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The Great Mimicker: Primary Extranodal Burkitt Lymphoma Masquerading as Base of Tongue Carcinoma

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Abstract:

- ***Introduction:***
Primary extranodal Non-Hodgkin's Lymphomas (NHLs) of the oral cavity are extremely rare (<1% of cases). In adults, Burkitt lymphoma accounts for under 2% of malignant lymphomas in India. Due to their scarcity, base of tongue lesions frequently present as diagnostic dilemmas, often mimicking common epithelial malignancies like squamous cell carcinoma.

- ***Aim:***
To highlight a rare presentation of primary extranodal Burkitt lymphoma of the base of the tongue, initially masquerading as squamous cell carcinoma, and emphasize the necessity of timely histopathological and immunohistochemical evaluation.

- ***Methodology:***
A 35-year-old male presenting with foreign body sensation underwent clinical examination, OPD toluidine blue staining, contrast-enhanced MRI (CEMRI) of the oral cavity/neck, and a direct laryngoscopic biopsy. The tissue sample was evaluated via histopathology (H&E) and a targeted panel of immunohistochemical (IHC) markers.

- ***Result:***
Clinical examination revealed a non-tender 1 cm X 2 cm ulcerative growth at the right base of the tongue. CEMRI suggested a neoplastic lesion consistent with tongue base carcinoma. However, histopathology showed diffusely infiltrating atypical lymphoid cells with tingible-body macrophages. IHC confirmed Burkitt lymphoma.

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I. INTRODUCTION

Non-Hodgkin's lymphomas (NHLs, 86%), the predominant lymphomas, are the second most frequent oropharyngeal malignancies [1]. In developed world, peripheral T-cell lymphomas (PTCLs) account for 15–20% and 5–10% of aggressive lymphomas and NHLs, respectively. While, the Asian countries have greater disease burden, with around 15–20% of all lymphomas classified as PTCL or NK/T-cell lymphoma (NKTL) [2]. Though, T-cell NHL has a predilection toward Asian race, the pattern observed in Indian population is distinct from the rest of the Asia [3].

Around 23–30% patients with NHL have extranodal involvement, and frequently affect skin, Waldeyer's ring, gastrointestinal tract, and bones. While oral cavities are affected in only 2% patients [1]. Thus, rare involvement of oral cavity poses a diagnostic dilemma for the clinicians. Herein, we present a rare case of base of tongue carcinoma presenting as Burkitt Lymphoma.

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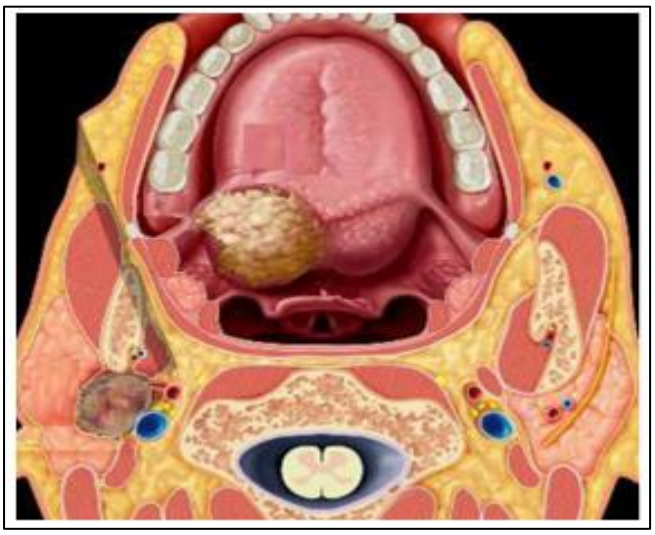

Fig 1 Base of Tongue Growth [4]

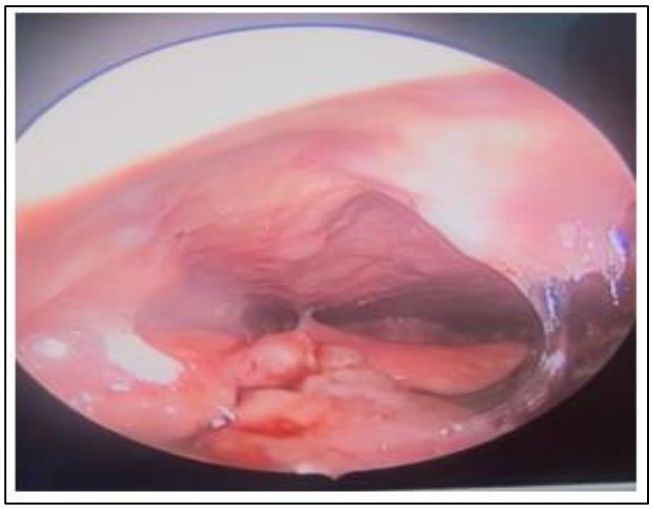

Fig 2 Laryngoscopic Picture

II. CASE REPORT

35-year male presented in ENT OPD with chief complain of foreign body sensation in throat since 3 months. The intraoral examination revealed an ulcerative growth at right side base of tongue of approx. 1cm x 2cm, with irregular margins, non- tender. Addiction history was absent. Patient underwent toluidine blue staining in OPD that was positive.

Contrast enhanced MRI oral cavity and neck (Figure 3) suggestive of a well-defined lobulated soft tissue signal intensity mass lesion is seen in involving base of the tongue, on right side of midline extending in right vallecula showing homogeneous enhancement. A mild enlarged right level II lymph node is seen. Posteroinferiorly, it growing in the preepiglottic fat and abutting epiglottis. Findings suggest neoplastic mass lesion like Carcinoma base of tongue.

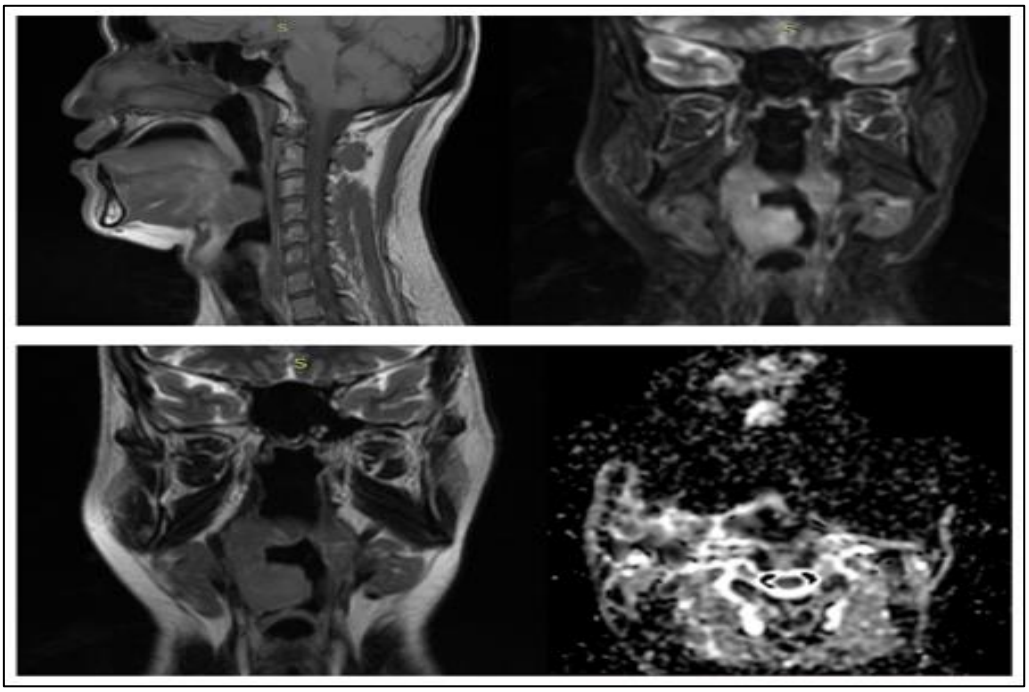

Fig 3 CEMRI Oral Cavity & Neck with T2 Weighted Fat Suppression with Diffusion Weighted Imaging

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Patient was planned for Direct laryngoscopic biopsy. Histopathologic section [Figure 4] shows a malignant tumor composed of diffusely infiltration by numerous medium sized atypical lymphoid cells admixed with tingible-body macrophages. The tumor cells exhibit uniform size ("monotonous"), round nucleus, small nucleoli, relatively abundant cytoplasm and Brisk mitotic rate. Suggestive of Non-Hodgkin's B-Cell lymphoma.

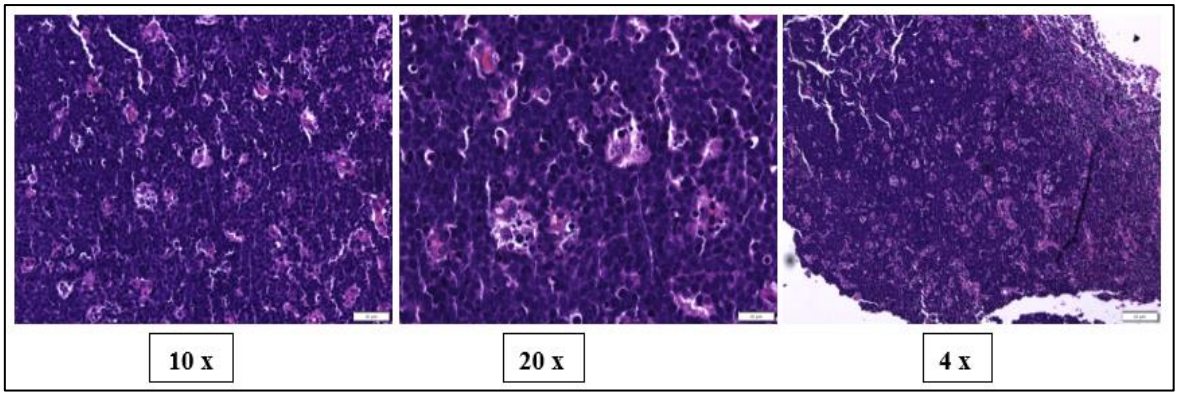

Fig 4 Histopathological Examination

On Immunohistochemistry, Favour Burkitt's lymphoma [Table 1] [Figure 5].

Table 1 IHC Marker & Results

IHC MARKER	RESULT
CD20 (B-cell marker)	Strong & diffuse membranous staining
C-Myc (Burkitt lymphoma marker)	Positive (diffuse)
MIB-1 labelling index	100 %
Pancytokeratin	Negative
CD3 (T-cell marker)	Negative
TdT (Terminal deoxynucleotidyl transferase)	Negative
Synaptophysin	Negative
CD-99	Negative

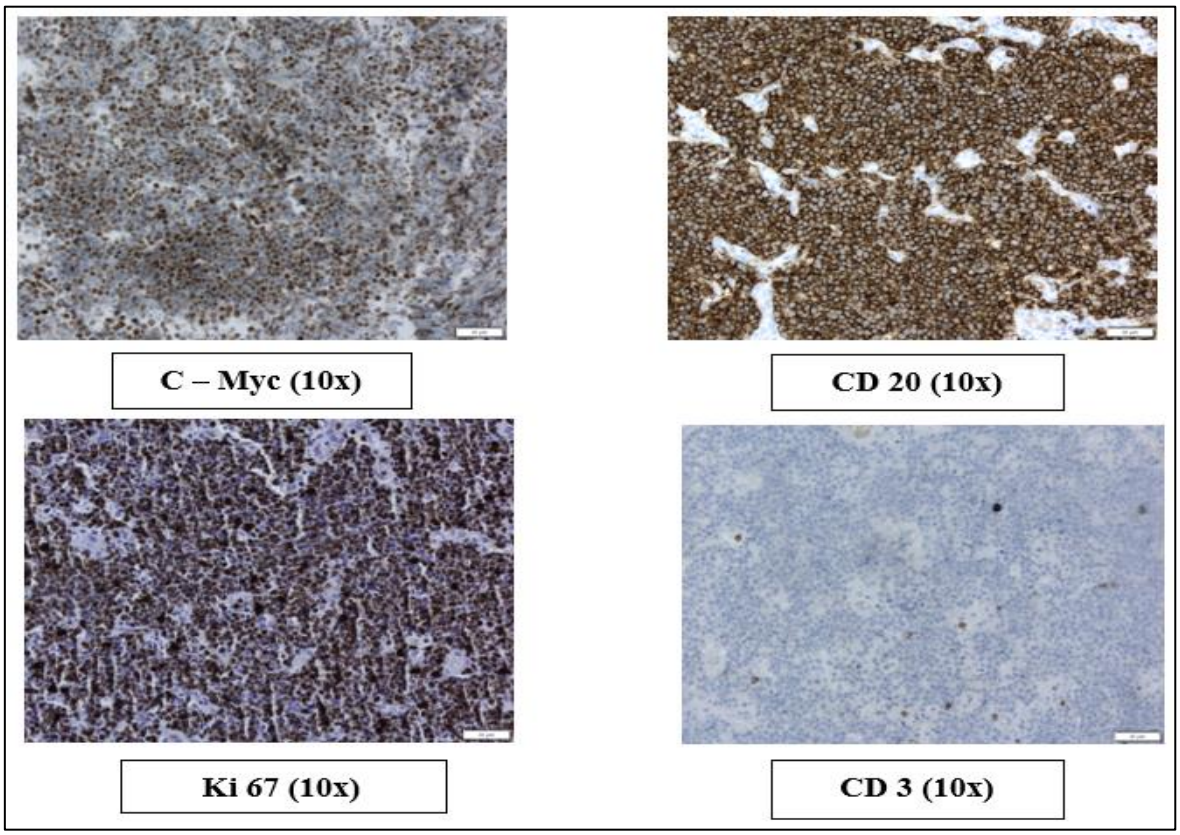

Fig 5 IHC

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Based on Ann Arbor staging system, the patient was classified as stage I with A symptoms. Thus, the patient was transferred to the medical oncology team and chemotherapy involving CHOP regimen (cyclophosphamide, hydroxydaunorubicin, vincristine, and prednisolone) was initiated.

III. DISCUSSION

Globally the proportion of lymphoma at extranodal sites has ranged from 20 % to 50%, however involvement of oral sites accounts for a mere proportion of <1 % [5]. In India, Burkitt lymphoma subtype constitutes < 2% of malignant lymphomas in adult population (15-59 years group) [6].

Epidemiologically, the presentation of Burkitt lymphoma follows a distinct geographic pattern [13]. While the endemic form is prevalent in equatorial Africa and typically involves the jaw in children, the sporadic form seen globally more commonly involves the abdomen. In India, adult Burkitt lymphoma is incredibly rare, representing less than 2% of all adult lymphomas, and its primary presentation as an extranodal lesion in the oral cavity—specifically the base of the tongue—is exceptional [14-15]. This distinct presentation underlines the necessity of documenting local demographic variances, as a low regional incidence can easily cause clinicians to overlook this highly aggressive but highly curable disease. Hence, the incidence of base of the tongue Burkitt lymphoma is indeed a rare incidence.

Burkitt lymphoma, particularly when presenting in the base of the tongue, remains a clinical challenge due to its aggressive nature and the complex anatomy of oropharyngeal anatomy.

The unique location can lead to late diagnosis, as symptoms may initially be attributed to more common conditions or benign tumours. Considering the benefit of prompt initiation of therapy, biopsies and diagnostic evaluations should be expedited when Burkitt lymphoma is suspected. Immunohistochemistry and cytogenetics play a significant role in the treatment and management of Burkitt Lymphoma.

The optimal imaging modality for the evaluation of tongue base tumour is MRI which delineates tumour extension more precisely than other modality however evaluation with CT scans also holds a significant role in evaluating lymph nodes status in head and neck region. With its rapid growth, Burkitt Lymphoma is a highly chemosensitive disease and was one of the early cancers in which cures were achieved with chemotherapy alone [7-8]. Appropriate treatment likely requires evaluation of a numbers of parameters including, age health status of the patient, co morbidities as well as high risks features such as CNS involvement.

The clinical and radiological presentation of primary extranodal lymphoma of the tongue base frequently overlaps with that of squamous cell carcinoma, creating a substantial diagnostic trap. In our patient, the presence of a lobulated, homogeneously enhancing base-of-tongue mass extending into the vallecula and preepiglottic fat, paired with regional lymphadenopathy, strongly mirrored an advanced squamous cell carcinoma. Because squamous cell carcinoma is epidemiologically dominant at this site, a clinician's natural inclination leans toward epithelial malignancies. This case demonstrates that relying solely on cross-sectional imaging can be deceptive, and emphasizes the mandate for prompt, deep tissue biopsies to differentiate haematological malignancies from solid tumors before any irreversible surgical intervention is contemplated.

Immunohistochemical (IHC) profiling is the definitive tool to differentiate Burkitt lymphoma from other small round blue cell tumors and aggressive high-grade lymphomas, such as Diffuse Large B-Cell Lymphoma (DLBCL). In this case, the tumor exhibited strong, diffuse membranous staining for CD20, establishing its B-cell lineage. Crucially, the definitive diagnosis of Burkitt lymphoma was solidified by a MIB-1/Ki-67 labelling index of 100% and diffuse positivity for C-Myc. A Ki-67 index approaching or reaching 100% reflects the extraordinarily high proliferation rate classic to Burkitt lymphoma, distinguishing it from DLBCL, which typically displays a lower, more variable proliferative fraction. The exclusion of T-cell markers (CD3 negative) and terminal deoxynucleotidyl transferase (TdT negative) further ruled out peripheral T-cell lymphomas and lymphoblastic lymphomas, respectively.

Approximately two-thirds of NHL is nodal at presentation and one-third extranodal, again in contrast to HL, where extranodal presentation is rare [9]. There is no absolute protocol for the treatment of the primary NHL at the base of the tongue [10]. There are distinct approaches in the treatment of oral cavity non-Hodgkin lymphomas. Certain studies suggest multiple chemotherapy combined with radiotherapy for primary lymphomas of Waldeyer's ring [10-12].

IV. CONCLUSION

While squamous cell carcinomas represent the vast majority of malignancies involving the base of the tongue, primary extranodal Non-Hodgkin Lymphomas (NHLs) at this site remain exceedingly rare. They typically present with insidious, non-specific oropharyngeal discomfort, frequently mimicking benign conditions or standard carcinomas under normal-like mucosal surfaces.

In the adult Indian population, the Burkitt lymphoma subtype is exceptionally uncommon, constituting less than 2% of all malignant lymphomas. This case highlights a profound diagnostic dilemma, where clinical and radiological features strongly pointed toward a base of tongue carcinoma, but definitive histopathology and immunohistochemistry (characterized by a 100% MIB-1 proliferation index and diffuse C-Myc positivity) revealed Burkitt Lymphoma.

Given that Burkitt lymphoma is a highly aggressive yet highly chemosensitive malignancy, establishing a high index of clinical suspicion is paramount. Clinicians must expedite

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tissue biopsies, tolluidine blue staining, and immunohistochemical profiling, as early differentiation from squamous cell carcinoma radically alters management—preventing unnecessary surgical resections and facilitating immediate, life-saving systemic chemotherapy.

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