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# Primary laryngeal T-cell lymphoma: A case report and review of the literature

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ABSTRACT

*INTRODUCTION:* Primary lymphoma of the larynx is extremely rare, representing less than 1% of all primary laryngeal neoplasms. It consists mainly of non-Hodgkin lymphomas (NHLs), represented particularly by diffuse large B-cell. Extranodal natural killer/T-cell lymphoma, presented in larynx is a rare condition that accounts for less than 11% of all lymphomas without distinctive clinicopathologic features, as well as challenging pathologic diagnosis.
*CASE REPORT:* We report here a case of a 64-years-old man who presented with primary lymphoma type T of the subglottic larynx. A histopathological examination of the biopsy confirmed non-Hodgkin T cell lymphoma. Given his age, he underwent chemotherapy and radiation therapy. The patient was disease-free after 18 months follow up.
*CONCLUSION:* The clinicopathological characteristics and rational treatment of primary laryngeal lymphoma are still unclear and need to be further defined due to the paucity of this pathology.


## 1. Introduction

Primary lymphoma of the larynx is extremely rare, representing less than 1% of all primary laryngeal neoplasms [1]. It consists mainly of non-Hodgkin lymphomas (NHLs), represented particularly by diffuse large B-cell and mucosa-associated lymphoid tissues and are predominantly located in the supraglottic region, as this area of the larynx contains follicular lymphoid tissue [2]. Extranodal natural killer/T-cell lymphoma, located in larynx is a rare condition that accounts for less than 11% of all lymphomas without distinctive clinicopathologic features, as well as challenging pathologic diagnosis [3].

In this article we report unrelated case of subglottic laryngeal lymphoma seen in our academic medical center.

This study has been reported in accordance with the SCARE criteria [4].

## 2. Case report

A 64-years-old man, who was a heavy smoker, was admitted in 2018 to otolaryngology department for hoarseness and shortness of breath for 12 months. The patient did not suffer from fever, weight loss, or night sweats. There was no other personal nor familial medical history. On physical examination, there was no palpable lymphadenopathy and no hepatosplenomegaly.

Flexible fiberoptic nasolaryngoscopy revealed a large exophytic mass limited in the subglottic region without complete airway obstruction (Fig. 1); the motion of the vocal folds was normal. A laryngeal computed tomography (CT) confirmed the existence of the subglottic tumor with no involvement of the vocal cords and no lymph node (Fig. 2).

A direct laryngoscopy has been performed revealing a subglottic, submucosal lump. No other tumor was found in the rhinopharynx or in the Waldeyer's ring lymphoid tissue. A biopsy revealed a mantle cell type T NHL with the following antigen constellation: cluster of differentiation (CD)3+, CD8+ and the granzyme B+; cells did not express CD20, CD5, PAX5, CD4, CD10 or CD30. Ki67 estimated at 70% (Fig. 3).

Full body PET scan did not showed any abnormality. Complete blood count (CBC) was normal thus, bone marrow biopsy did not show any anomalies.

The patient was referred to the hematology department for completion of staging and treatment where he received chemotherapy (cyclophosphamide, doxorubicin, vincristine and prednisone) and radiation therapy (60 Gy). The patient was disease-free after 18 months follow up.

## 3. Discussion

The most frequently encountered cancer of the larynx is squamous cell carcinoma; less commonly adenocarcinoma, adenoid

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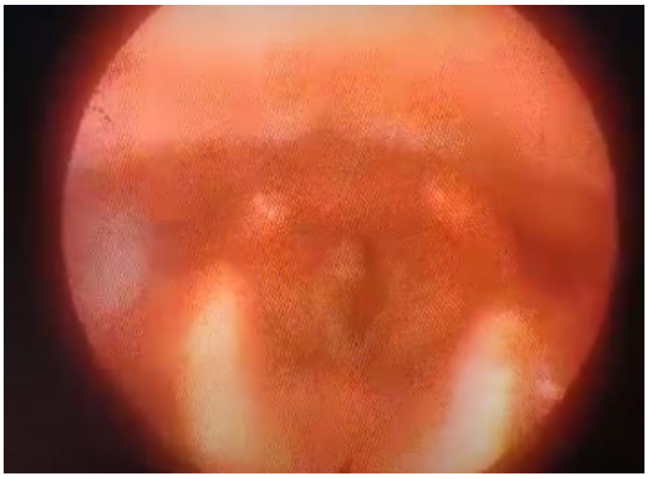

**Fig. 1.** Large exophytic mass limited in the subglottic region without complete airway obstruction.

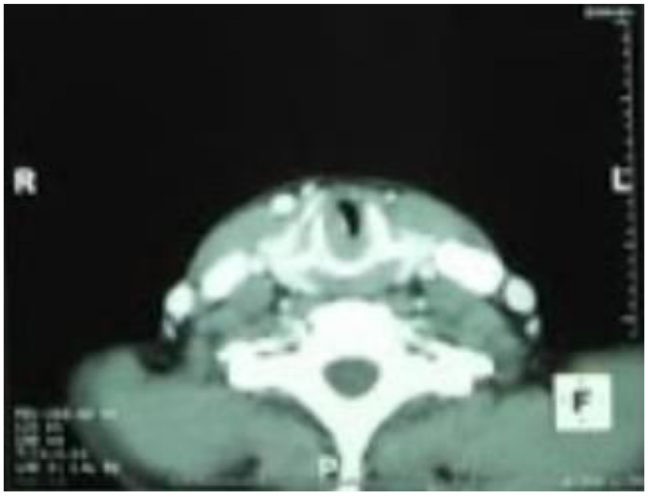


**Fig. 2.** CT showing the subglottic tumor.

cystic carcinoma, and neuroendocrine tumors [5]. As known, the head and neck is the second most common extranodal site of involvement in non-Hodgkin lymphoma (NHL). Most structures typically affected are Waldeyer's ring, ocular adnexal structures, nasal cavity, paranasal sinuses, nasopharynx, thyroid gland, as well as salivary glands [6,7], yet the larynx is rarely involved. A 10-years cohort study involving 2631 laryngeal biopsies revealed only one case of diffuse large B cell lymphoma [8]. The supraglottic larynx is most commonly involved [9]. Mean age is in the seventh decade, without difference in distribution between males and females [10]. The most common symptom is hoarseness and/or dysphagia, other patients also suffer from chronic cough unresponsive to oral corticosteroid [11,12].

Endoscopic examination usually shows a smooth surface or submucosal mass; ulcerated lesions are extremly rare. Even when imaging techniques such as CT and magnetic resonance imaging (MRI) may help in the assessment of any laryngeal neoplasm [13]; the definitive diagnosis requires histological examination of a biopsy. Data of a retrospective study including 31 cases showed that CD3 and cytotoxic granules (granzyme B and/or TIA-1) were present in all cases, while CD20 was absent. CD56 was present in 28 cases (90.3%), CD5 was present in 4 cases. CD43 was present in 3 cases and neoplastic cells contained CD2 and CD7 present in 2 cases. The Ki-67 index ranged 30%–80% [3]. Authors suggested that

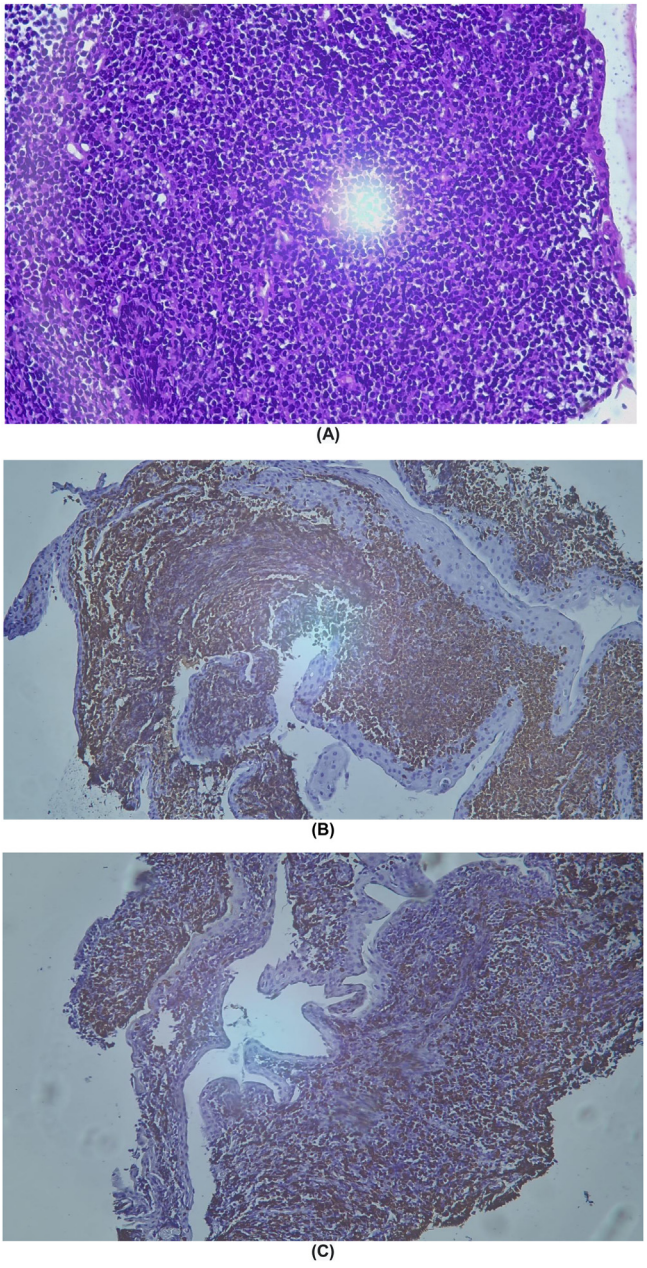


**Fig. 3.** Basophil cells with atypical hyperchromatic nuclei. The stroma is small and rich in vessels (x200) (A). Tumor cells diffusely express CD3 (B). The ki-67 proliferation index is estimated at 70% (C).

the outcome of laryngeal T lymphoma was bleak, more than half of them (73.1%) died with earlier treatment either from tumor dissemination or severe laryngeal complications, such as bleeding or obstruction [5,14].

## 4. Conclusion

Larynx is a rare location of extranodal site of NHL. The clinicopathological characteristics and rational treatment of primary laryngeal lymphoma are still unclear and need to be further defined and improved due to the paucity of this pathology and the poor prognosis.

## Declaration of Competing Interest

The authors declare no conflict of interest.

## Funding

This study did not receive any sources of funding.

## Ethical approval

This type of study does not require any ethical approval by our institution.

## Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Author contribution

K. Chaker: drafting the article.
M. Beghdad: acquisition of data.
M. A. Mennouni: study design.
A. Mkhatri: revising the article.
Y. Oukessou: study concept, writing the article.
M. Mahtar: final approval.

## Registration of research studies

Not Applicable.

## Guarantor

A. Mkhatri.

## Provenance and peer review

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