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PRIMARY NON-HODGKIN'S LYMPHOMA OF THE LARYNX – TWO DIFFERENT CASE SCENARIOS

Otolaryngology

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ABSTRACT

Primary NHL arising from larynx is unusual. Prevalence of NHL among immunocompromised patients is 19%, finding a plasmocytic lymphoma of larynx is extremely rare. Due to scarcity of lymphoid tissue in the glottis, most primary lymphoid tissue arises either in supraglottic and sub glottic region. This case series is being presented in view of the rarity of the case and paucity of literature. Here we retrospectively analysed the background, clinical profile of patients, management and histopathological confirmation of Non-Hodgkin's lymphoma of the larynx.

KEYWORDS

Larynx, Non-hodgkins Lymphoma, Coblation

INTRODUCTION

Lymphomas arise from the proliferation of lymphoid tissue. Head & neck region is a favorable place for the development of lymphoma . The reason is the presence of extensive lymphatics in head & neck region .As a variation from the other regions , larynx has sparse lymphatics and hence the incidence of lymphoma is less(1) .There is only few hand full of cases reported till date about non Hodgkin's lymphoma of the larynx(2).The objective is to study the clinical profile , histopathological confirmation ,surgical aspect and prognosis of patients who are diagnosed with Non Hodgkin's lymphoma of larynx.Non-Hodgkin lymphomas (NHL) and Hodgkin disease are two types of malignant lymphomas . Laryngeal primary lymphomas are primarily NHL and are predominately found in the supraglottic region of the larynx because follicular lymphoid tissue is present there. Diffuse large B-cell and mucosa-associated lymphoid tissue (MALT)-type marginal zones are two NHL subtypes. The most frequent primary laryngeal hematopoietic tumor's are B-cell lymphomas. Rare reports exist for other forms of lymphomas, such as T- or natural killer (NK)-cell lymphomas. The most common presentations are dysphagia, dysphonia, hoarseness of voice & cervical lymphadenopathy (3–7).Unlike to the usual symptoms, the first patient presented with breathing difficulty, which is due to the position(interarytenoid region) of the lymphoma Retrospectively we analysed the case records of patients who presented with laryngeal mass in the department of Otorhinolaryngology,St John's Medical college.

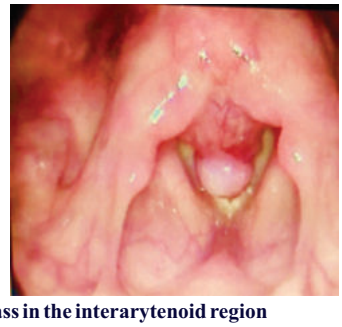

Figure 1: Mass in the interarytenoid region

Table – 1 Clinical Data Of Cases

Case	Age/ sex	Comor bidity	Location Of Lymphoma	Duration Of Symptom	Histopathology
1	37/F	PLHIV	Interarytenoid region	3 months	Non-Hodgkin's lymphoma with plasmacytic differentiation. IHC -positive for CD3+, CD138,LCA & MUM1 and negative for CD 15,CD 30,CD56.
2	72/F	Hep B positive K/C/O Bcell nodal marginal lymphoma,Hypothyroidism,HTN	Subglottic region	20 days	Low grade B cell lymphoma CD 20 +,BCL-2 +

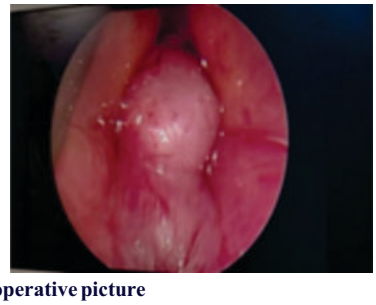

Fig 2-Intra operative picture

Case Study

CASE 1: 37-year-old Known case of PLHIV on treatment, presented with complaints of progressively increasing hoarseness of voice and shortness of breath for 3 months. She was unable to lie down in recumbent position. On examination neck was Normal with no laryngeal framework widening.

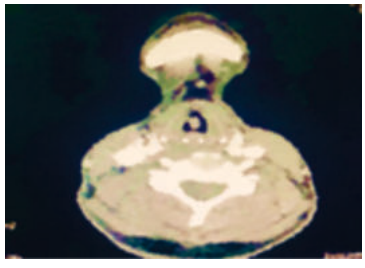

Fig 3-CECT Neck

Office laryngoscopy revealed a smooth pedunculated mass from the interarytenoid region obstructing the laryngeal inlet. Based on history & clinical finding, we suspected interarytenoid benign lesion .CECT neck revealed an enhancing lesion in the inter arytenoid region. It is noted projecting anteriorly just above the vocal cord, causing narrowing of laryngeal inlet. In view of difficult intubation tracheostomy + coblation assisted excision + biopsy of the mass was performed. Histopathology report came as Non-Hodgkin's lymphoma with plasmacytic differentiation. IHC was positive for CD3+, CD138,LCA & MUM1 and negative for CD 15,CD 30,CD56.Case was discussed in multidisciplinary tumor board and chemotherapy was done with CHOP regimen.6 months after her treatment follow up PET-CT was done, which showed regression of the disease

Case 2:

A 72 year old female with presented to the Emergency Medicine

department with stridor. She had complaints of progressively worsening change in voice for 20 days along with low grade fever with productive cough.

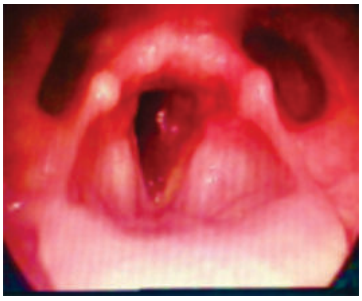

Fig 4-Mass in subglottic region

She was a known case of B cell nodal marginal zone lymphoma and has completed chemotherapy. On examination neck was normal. Office video laryngoscopy- was suggestive of a subglottic mass with more than 50 % occlusion of the lumen. CECT neck revealed an enhancing lesion in the in the sub glottis ,extending from c4-c5 to c5-c6 level likely neoplastic origin. There was loss of fat plane in left vocal cord. And subglottic narrowing.

In view of stridor-emergency tracheostomy was done with direct laryngoscopy and biopsy of the lesion. Histopathology report came as Low grade B cell lymphoma and was CD 20 +,BCL-2 +.The case was discussed in multidisciplinary tumour board,received radiation therapy by IMRT technique.PET-CT was done 4 months later.There was no abnormal tracer avid disease in subglottic region. This was followed by decannulation of tracheostomy tube after 6 months.

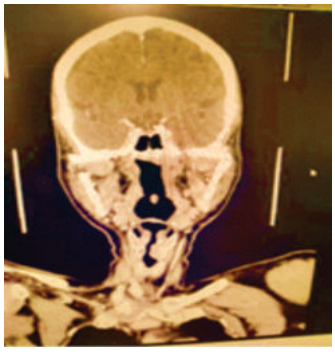

Fig 5- Histopathological appearance of cd 20 positive lymphoma

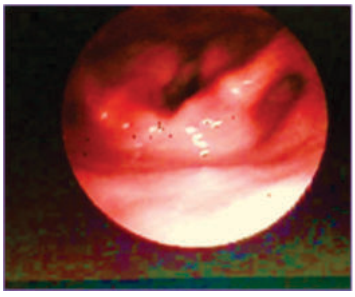

Fig 6-Enhancing lesion in the subglottic region

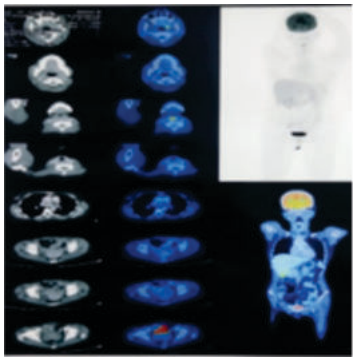

Fig 7- Post operative picture after 1 month

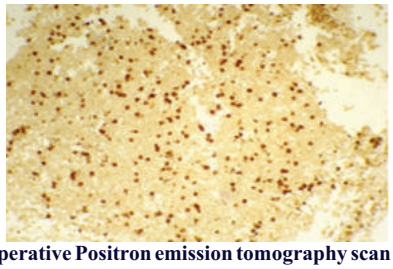

Fig 8 Pre-operative Positron emission tomography scan

DISCUSSION

In our study we are presenting two cases of rare laryngeal lymphoma. First one was a PLHIV patient, whose histopathology report was consistent with plasmacytic differentiation. She was treated with chemotherapy

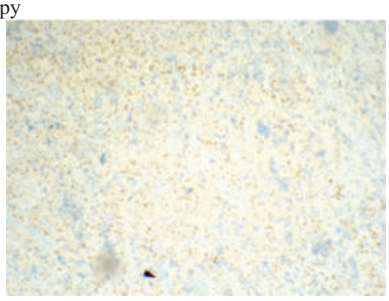

Fig 9-Histopathological appearance of MUM 1 positive lymphoma

There are several different types of B-cell non-Hodgkin lymphomas with plasmacytic differentiation, and their morphologic characteristics can vary greatly. Differentiating lymphoplasmacytic lymphoma from marginal zone lymphoma and other low-grade B-cell lymphomas might be difficult from a diagnostic standpoint. Second case was a Hepatitis B positive patient and the histopathology report was Low grade-B cell lymphoma. The most prevalent non-epithelial tumor's in the head and neck are lymphomas(8).The most common mode of presentation in lymphomas is lymphadenopathy.25 % of cases has extranodal involvement(9–11).In our case series,both patient had negative nodal ststus. In the majority of these instances, lymphomas are found in areas that typically have lymphoid tissue. Extranodal lymphomas, however, can also develop in healthy, non-lymphoid tissues like the thyroid(12–14). The vast majority of extranodal lymphomas in the head and neck are NHLs, with Waldeyer's ring being the most often involved site. Extranodal NHL accounts below 1 % of all laryngeal neoplasms(9).In our cohort study the incidence of Non-Hodgkin's lymphoma of larynx was approximately 0.2 %.The age of onset of laryngeal lymphoma varies between 37 to 67 years, with the mean age of occurrence to be 71 years. In our study both patients are females. Data from previous studies also showed female preponderance(9,15,16) in line with our study. The usual symptoms at the time of presentation are dyspnoea, hoarseness of voice, cough, dysphagia, stridor or systemic signs such as weight loss and fever. There were no systemic symptom/sign for both patients. Duration of symptom varied between 20 days to 3 months

Due to their lack of clinical and gross differential criteria in comparison to SCC, primary laryngeal lymphomas provide a diagnostic challenge. In a study conducted by Horny et al(1,17) mentioned the appearance as non proliferative, smooth pedunculated mass. In our study also supraglottic mass had a similar appearance. However imaging techniques such as CT/MRI is useful in differentiating this from neoplasms of larynx. The definitive diagnostic modality is by biopsy followed by histopathological examination. In our study, we did excision of the mass using coblation, unlike in our previous studies. It had better post operative outcome in terms of language and swallow outcomes(18).Several pathological classification schemes have been employed, it is difficult to compare the histological findings of prior studies of laryngeal lymphomas. In order to classify B-cell or T-cell lineage fractions more precisely and forecast the lymphoid nature of the cells, several antibodies are utilised. According to reports, B-cell lymphomas are more common than T-cell lymphomas (6:1)(1). Due to the various pathological classification schemes that have been employed, comparing the histological results of prior studies of laryngeal lymphomas is challenging. Immunohistochemical analysis of paraffin-embedded or

frozen tissue is required for an appropriate diagnosis. The lymphoid nature of the cells can be predicted using a variety of antibodies, which can also provide more detailed information regarding the classification of B-cell or T-cell lineage subset. The two most popular therapeutic modalities used to treat primary laryngeal lymphomas are radiation therapy and chemotherapy . As the best treatment, radiotherapy has been recommended. Several studies have shown that lymphomas that have spread to the larynx are among the most radiosensitive cancers, and that moderate-dose therapy (30–50 cGy) is typically required to control them(11,16,19,20).In our institution we treated first patient with Chemotherapy and second one with radiotherapy. Post treatment follow up was done with PET-CT, and had shown regression for both.

CONCLUSIONS

These cases show a diverse modality of presentation of Non Hodgkin's lymphoma of the larynx. The reason why most of the patients with lymphoma presents to ENT because of neck node. Hence these 2 cases are rare presentations

Case 1 - Presented with hoarseness of voice with a benign looking mass on examination which can easily get misdiagnosed .However early treatment & timely intervention can provide complete recovery.

CASE 2 – Suspicion of lymphoma was made as she was a known case of the same.

Obstructive lesions in the larynx is challenging in view of the location and clinical presentation with stridor.

Thorough clinical evaluation with laryngoscopy and imaging is essential to plan further management Once airway is secured, direct laryngoscopy and biopsy can be planned for tissue diagnosis. This should be followed by chemoradiotherapy for complete cure

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