Primary Mantle Cell Lymphoma of Thyroid- A Rare Case Report

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ABSTRACT

Introduction

Primary lymphoma of the thyroid is a rare malignancy. DLBCL is also the most common subtype followed by mucosa-associated lymphoid tissue (MALT) lymphoma.

Case Report

Here we present a case of a 62-year-old female presented to out-patient-department with a complaint of swelling in front of her neck for four years. The swelling was diffuse, hard in consistency, approximately 9x8 cm. On USG both the lobes were bulky heterogenous with a bulky isthmus. Total thyroidectomy was performed and sections taken showed diffuse lymphomatous infiltration of the gland by atypical lymphoid cells. On applying IHC CD20, CD5, Bcl2, Cyclin D1 were diffusely positive while CD 10 was negative and ki67 showed high proliferation index. So, the final diagnosis on the basis of clinical, radiological, histomorphological, and immunohistochemical grounds was made as Mantle Cell lymphoma of the thyroid gland.

Conclusion

Only four cases of MCL affecting the thyroid have been reported till now to the best of our knowledge. Hence, this is a rare case of primary mantle cell lymphoma of thyroid.

Keywords

Mantle Cell Lymphoma; thyroid malignancy;DLBCL thyroid

INTRODUCTION

Extranodal NHL constitutes to only 25% of all cases. Some rare sites include thyroid and muscles.[1] Among thyroid malignancies primary lymphoma is very rare contributing to <5% and accounting for <2% of extranodal lymphomas.[2] It affects women more than men in their sixth to seventh decade.[3] Lymphocytic thyroiditis is the most common risk factor in thyroid lymphoma with an estimated risk increment of nearly 40 to 80 times with an average interval of 20 to 30 years in developing lymphoma after the diagnosis of thyroiditis.[4] Most primary thyroid lymphomas are B cell NHL. Among B cells, DLBCL is also the most common subtype, accounting for 60-70% of cases, followed by mucosa-associated lymphoid tissue (MALT) lymphoma. T cell lymphoma and other subtypes of B cell lymphoma are among rare entities.[5]

Case Report

A 62-year-old female presented to out-patient-department with a complaint of swelling in front of her neck for four years, which was gradually progressive and not associated with pain. There was a history of on-and-off low-grade fever for six months, along with a history of weight loss. The patient had no history of swelling at any other site, no similar complaints in the past, or any other family member. She was not on any medication. She gave a history of difficulty in breathing, which was a new symptom she started experiencing for 25 days.

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On examination, the patient's vitals were stable. The swelling was diffuse, hard in consistency, approximately 9x8 cm, and was moving with deglutition. No ulceration was noted on the overlying skin. She underwent ultrasonography of the neck, which showed diffuse enlargement of both lobes, with the left lobe being larger than the right. Both the lobes were bulky heterogenous with a bulky isthmus. Multiple tiny hypoechoic areas were noted within lobes, giving a spongiform appearance. These findings pointed towards neoplastic etiology. Computed tomography was performed, and similar findings were obtained without the involvement of surrounding structures and lymph nodes. Fine needle aspiration cytology was performed, which showed cellular smears containing small to intermediate-sized monotonous lymphoid cells with indistinct nucleoli and stippled chromatin. Her counts and general blood picture were within normal limits. Total thyroidectomy was performed, and the gross specimen measured 10x6.5x5cm. The left lobe was larger than the right, which collaborated with USG and CT findings. On the cut section of both lobes, white homogenous areas were observed. The outer surface of both lobes appeared bosselated (Figure 1). Sections taken showed diffuse lymphomatous infiltration of the gland by atypical lymphoid cells (Figure 2). Although the gland was diffusely effaced, few surviving follicular acini were noted in the subcapsular area. Lymphoid infiltration was seen involving all the margins. Keeping in mind the diagnosis of NHL in the setting of preexisting Hashimoto's thyroiditis, immunohistochemistry was applied. CD20 showed strong diffuse membranous positivity, establishing monoclonality and pointed toward B NHL. Since DLBCL is the most common subtype, CD5 and CD10 were applied. CD5 showed strong diffuse positivity, and CD10 turned out to be negative in tumor cells. For further typing, we applied CD23, which highlighted FDC meshwork, Bcl2 showed strong diffuse positivity, and ki67 was 65-70%. Finally, we applied Cyclin D1, which, to our surprise, showed strong nuclear positivity (Figure 3). So, the final diagnosis on the basis of clinical, radiological, histomorphological, and immunohistochemical grounds was made as Mantle Cell lymphoma of the thyroid gland.

DISCUSSION

Primary thyroid lymphoma is a rare malignancy and is defined as lymphoma arising from the thyroid gland without contiguous involvement of other areas and distant metastasis when diagnosis.[6] An expanding neck mass including cervical nodes is the most typical clinical manifestation. Compressive symptoms such stridor, dyspnea, and hoarseness of voice are present in over 40% of cases.[2] Difficulty in breathing was also present in our patient. Nearly 10% of patients present with B-symptoms such as fever, night sweats, and weight loss.[2] The primary diagnostic tool is ultrasonography, which is capable of categorizing lymphoma into diffuse, nodular, and mixed types.[7] In our case, diffuse swelling involving both lobes was noted on ultrasonography. FNAC has an important diagnostic value, but the difficulty arises in differentiation from Hashimoto's thyroiditis and poorly differentiated, undifferentiated, or anaplastic thyroid carcinomas. Smear containing atypical lymphoma cells and few non-neoplastic thyroiditis elements pose a diagnostic challenge. FNAC, in collaboration with the cell block, serves as a crucial tool for conclusive diagnosis.[8] In cases of enlarged cervical lymph node FNAC followed by biopsy should be done to see lymph node architecture and metastasis.[9] In our has patient started developing compressive symptoms, so a total thyroidectomy was performed, which led to the final diagnosis. H & E stained sections in our case showed diffuse effacement of the thyroid gland by atypical lymphoid cells. Since DLBCL is the most common subtype, followed by MALT lymphoma, we also kept these two as the first differentials.[5] But to our surprise, immunohistochemistry results pointed towards mantle cell lymphoma. MCL has a proliferation of mature B cells that co-express CD5. The lymphoma cells have t(11;14)(q13;q32), which leads to Cyclin D1 being overexpressed. However, a subset of MCL may be negative for Cyclin D1.[10] In our case, expression of CD5 and Cyclin D1 was diffusely positive (Figure 3). MCL is a rare subtype of B cell NHL, and its presentation is thyroid, as PTL is even rare. Only four cases of MCL affecting the thyroid have been reported. Hence, it can be inferred that MCL in the thyroid is rare or underdiagnosed.[11]

There is no defined standard care for MCL in older patients, but chemoimmunotherapy followed by rituximab is the most commonly used regimen. Even after response to therapy, patients usually end up in relapse.[12] Since MCL is an aggressive lymphoma, prognosis is difficult to assess. However, the International Prognostic Index (IPI) takes into account patient age, stage of disease, serum lactate dehydrogenase, performance status, and

presence of extranodal sites to look for prognosis. Median survival in MCL is estimated to be 3-5 years.[13]

Since our patient is a poor elderly female, treatment options are being explored to give her the best therapy possible. However, after surgery, her compressive symptoms are relieved, and she has developed no new symptoms.

CONCLUSIONS

This is a rare case of lymphoma occurring primarily in thyroid. Such entities are difficult to suspect and diagnose as mostly it occurs in setting of thyroiditis as was true in our case also. Diffuse effacement of architecture provides a hint. However, immunohistochemical examination is relevant for exact diagnosis and typing. This case provide insight that thyroid lesion in elderly female could be a lymphoma so it should be kept in differential diagnosis if the morphology suggests.

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Authors's contribution

Data gathering and idea owner of this study: Dr. Mohd. Rafey and Dr. Syeda Iqra Usman

Study design: Dr. Nishat Afroz

Data gathering: Dr. Syeda Iqra Usman and Dr. Mohd. Rafey

Writing and submitting manuscript: Dr. Syeda Iqra Usman

Editing and approval of final draft: Dr. Mohd. Rafey and Dr. Murad Ahmad

Legends to figures

Fig 1- Gross: Bulky right and left lobe of thyroid with cut section showing white homogenous areas.

Fig 2- H and E: Diffuse infiltration of thyroid by lymphoid cells.

Fig 3- Immunohistochemistry of Mantle cell lymphoma

Abbreviations- DLBCL- Diffuse Large B cell lymphoma

MCL- Mantle cell lymphoma

IHC- Immunohistochemistry

H and E- Hematoxylin and Eosin

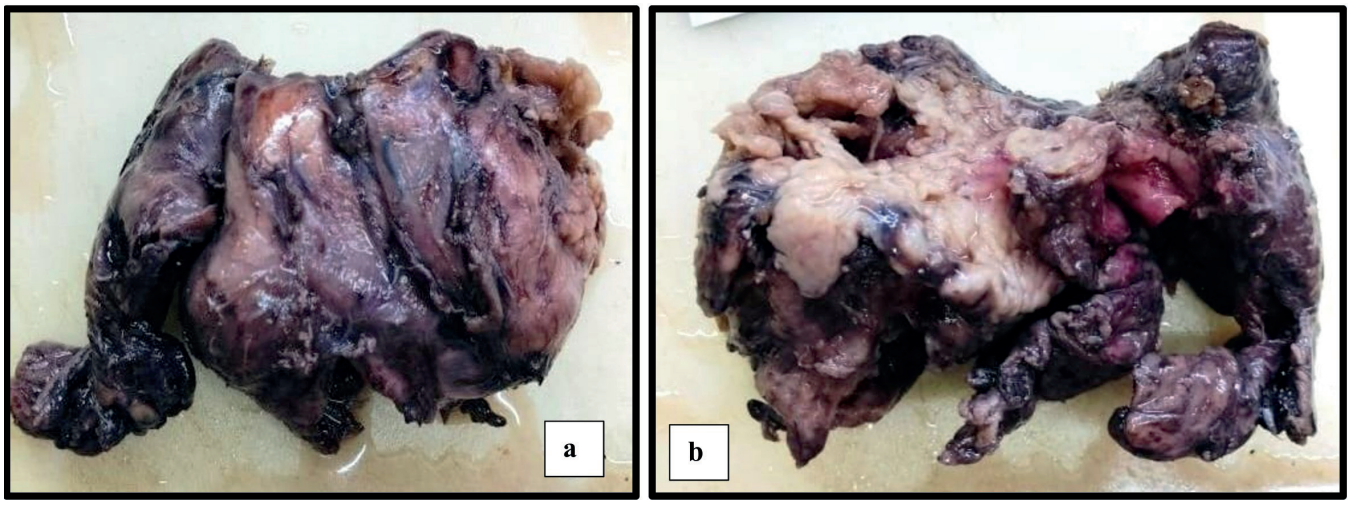


Fig 1- Gross: Bulky right and left lobe of thyroid with cut section showing white homogenous areas.

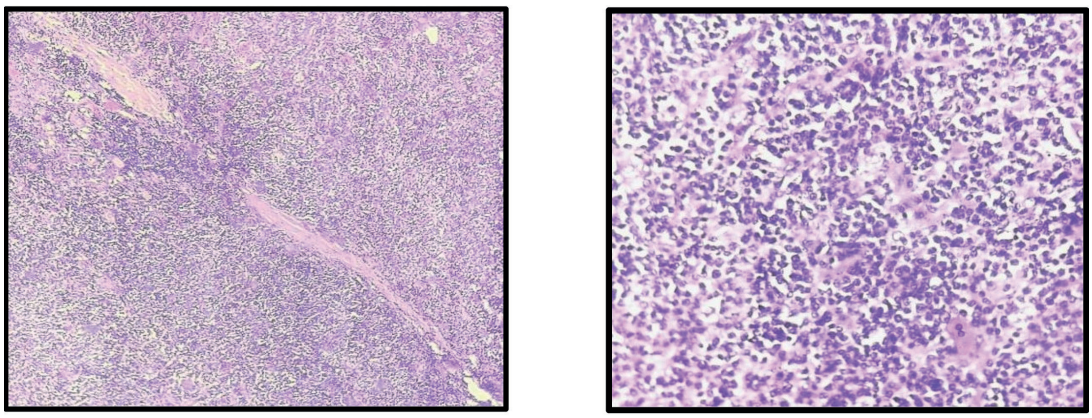
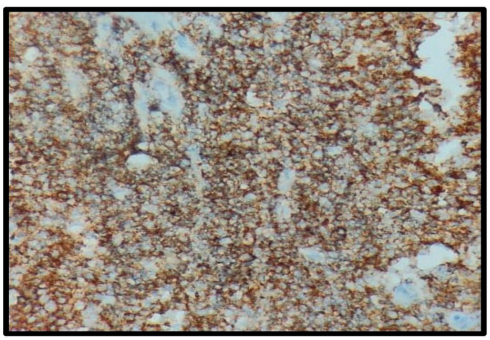
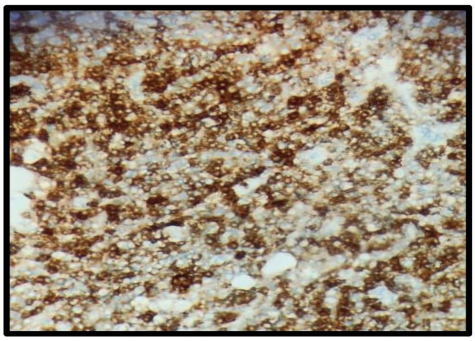


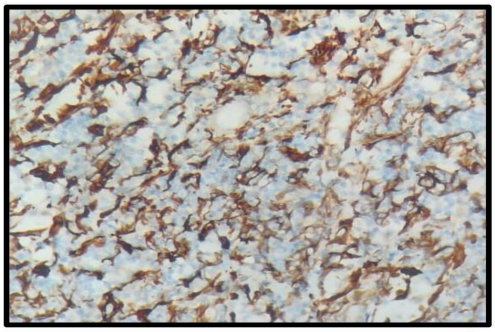
Fig 2- H and E: Diffuse infiltration of thyroid by lymphoid cells.



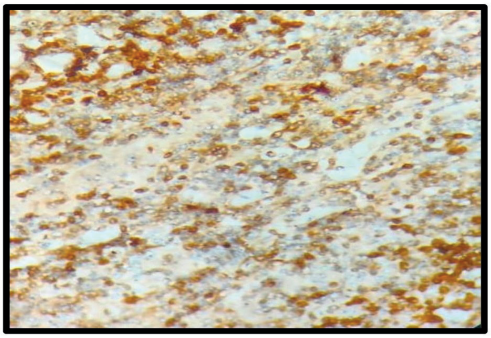
CD 20: Diffuse Positivity



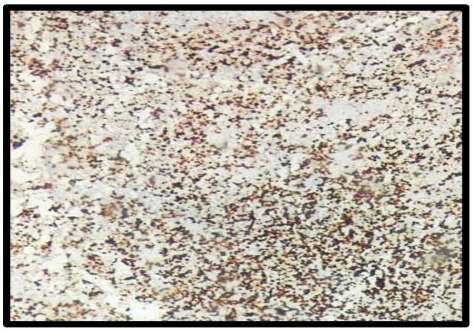
CD 5: Diffuse Positivity



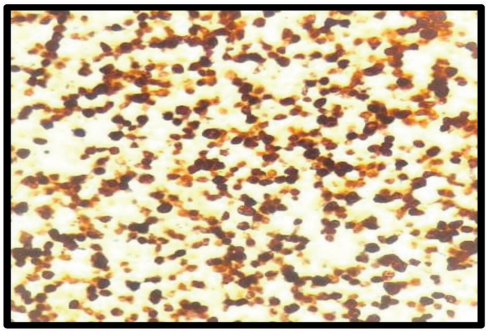
CD 23: Highlighting FDC Meshwork



Bcl 2: Strong Diffuse Positivity



Ki 67: 60-70% Positivity



Cyclin D1: Strong Diffuse Nuclear Positivity

Fig 3- Immunohistochemistry of Mantle cell lymphoma