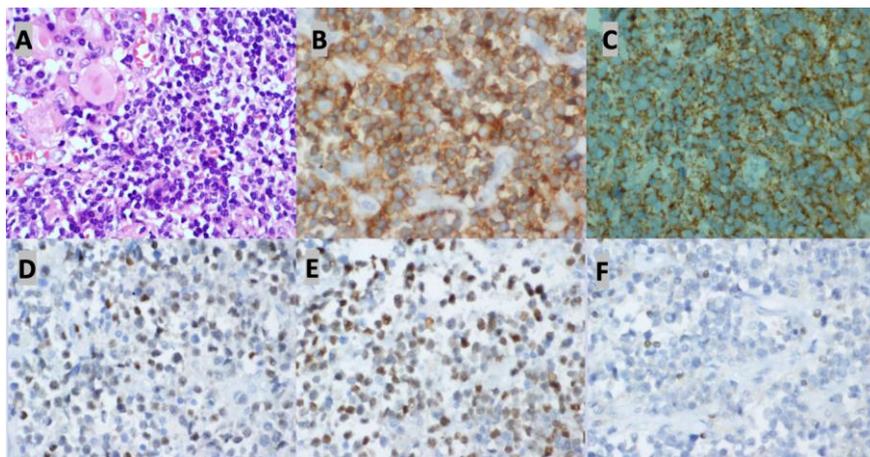


Figure 2 A: Histopathological examination of the specimen showed neoplasm with tumor cells showing scanty cytoplasm, round nuclei with clumped chromatin in diffuse sheets. On immunohistochemistry these cells were positive for CD 10 (B), CD20 (C), c-Myc (D), BCL6 (E) and negative for BCL2 (F)

DISCUSSION

BL is one of the highly aggressive B-cell non-Hodgkin’s lymphomas with three distinct forms: endemic (African), sporadic and immunodeficiency-associated [4]. First described by Denis Burkitt, an Irish surgeon [5]. Burkitt’s lymphoma (BL) accounts for only 2.5% cases of PTL [3].

The Hallmark “starry sky” appearance is due to scattered tingible-body-laden macrophages that contain apoptotic tumour cells [6]. BL arises from mature B cells derived from a germinal centre. Immunohistochemistry on tissue biopsy or flow cytometry from fine needle aspiration specimens will show positivity for CD20, CD10, BCL6, CD79a, CD45 and BCL2 will be negative [7]. Cytogenetic studies will show characteristic translocation t(8;14) (q24;q32) in

70–80% of patients while in another 10–15% of patients will show variant translocations, t(2;8)(p12;q24) and t(8;22) (q24;q11). These translocations will eventually lead to deregulated expression of the MYC oncogene, which plays a major role in cell cycle control [8].

After getting high response rates in an initial favourable publication from the National Cancer Institute (NCI) Cyclophosphamide, doxorubicin, vincristine, methotrexate, ifosfamide, etoposide, and high-dose cytarabine, with intrathecal cytarabine and methotrexate (CODOX-M/IVAC) is among the most commonly used regimens for adults with BL [9]. Rituximab is routinely incorporated in treatment regimens as per encouraging results in several studies [10, 11]. Other commonly used regimens are Hyper CVAD and dose-adjusted R-EPOCH. Aggressive high-dose therapy is indicated in adult BL.

Quesada *et al.*, reviewed 7 patients with Burkitt lymphoma of the thyroid gland [12]. It included 4 men and 3 women with a median age of 41 years who presented with a rapidly growing neck mass. Upper airway compression was seen in 5 patients. Six patients received R-Hyper-CVAD while one patient was treated with R- EPOCH. At the end of the study period, 5 patients were alive, 4 in complete remission, and 1 with persistent disease. Yildiz *et al.*, and Mweempwa A. *et al.*, also reported BL of the thyroid gland [13, 14].

Our patient is a middle-aged lady belonging to typical age group and sex for primary thyroid lymphomas. She also gives history of taking thyroxine supplementation for hypothyroidism and USG of neck, FNAC is showing features of lymphocytic thyroiditis. It was seen in multiple studies that patients with chronic lymphocytic thyroiditis are at increased risk of subsequently developing thyroid lymphoma. Our case reminds that differential diagnosis of lymphoma should be kept in mind when dealing with a thyroid mass, as the management is different.

CONCLUSION

Primary thyroid lymphomas are rare neoplasms and among them Burkitt's lymphoma of thyroid is exceedingly rare. Diagnosis may be delayed due to unusual presentation. Even though it is a highly aggressive tumour if diagnosed early can be potentially cured with combination chemotherapy regimens.

REFERENCES

1. KOSSEV, P., & LIVOLSI, V. (1999). Lymphoid lesions of the thyroid: review in light of the revised European-American lymphoma classification and upcoming World Health Organization classification. *Thyroid*, 9(12), 1273-1280.
2. Derringer, G. A., Thompson, L. D., Frommelt, R. A., Bijwaard, K. E., Heffess, C. S., & Abbondanzo, S. L. (2000). Malignant lymphoma of the thyroid gland: a clinicopathologic study of 108 cases. *The American journal of surgical pathology*, 24(5), 623-639.
3. Xiang, N., Dong, F., Zhan, X., Wang, S., Wang, J., Sun, E., & Chen, B. (2021). Incidence and prognostic factors of primary thyroid lymphoma and construction of prognostic models for post-chemotherapy and postoperative patients: a population-based study. *BMC Endocrine Disorders*, 21(1), 1-15. doi: 10.1186/s12902
4. Campo, E., Swerdlow, S. H., Harris, N. L., Pileri, S., Stein, H., & Jaffe, E. S. (2011). The 2008 WHO classification of lymphoid neoplasms and beyond: evolving concepts and practical applications. *Blood, The Journal of the American Society of Hematology*, 117(19), 5019-5032. doi:10.1182/blood-2011-01-293050
5. Burkitt, D. (1958). A sarcoma involving the jaws in African children. *Br J Surg.*, 46(197), 218-23. doi: 10.1002/bjs.18004619704. PMID: 13628987
6. Fujita, S., Buziba, N., Kumatori, A., Senba, M., Yamaguchi, A., & Toriyama, K. (2004). Early stage of Epstein-Barr virus lytic infection leading to the "starry sky" pattern formation in endemic Burkitt lymphoma. *Arch Pathol Lab Med*, 128, 549-52.
7. Dave, S. S., Fu, K., Wright, G. W., Lam, L. T., Kluin, P., Boerma, E. J., ... & Staudt, L. M. (2006). Molecular diagnosis of Burkitt's lymphoma. *New England Journal of Medicine*, 354(23), 2431-2442.
8. Kelemen, K., Braziel, R. M., Gatter, K., Bakke, T. C., Olson, S., & Fan, G. (2010). Immunophenotypic variations of Burkitt lymphoma. *American journal of clinical pathology*, 134(1), 127-138.
9. Magrath, I., Adde, M., Shad, A., Venzon, D., Seibel, N., Gootenberg, J., ... & Horak, I. D. (1996). Adults and children with small non-cleaved-cell lymphoma have a similar excellent outcome when treated with the same chemotherapy regimen. *Journal of Clinical Oncology*, 14(3), 925-934.
10. Barnes, J. A., Lacasce, A. S., Feng, Y., Toomey, C. E., Neuberg, D., Michaelson, J. S., ... & Abramson, J. S. (2011). Evaluation of the addition of rituximab to CODOX-M/IVAC for Burkitt's lymphoma: a retrospective analysis. *Annals of Oncology*, 22(8), 1859-1864.
11. Ribrag, V., Koscielny, S., Bouabdallah, K., Salles, G., Casasnovas, O., Recher, C., ... & Coiffier, B. (2012). Addition of rituximab improves outcome of HIV negative patients with Burkitt lymphoma treated with the Lmba protocol: results of the Randomized Intergroup (GRAALL-Lysa) LMBA02 protocol.(IGR sponsored LMBA02, NCT00180882). *Blood*, 120(21), 685.
12. Quesada, A. E., Liu, H., Miranda, R. N., Golardi, N., Billah, S., Medeiros, L. J., & Jaso, J. M. (2016). Burkitt lymphoma presenting as a mass in the thyroid gland: a clinicopathologic study of 7 cases and review of the literature. *Human Pathology*, 56, 101-108. doi:10.1016/j.humpath.2016.05.018
13. Yildiz, I., Sen, F., Toz, B., Kilic, L., Agan, M., & Basaran, M. (2012). Primary Burkitt's Lymphoma Presenting as a Rapidly Growing Thyroid Mass. *Case Reports in Oncology*, 5(2), 388-393. doi:10.1159/000341260
14. Mweempwa, A., Prasad, J., & Islam, S. (2013). A rare neoplasm of the thyroid gland. *N Z Med J.*, 126(1369), 75-8. PMID: 23463113.