

## Primary Soft Palate Lymphoma: A Case Report of Unique Entity in A Tertiary Institution, South-South, Nigeria

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### Abstract

### Case Report

Lymphomas are malignant neoplasms of the lymphocytic cell lines, involving mainly lymph nodes and also extra nodal sites. They are generally classified as either Hodgkin's lymphoma, or Non-Hodgkin's lymphoma; which may be of either B – lymphocyte or T – lymphocyte in origin. Oropharyngeal lymphomas are relatively rare and diagnosis is a challenge. Most frequently arising from the Waldeyer's ring after gastrointestinal tract is the second most common site for extra nodal lymphomas. We report a case of primary Non –Hodgkin's lymphoma arising from the soft palate in a 71- year- old male who presented with progressive difficulty in swallowing, slurred speech and choking on lying supine. There was no other lymphomatous lesion detected elsewhere in the body. Postnasal X- ray revealed a pedunculated mass arising from the soft palate down the laryngeal inlet. Excisional biopsy of the lesion showed sections of lymphoid tissue with partial effacement of the architecture replaced by ill- defined follicles and diffused pattern in some areas. Follicles are of various sizes and shapes, containing varying proportions of centrocytes and centroblasts suggestive of follicular mixed type of primary extra nodal lymphoma. Immunohistochemistry confirmed the diagnosis. Our patient has been followed up for 15months and currently doing well.

**Keywords:** Extra nodal Non-Hodgkin's lymphoma; Soft palate; Follicular lymphoma.

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## INTRODUCTION

Lymphoma is the second most common neoplasm of the Head and Neck region after squamous cell carcinoma. They are divided into two main categories: Hodgkin and non-Hodgkin's lymphomas. Non –Hodgkin's lymphoma (NHL) may be of B cell, T cell or both. In adults, majority of non-Hodgkin's lymphoma are of B cell origin [1]. Oral lymphomas are relatively rare lesions mimicking other pathologies such as periodontal disease, osteomyelitis and other malignancies, thus making diagnosis a challenge [2].

The soft palate is a complex sub site of the oropharynx with a variety of native tissue types that can give rise to a plethora of pathological conditions both benign and malignant [3]. Also, exposure to carcinogens such as tobacco and alcohol is common; making squamous cell carcinoma the most common malignancy of the Head and Neck. Others like inflammatory lesions, leukoplakia and erythroplakia are well known premalignant lesions [4]. There is general agreement that involvement of lymph nodes is common

with oral lymphoma [5]. However, we noted that our patient did not have cervical lymph nodes involvement.

## CASE REPORT

A 71-year-old male retired primary school teacher who was referred to the ENT Clinic of the University of Calabar Teaching Hospital from the General Outpatient Clinic for review and definitive management on account of difficulty in swallowing and slurred speech of 3-years duration. This was followed by inability to lie supine as a result of choking of one-year duration.

Dysphagia was progressive and not specific to solids. Patient manually pushes the food through with his index finger during swallowing. No odynophagia but feeling of lump in the throat and associated hot potato voice. There was no drooling of saliva. Patient was a non- smoker with no history of tobacco chewing but consumption of alcohol was sparingly. Similarly, there were no systemic symptoms of fever, night sweat,

pruritus or fatigue. No significant medical or surgical history.

General examination revealed elderly man, healthy looking, afebrile, no pallor and not dehydrated. He was not in any respiratory distress. Vital signs were essentially within normal range. Pulse 88beats/min. regular, BP 110/90mmHg, respiratory rate 22 cycles/min. and temperature 36.7oc Otorhinolaryngological examinations showed bilateral cerumen auris, moderate engorgement of the inferior turbinates, no rhinorrhoea and both nasal cavities were essentially patent. Examination of the oropharynx revealed good mouth opening, fair oral hygiene with incomplete dentition as the 1<sup>st</sup> -3<sup>rd</sup> molars of the lower jaws was absent. The tongue was mobile in all directions, the hard and soft palate apparently intact; but marked bulging and elongation of the soft palate down the base of tongue as it could not be retracted upwards.

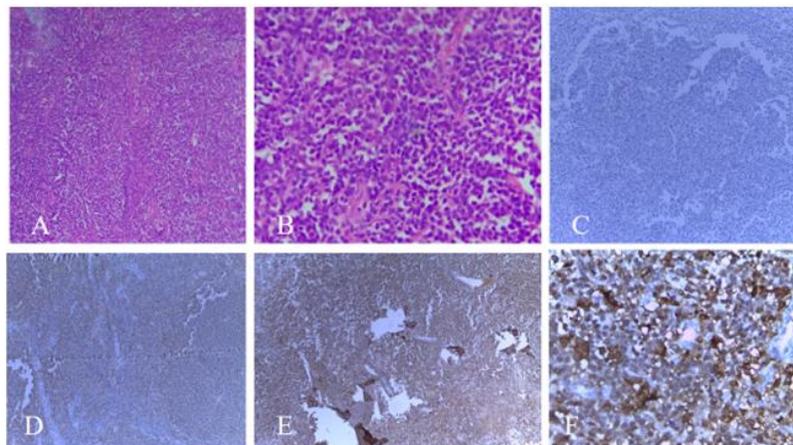
The uvula was not visible. There were no signs of ulceration, bleeding or discharge. No palpable lymph nodes in the head and neck region, or any other parts of the body. Investigations revealed normal complete hemogram, blood sugar, and liver and kidney function tests. Chest and Para nasal radiographs were essentially normal. Also, HIV screening and ECG were normal. A CT Scan could not be done because of financial constraints. However, Plain X – ray of the post nasal space revealed a pedunculated mass arising from the soft palate down the laryngeal inlet obstructing it. A provisional diagnosis of benign Oropharyngeal tumour was made. To establish the diagnosis, patient underwent elective tracheostomy under local anaesthesia in a semi sitting position, because he could not lie down supine as the tumour was obstructing the airways. This was followed by examination under anaesthesia and complete excision of the tumour (Fig. 1).



**Fig-1: A 71-year-old man with follicular lymphoma of the palate**

A: Pre-anaesthesia; B: Tracheostomized for anaesthetic access; C: Exposure of operation site showed tumour obstructing the oropharynx; D: Tumour

excised; E: Post operation; F: External surface of tumor after fixation and G: Cut surface showing a greyish white tumor.



**Fig-2: Photomicrograph showing sections of lymphoid tissue with partial effacement of the architecture that is replaced by ill - defined follicles (Fig 2A: HE x100). Follicles comprises of varying proportions of centrocytes and centroblasts (Fig 2B: HE x400). Fig: C-CD5 negativity, Fig: D- Bcl 6 positivity, Fig: (E-F) - CD10 positivity**

The histopathological report showed sections of lymphoid tissue with partial effacement of the

architecture that is replaced by ill - defined follicles and diffuse pattern in some areas. Follicles are of various

sizes and shapes and contain varying proportions of centrocytes and centroblasts. The centrocytes are small with angulated nucleoli, condensed chromatin and inconspicuous nucleoli within a scanty cytoplasm. The centroblasts are large with irregular nuclei and clotted nuclear membrane with moderate amount of cytoplasm (Fig. 2A-C). Also seen are sclerotic bands separating follicles, areas of haemorrhage, mucous glands and non-keratinized squamous epithelial cells and immunohistochemistry further confirmed the diagnosis (Fig. 2C-F). The tumour shows strong positivity for CD10 and Bcl-6 and negative for CD5. So far, patient is doing well on follow up; yet to have involved site radiotherapy (ISRT) because of financial constraints.

## DISCUSSION

Non-Hodgkin's lymphoma (NHL) belongs to a group of lymphoid neoplasms, which are diverse in manner of presentation, response to therapy and prognosis [5]. The most common site of extra nodal lymphoma in the head and neck is the palate; but NHL may involve both osseous and soft tissues. Also, it appears as non-tender, diffuse and rarely ulcerated lesion. Our index case had non-ulcerated swelling of the soft palate with intact overlying mucosa. Non-Hodgkin's lymphoma commonly affects the middle age and the elderly with a slight male preponderance [5]. Our patient is a 71-year old man. Similarly, one third of NHL may present in extra nodal sites [6]. In addition, most of NHL is hardly ever accompanied by 'B symptoms' such as weight loss, unexplained fever, or night sweats [7] as presented by our index patient. Lymphoid lesions of the palate can be classified into three main categories: primary lymphoma of the palate; infiltration of the palate as a part of disseminated disease and benign lymphoid hyperplasia (BLH) of the palate [8]. Benign lymphoid hyperplasia (BLH), is a strong differential of low grade lymphoma of the palate. However, in BLH, the cells demonstrate all stages of follicular center cell transformation as different from malignant lymphoma where the range of cells is narrow with nuclear atypia [9] Furthermore, no major clinical or histologic differences between BLH and the palatal lymphoma, hence the role of immunohistochemical study in differentiating them. WHO staging is based on 3 grades: grade 1 with zero to five centroblasts, grade 2 with six to 15 centroblasts and grade 3 with more than 15 centroblasts by field view (x 40 magnification). However, grade 3 is divided into 3A with persistence of some centrocytes and 3B with centroblasts only. Follicular lymphoma (FL) grade 1, 2 and 3A are considered as an indolent disease, while grade 3B as an aggressive lymphoma. Our patient had grade 1 tumour. Extra nodal lymphomas of B-cell origin especially low-grade types tend to remain localized for long period of time; therefore local treatment (surgery or radiotherapy) is effective at long-term control of disease [5]. In some cases involved site radiotherapy (ISRT) with radiation dose of 24–30Gy (2Gy per fraction) is the most effective treatment for low grade lymphoma of the

oropharynx; observation alone can be an option if biopsy is excisional like in our patient. However, if the margins are positive post-surgery local radiotherapy and chemotherapy (cyclophosphamide, doxorubicin, vincristine, and prednisolone) are considered [10]. Similarly, monoclonal antibodies directed against antigens or within the lymphoma and injection of interferon have been used [5]. Prognosis of follicular lymphoma (FL) is very good with early diagnosis and appropriate therapy, but delayed diagnosis may be detrimental for the patients. The index patient had a localized lesion with histological grade 1. He had complete excision of his tumour, and had been followed up for 15 months without recurrence.

## Management challenges

- 1). CT – Scan could not be done because of financial constraint, therefore the tumour extent could not be ascertained preoperatively.
- 2). Oral Endotracheal intubation could not be instituted due to inability of the patient to lie supine without being choked, hence elective tracheostomy in semi-sitting position for anesthetic access.
- 3). There was immediate post – operative velopharyngeal insufficiency, but it was successfully ameliorated with speech therapy.

## CONCLUSION

Follicular lymphoma of the soft palate though rare should be considered in the differential diagnosis of palatal lesions. Histopathological and immunohistochemical criteria are essential to differentiate it from benign lymphoid hyperplasia. Early diagnosis and treatment offer the best prognosis. The rarity of follicular lymphoma of the soft palate coupled with the above challenges made this case to be a unique one.

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## Consent

Written consent to display the patient's photographs was duly obtained from him.

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