



LYMPHOMA PRESENTING AS MULTINODULAR GOITRE

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1. Introduction

Head and neck are the second most common site for extra-nodal lymphoma. It can involve virtually any region, including the orbit, paranasal sinuses, Waldeyer's ring, salivary glands, or thyroid but the commonest is the cervical lymph nodes. The diagnosis of lymphoma is based on the histopathology examination of tissue biopsy taken from the enlarged lesion or lymph nodes. The purpose of this report is to make the otolaryngologist aware of this entity as a cause of an anterior neck mass.

Lymphomas are cancers of lymphatic system which composed of lymph nodes in the neck, armpits, groin, chest, and abdomen. It divided into two categories: Hodgkin lymphoma and non-Hodgkin lymphoma. Non-Hodgkin lymphomas (NHL) represent a group of neoplasm that commonly involve extra-nodal structures. NHL of the head and neck arises from lymph nodes in 65% of cases and, 25% to 30% in extra-nodal cases².

Twenty four percent of all non-Hodgkin lymphomas originate in extra-nodal sites, with thirty four percent of these involves head and neck structures.⁷. Primary malignant lymphoma of the thyroid gland is uncommon, and represents less than 5% of all thyroid malignancies¹. NHL of the thyroid gland generally presents in the sixth or seventh decades of life^{4,5}.

Keywords : neck tumor,lymphoma,thyroid mass,chronic lymphocytic leukemia

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2. Case Report

A 55-year-old gentleman presented to our otorhinolaryngology clinic with a one-year history of painless swelling in the anterior region of the neck. The swelling had been gradually increasing in size but not associated with dysphagia, hoarseness or dyspnea. There were no constitutional symptoms present in the patient and no symptoms of hypothyroidism or hyperthyroidism. In addition, patient denied any history of trauma or insect bite related to the neck.

Upon examination patient was comfortable and no stridor. Physical examination revealed a firm, non-tender, fixed anterior neck mass along the midline measuring about 8x8 cm. The surface of the mass is smooth, with no overlying skin changes and does not move with deglutition.

Routine hematological biochemical investigations were normal. Thyroid function tests were also normal. Ultrasound neck shows multinodular goiter bilaterally and subsequently fine needle aspiration cytology which revealed features suggestive of lymphoma.

We then proceeded with computer tomography of brain, neck, abdomen and pelvis with contrast which then revealed anterior neck mass along the midline level four measuring 5x10x7, thyroid homogeneously enhanced with no calcification within. Subsequently an incisional biopsy was done.

The biopsy showed fragments of lymphoid tissue harboring small to medium size lymphoid cells arranged in diffuse pattern. The darker areas consisted of small neoplastic lymphoid cells exhibit round nuclei with clumped chromatin and small nucleoli. The clusters of paler areas were composed of

medium cells have round nuclei with dispersed chromatin, centrally located eosinophilic nucleoli and slightly basophilic cytoplasm. Foci of sclerotic bands were present. There are scattered reactive mature T-cells are observed in the background. The neoplastic lymphoid cells demonstrated strong positive immunostaining for B-cell markers, CD 20 and CD 79a. The CD3, CD 23, PAX5 were also strongly positive. Mitoses were minimal. The Ki-67 proliferative index were generally low, but show high proliferation rate of the cells within the proliferation centers.

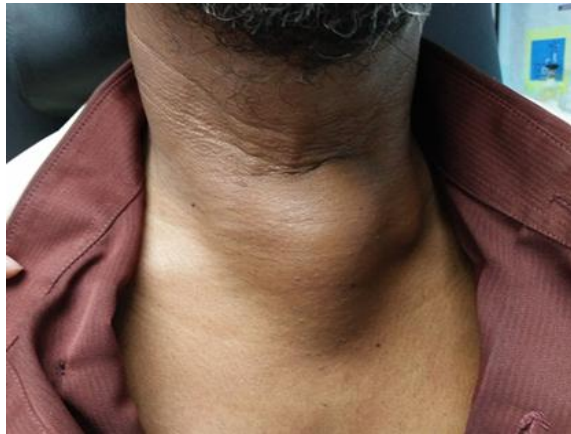


Figure 1- 55 years old gentleman with an anterior neck swelling.



Figure 2. Anterior neck mass along the midline, homogeneously enhanced in post contrast with no signs of calcification.



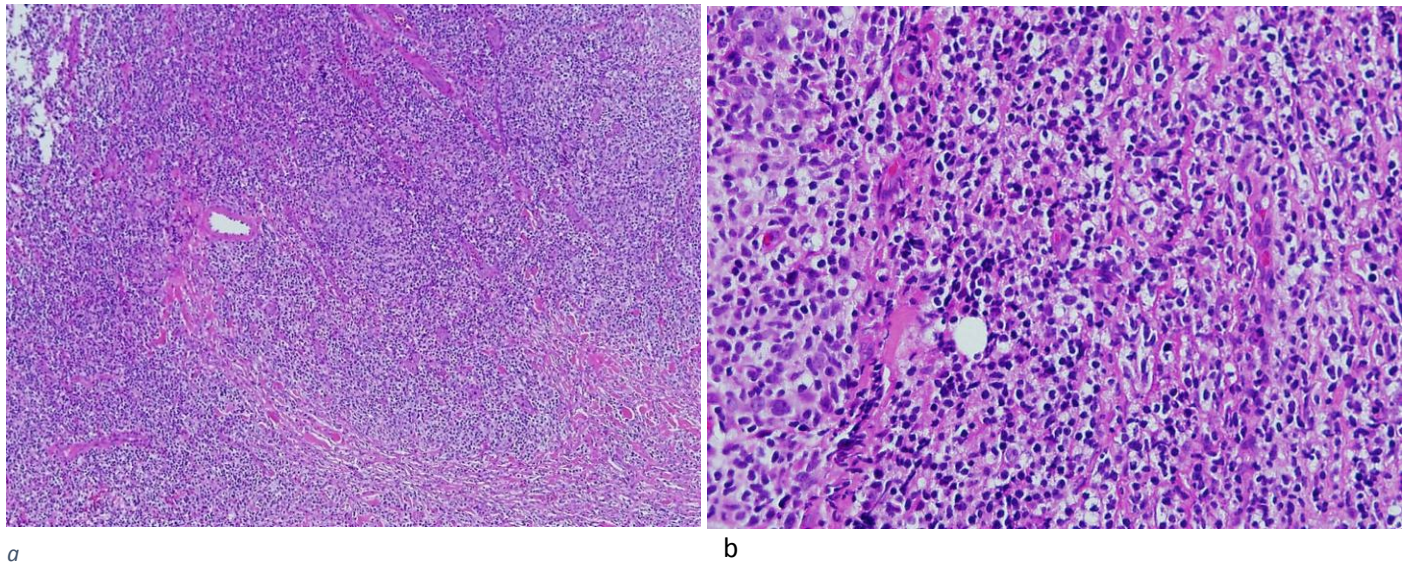
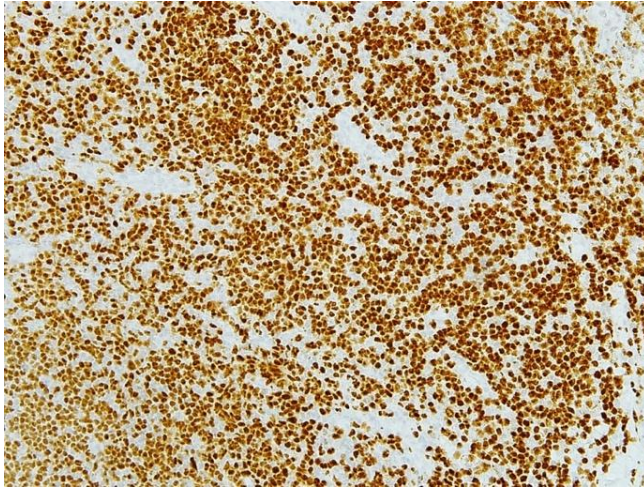


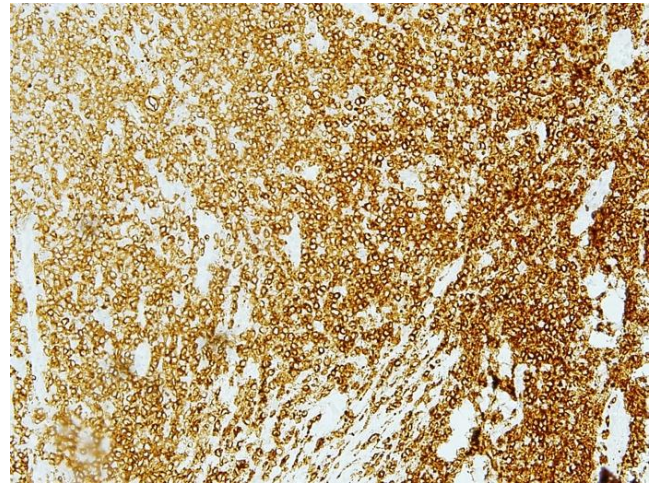
Figure 3. Microscopic findings (hematoxylin-eosin stain)

(a) Low magnification exhibits diffuse proliferation of neoplastic lymphoid cell (dark and paler areas) (x40).

(b) Higher magnification of monotonous neoplastic lymphoid cells exhibits regular nuclear outline and minimal cytoplasm(x400).



A



B

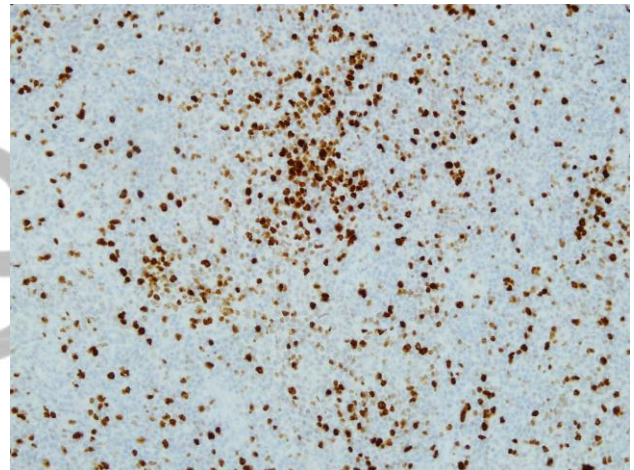
Figure 4:

(A) Immunostaining of CD20 show diffuse positivity of membrane and cytoplasm.

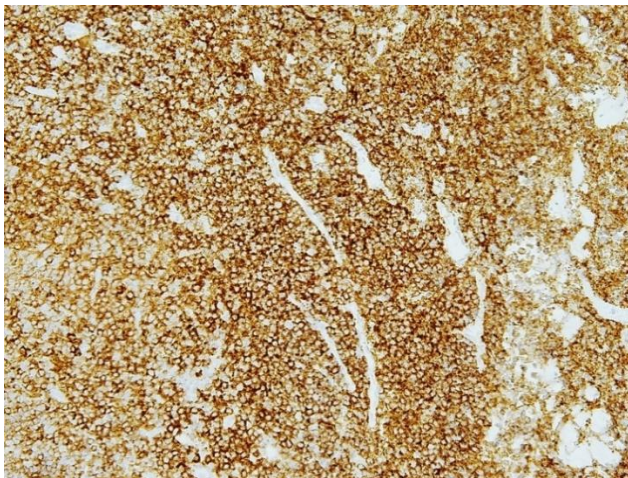
(B) PAX5 show nuclei staining.

(C) CD23 show diffuse positivity of membrane and cytoplasm.

(D) The ki67 staining is >30%, shows high proliferation rate of the cells in the proliferation centers.



D



C

3. Discussion and Conclusion

Malignant lymphomas represent approximately 5 percent of all malignant neoplasm of the head and neck and may involve nodal or extra-nodal sites. The head and neck is the second most frequent anatomical site of extra-nodal lymphomas and should be considered as the differential diagnosis of any patients who presented with neck mass especially in the young adult patients.

Small lymphocytic lymphoma (SLL) is synonym with Chronic lymphocytic leukemia (CLL). It is a neoplasm constituted of monomorphic small mature B cells that co-express CD5 and CD23 immunostains. CLL/SLL counts for 7% of non-Hodgkin lymphomas^{9,10}.

CLL is established by present of monoclonal B-cell count $\geq 5 \times 10^9/L$, with the characteristic morphology and phenotype of CLL in the peripheral blood. The term of SLL is used when the circulating CLL cell count $< 5 \times 10^9/L$ and involvement of nodal, and secondary lymphoid tissues such as the spleen, lymph nodes, and Waldeyer ring. The common extra-nodal involvements are skin, gastrointestinal tract, kidneys or CNS¹². SLL is diagnosed in 10- 20% of cases, and about 20% evolve into frank CLL¹³.

Microscopically, SLL shows diffuse architectural effacement by a proliferation of small mature lymphocytes with patchy paler proliferation centres¹⁴. The small lymphocyte exhibits scant cytoplasm, round nucleus, clumped chromatin, and occasionally a small nucleolus. There are some cases which the small lymphoid cells show moderate nuclear irregularity, which can lead to a differential diagnosis of mantle cell lymphoma. Mitotic activity is usually very low¹⁵. CD20 and CD23 expression is usually stronger in cells of the proliferation centres than in the diffuse areas. Follicular dendritic cell meshworks are present in some cases, and may be associated with the proliferation centres. About 2-8% of patients with CLL progress to DLBCL (Reiter's disease), and $< 1\%$ develop classic Hodgkin lymphoma¹⁶.

CT and MR are preferred imaging techniques for assessment of enlarged lymph nodes, bony abnormalities, and calcifications. The CT appearance of lymph nodes is isodense with muscle and necrosis is seldom seen. MR imaging is preferred to view evident of extension of lymphoma to different fascial planes and for intracranial extension. CT and MR imaging however cannot distinguish HL from NHL, and cannot differentiate their various subtypes, necessitating a pathologic diagnosis.

The diagnosis of lymphoma is based on the histopathology examination of tissue from enlarged lymph nodes or from extra lymphatic tissue in Non-Hodgkin's Lymphoma (NHL). The technical criteria required for the diagnosis includes proper selection of a biopsy site, preferably the largest accessible lymph node or a definitively involved extra-nodal site, good representative sample size and avoiding crushing of the specimen. Excisional biopsy of lymph nodes is the procedure most likely to yield a diagnosis, therefore communication between the surgeon and pathologist is essential to prevent an incorrect or delayed diagnosis.

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