Ahmad Rejab et al
Brunei International Medical Journal
2025;21:78-81

Brunei International Medical Journal

Case Report

Open Access

Supraglottic Non-Hodgkin Lymphoma

Ahmad Rejab RAF[1], **Lott** A[2], **Abdullah** K[2], **Musa** Z[2], **Mohamad** I[1*]

Abstract

Head and neck are the second most common region for the extra-nodal lymphomas and isolated presentation of laryngeal and hypopharyngeal lymphoma is rare. The most common laryngeal neoplasm is squamous cell carcinoma. We report a case of a 72-year-old Malay lady with primary extra-nodal supraglottic lymphoma who presented with a mass at the lingual surface of epiglottis with history of foreign body sensation in the throat and throat pain for three weeks. She was started on chemotherapy and responded well to the treatment.

Keywords: Laryngeal Neoplasm, lymphoma, B-cell, non-Hodgkin.

Author Details:
1 Department of Otorhinolaryngology-Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, Kota Bharu, Kelantan, Malaysia
2 Department of Otorhinolaryngology, Hospital Kemaman, Kemaman, Terengganu, Malaysia.

***Correspondence:**
Dr Irfan Mohamad,
irfankb@usm.my

INTRODUCTION

Primary extra-nodal lymphoma most commonly arises from the gastrointestinal tract with head and neck being the second most common, accounting about 2.5%.[1] Out of these, only 1% of all laryngeal tumours are attributed to the laryngeal lymphoma. Most common histopathological type of the primary laryngeal malignancy is of squamous cell carcinoma (SCC) type which accounts about 90%, making primary laryngeal and hypopharyngeal lymphoma rare.[1] It is very important to differentiate the histopathological entity in the laryngeal tumour as the treatment modalities for different type of malignancy differs.

CASE REPORT

A 72-year-old Malay lady with underlying diabetes mellitus, hypertension and ischaemic heart disease, presented with throat pain and foreign body sensation for three weeks. There was no history suggestive of aerodigestive obstruction, as well as other systemic symptoms. There was neither history of smoking nor family history of malignancy.

On examination, patient was stable hemodynamically, comfortable under room air with no signs of airway obstruction. Intraoral examination was normal, and no visible foreign body. Laryngoscopy showed a lobulated, reddish, non-tender, smooth surface mass at the right lingual surface of the epiglottis, with no contact bleeding (**Figure 1**). The mass extended into the right vallecular region, laterally to just anterior to the right aryepiglottic fold. Otherwise, vocal folds, pyriform fossae and arytenoids were normal bilaterally. There was no palpable neck swelling on neck examination with normal external laryngeal mark.

A biopsy was taken from the mass using biopsy forceps under 70-scope guidance, at the clinic setting using lignocaine spray as analgesia. Patient tolerated the procedure well. The biopsy result was reported as Diffused Large B-cell Lymphoma (DLBCL).

A computed tomography (CT) scan of the neck and thorax was done and reported as mild enhancing thickening of the right epiglottic fold (0.8cm) with fullness of the right para-glottic space. Minimal fluid was seen within the left pyriform sinus, the left aryepiglottic fold was not significantly thickened or enhanced. There were also scattered lung nodules which suggestive of metastasis. No bone erosion seen. The nasopharynx, Fossa of Rosenmuller (FOR), torus tubarious, oropharynx, and vocal cord appeared normal. The rest of the structures in the neck grossly were normal.

The patient was subsequently referred to Haematology team for further treatment. She was started on R-CHOP regime (Rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone) and successfully completed six cycles. Three months post chemotherapy follow up showed significant improvement with disappearance of the mass (**Figure 2**). However, the lung nodules persisted. She was not subjected to further treatment or cycle and was kept for surveillance.

DISCUSSION

Lymphomas encompass a group of lymphoproliferative malignant diseases that originate from T- and B-cells in the lymphatic system. Traditionally, lymphomas have been subcategorised into two groups: Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL).[2] HL is a malignancy is characterised histopathologically by the presence of Reed-Sternberg cells in the appropriate cellular background, while NHL is characterised by absence of Reed Sternberg Cells with abnormal clonal proliferation of B and T cells or both.[2] Most reported cases of laryngeal lymphoma have been of the B-cell phenotype, with only a small percentage being of the Natural Killer/T-cell phenotype with DLBCL being the most common immunophenotype.[3]

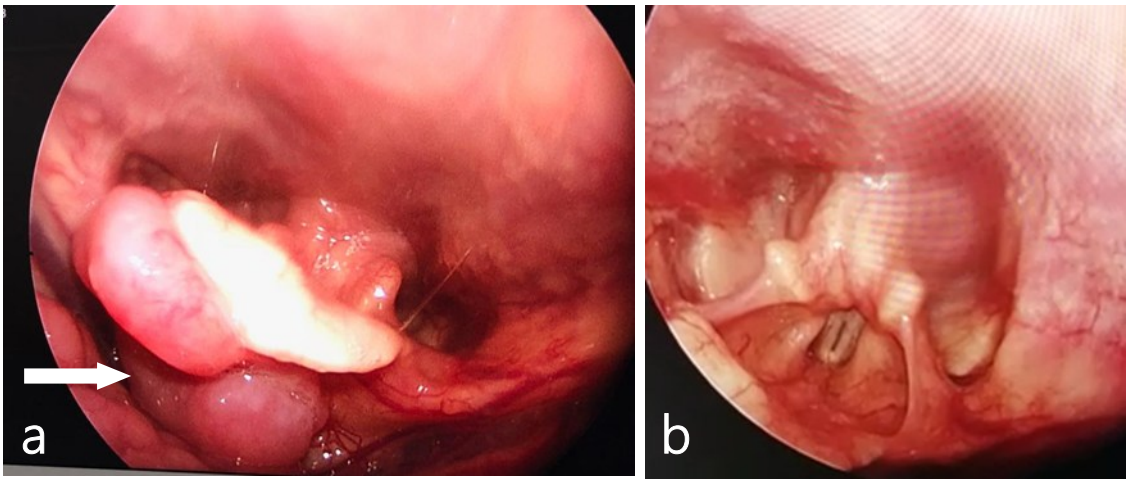

Figure 1 The mass seen over the right lingual surface of the epiglottis extending till the vallecula (arrow) (a), with a normal laryngeal inlet (b).

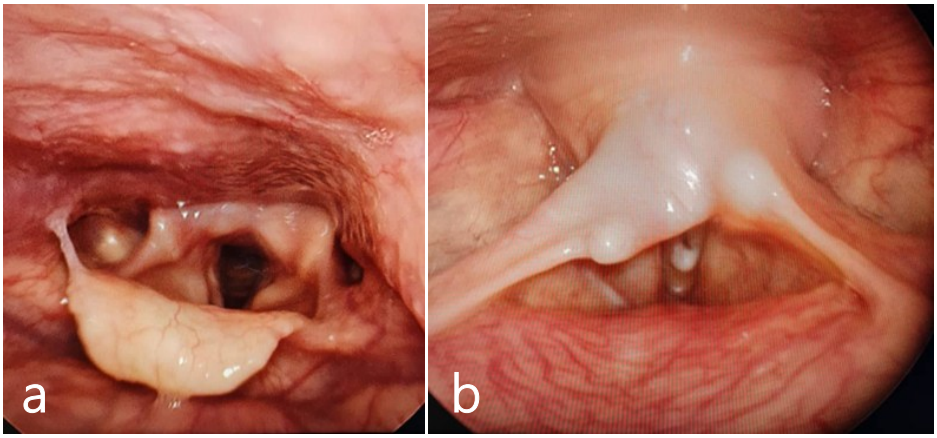

Figure 2 - Laryngoscopic findings three months after completion of chemotherapy showed no residual mass on epiglottis (a) and laryngeal inlet (b).

Fewer than 100 cases of lymphoproliferative tumours arising from the larynx, including both NHL and immunosuppression-related lymphoproliferative diseases, have been previously reported in the literature.[3,4] Extra-nodal NHL in epiglottis is extremely rare, accounting for 0.7% of all NHL and 1% of laryngeal tumours.[1,3] Supraglottic carcinoma is almost exclusively SCC which is about 90%, therefore, biopsy from the mass should be taken for histopathological examination as the treatment of the two are different. Despite being a part of the supraglottis, the epiglottis is rarely reported as the primary site of laryngeal lymphoma.[1]

Presentation of primary supraglottic lymphoma is the same as other supraglottic SCC which may include foreign body sensation, sore throat, hoarseness, dyspnoea, stridor, and dysphagia depending on the size and extension of the mass.[5] In our case, this patient came with complain of foreign body sensation and pain during swallowing which is typical presentation of laryngeal carcinoma. Patient may as well present with history of night sweats, fever, pruritus, fatigue which could suggest lymphoma, but in our case, the patient denied any of the B symptoms. Patient might present with painless multiple neck swelling due to the infiltration of the malignant cells to the regional lymph nodes which was absent in our case.

The mean age at diagnosis is 70 years with variable sex-ratio.[6] Our case was a 72-year-old female. Histologically, primary laryngeal lymphoma is more commonly of B-cell origin even though some T-cell and NK cell lymphomas were found before.[6] Commonly used markers for subtype diagnosis are leucocyte common antigen (LCA). B-cell markers (CD20 and CD79a), T-cell markers (CD3, CD5) and other markers like CD23, BCL2, BCL6, and CD10.[1] Imaging modalities such as CT scan is one of the investigative tools required to see the extension of the mass, the lymphatic spread and distant metastasis. CT scan is also commonly used to see the response of the disease after mid-cycle or end of therapy, however Positron Emission Tomography (PET) scan is a better imaging tool as it can distinguish between residual fibrotic masses from the masses containing viable tumour.[7,8] Other investigations include cardiac function should be done before starting the therapy because most chemotherapy drugs include anthracycline can damage the heart.[8] In this patient, the cardiac function was fairly normal, therefore chemotherapy was warranted.

After all the investigations were done, patients need to be staged according to the Cotswolds Modification of Ann Arbour Staging System.[8] Patients are categorised into limited (stages I and II) or advanced (III and IV) disease. The parameters are the distribution and numbers of the involved sites, the presence of extra-nodal involvement, and constitutional symptoms.[8] In our case, the patient was in stage II.

It is crucial to stage the disease correctly, as treatment varies according to the grade and stage. The most common therapeutic approaches for treatment of laryngeal lymphoma are radiotherapy or in combination with chemotherapy. In our case, the patient was given R-CHOP regime for six cycles with improvement seen three months post-chemotherapy.

Standard first-line chemotherapy for all CD20+ DLBCL is R-CHOP given every 21 days. For limited disease (stages I and II), the strategy is to give abbreviated chemotherapy (3-4 cycles) with anthracycline-containing regimen plus field radiotherapy (35-40Gy)

or a full course (6-8 cycles) of chemotherapy alone. This will result in a complete remission rate of 75-80% and 3-5year progression-free survival of 50-80%.[8]

R-CHOP treatment can usually be given to elderly patients up to 80 years of age in fit patients but with modulation of treatment according to geriatric assessment. In fit elderly patients aged more than 80 years old, a combination of rituximab with attenuated chemotherapy, R-mini CHOP can induce remission and improve survival rate. In unfit or frail elderly patients or those with cardiac dysfunction, replacement doxorubicin with gemcitabine, etoposide, or liposomal doxorubicin can be considered.[9]

There is no role of surgery in treating lymphomas. However, surgery may be needed if the patient comes with obstructed aerodigestive symptoms in terms of tracheostomy, laser debulking, or gastrotomy.[1,7,8]

Those who have complete remission post chemotherapy or radiotherapy need to be monitored regularly and need special attention to any long-term side effects of chemotherapy or development of secondary cancer like leukemia, breast and thyroid carcinoma.[8]

Radiological imaging such as CT scans at 6, 12, and 24 months after completed treatment is common practice, but there is no conclusive evidence to support this approach and it may even increase the risk of secondary malignancies.[9]

As for the treatment outcomes, the treatment method was not significantly important in predicting the result of therapy and prognosis. Kim *et al* reported that the survival rate of patients with T-cell lymphoma was poorer than that of patients with B-cell lymphoma. The high percentage of histologically aggressive lymphomas appears to be the cause of the poor prognosis associated with T-cell lymphoma.[10] Given that our patient's histology revealed B-cell lymphoma, she had an excellent prognosis and responded well to treatment.

CONCLUSION

Laryngeal malignancy is almost exclusively of SCC type, hence making epiglottic lymphoma a rare event. Thorough clinical evaluation as well as other supporting investigations such as biopsy and CT scan with proper staging, are mandatory to design the best treatment for the patient diagnosed with supraglottic lymphoma. The R-CHOP chemotherapy alone or combined with field radiotherapy is the first-line treatment for DLBCL so far with no role of surgery involved, unlike SCC. Patients who survived, need regular monitoring so that relapse can be detected early.

Abbreviations

SCC	Squamous cell carcinoma
DLBCL	Diffuse large B-cell lymphoma
CT	Computed tomography
FOR	Fossa of Rosenmuller
CHOP	Rituximab, cyclophosphamide, doxorubicin, vincristine & prednisolone
HL	Hodgkin lymphoma
NHL	Non Hodgkin lymphoma

Declarations

Conflict of interests

The authors declare no conflict of interests.

Consent

Consent has been obtained from patient's parents for publication.

Acknowledgement

None.

References

1. Liu C, Delaney S, Brown TN. An Unusual presentation of B-Cell lymphoma as a large isolated epiglottic mass: Case report and Literature Review. Case Rep Otolaryngol. 2016;2016:9787432.
2. Peh, SC Poppema S. Lymphoma: an introduction into historical background, classification schemes, aetiology, geographical variation and epidemiological trend. Malaysian J Pathol. 2001;23:49-63
3. Siddiqui NA, Branstetter BF 4th, Hamilton BE, Ginsberg LE, Glastonbury CM, Harnsberger HR, et al. Imaging characteristics of primary laryngeal lymphoma. Am J Neuroradiol. 2010;31:1261-5.
4. Horny HP, Kaiserling E. Involvement of the larynx by hemopoietic neoplasms. An investigation of autopsy cases and review of the literature. Pathol Res Pract. 1995;191:130-8
5. Mark P, Najihah Hanim A, Eshamsol Kamar O, Suhaila A, Irfan M. A rare case of primary extranodal laryngeal non hodgkin lymphoma. Inter Med J Malaysia. 2018;17:85-8.
6. Kato S, Sakura M, Takooda S, Sakurai M, Izumo T. Primary Non -Hodgkin Lymphoma of the larynx. J Laryngol Otol. 1997;111:571-4.
7. Junior NT, Zago TM, Pauna HF, Crespo AN. Non Hodgkin Lymphoma in Supraglottis: Case Report. Otolaryngol (Sunnyvale) 2017;7:285
8. Fadilah SAW. Fundamental management of Non-hodgkin lymphoma. Med J Malaysia. 2009; 64:333-40.
9. Tilly H, Gomes da Silva M, Vitolo U, Jack A, Meignan M, Lopez-Guillermo A, et al, ESMO Guidelines Committee. Diffuse large B-cell lymphoma (DLBCL): ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol. 2015;26 Suppl 5:v116-25.*
10. Kim KH, Kim RB, Woo SH. Individual Participant Data Meta-Analysis of Primary Laryngeal Lymphoma: Focusing on the Clinical Characteristic and Prognosis. Laryngoscope. 2015;125:2741-8.