

## CASE REPORT

# Non-Hodgkin's Lymphoma: An Unusual Presentation

S Juthika Rai<sup>1</sup>, Rakshita R Kamath<sup>2</sup>

## ABSTRACT

Non-Hodgkin's lymphoma especially the diffuse large B-cell lymphoma variety accounts for a considerable fraction of lymphomas. However, their *de novo* occurrence in the Waldeyer's ring is rare. The independent origin of this condition as a large unilateral tonsillar swelling makes it an interesting presentation in clinical practice. Here, we present such a case with the few known aspects of this condition.

**Keywords:** Diffuse large B-cell lymphoma, Non-Hodgkin's lymphoma, Tonsillar disease.

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## INTRODUCTION

Non-Hodgkin's lymphoma (NHL) of the Waldeyer's ring is a clinical rarity. Among these tumors, palatine tonsils are most frequently involved. A variety of predisposing factors, such as human immunodeficiency virus, Epstein-Barr virus, has been associated but the exact etiology remains unidentified. NHL of primary extranodal type involving the oral cavity as well as oropharynx amounts to 13% with almost 70% fraction occurring in the tonsils.<sup>1</sup> Most common high-grade rapidly growing NHL is diffuse large B-cell lymphoma (DLBCL), reaching about 80% in some of the studies.<sup>1,2</sup> Many reported cases of tonsillar NHL are bilateral and range between 2 and 4 cm sizes.<sup>2</sup> This study reports a rare case of a large independent localized unilateral extranodal NHL of the left tonsil.

## CASE DESCRIPTION

A 65-year-old male came to us with swelling in the left tonsillar fossa since 10 days, which was sudden in onset, associated with difficulty while swallowing food, and change in voice since 5 days. Ear, nose, and throat examination defined a unilateral growth with smooth bosselated surface arising from the left tonsillar fossa around 5 × 8 cm pushing anterior and posterior tonsillar pillars, inferiorly extending up to left valleculae crossing the midline. **Figure 1** shows the extent of the mass noted on the clinical examination. Indirect laryngoscopy appeared normal. No palpable salivary gland mass or metastatic lymphadenopathy was found clinically. Contrast-enhanced computerized tomography (CT) skull base to mediastinum and chest was done. The CT image, as in **Figure 2**, showed a well-defined globular soft tissue density along the left posterior pharyngeal wall extending up to soft palate. No significant cervical lymphadenopathy was noted and hence a benign etiology was suspected.

The patient underwent tonsillectomy and the surgical specimens of both tonsils were formalin fixed and paraffin embedded for histological examination. Macroscopically, there was a 9 cm × 5.5 cm × 2 cm mass with grey-white to grey-brown soft tissue bits. Microscopy, as shown in **Figure 3**, depicted complete effacement of tonsillar architecture replaced by tumor tissue composed of sheets of monomorphic medium to large round to mildly indented cells with vesicular nuclei and variable number of nucleoli. Atypical mitotic figures and numerous

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karyorrhectic debris are seen dispersed in tumor tissue. Also seen are thin-walled vessels amid tumor cells. Immunohistochemistry with CD20, CD45, CD10, and Ki-67 confirmed diagnosis of diffuse large B-cell Non-Hodgkin's type of lymphoma. Postoperatively oncology reference was given. Chemoradiation was planned for the patient subsequently.

## DISCUSSION

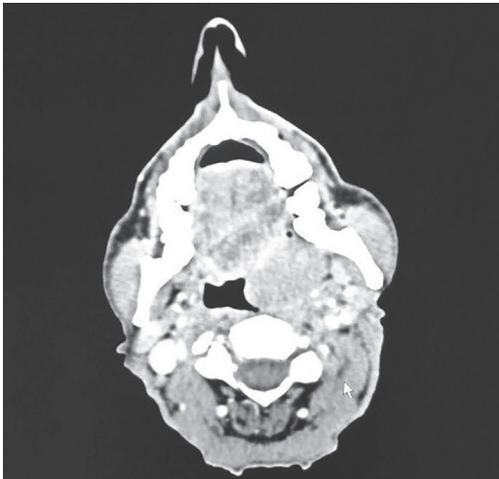
Malignant lymphomas in majority of the cases arise from lymph nodes. However, 24–48% of them arise in extranodal sites.<sup>1</sup> Guevara-Canales et al. reviewed 215 articles to conclude that Waldeyer's ring along with gingiva was the most commonly affected site with palatine tonsil being the most common.<sup>2</sup>

Primary tonsillar lymphoma occurs in <1% of all head of neck malignancies.<sup>3</sup> The lymphoma has peaks in the 6th and 7th decades of life in published studies<sup>4</sup> and the incidence is male predominant.<sup>2</sup> The nonspecific clinical signs and symptoms include but are not limited to fullness in the throat, sore throat, pain or difficulty in swallowing, earache, cervical lymphadenopathy, swelling in tonsils, or snoring. Advanced disease may also show fever, weight loss, and night sweats, albeit rare. Mild-to-moderate tonsillar enlargement on clinical presentation can easily be misdiagnosed for tonsillitis when it is the only presenting feature in this cohort of cases.<sup>5</sup>

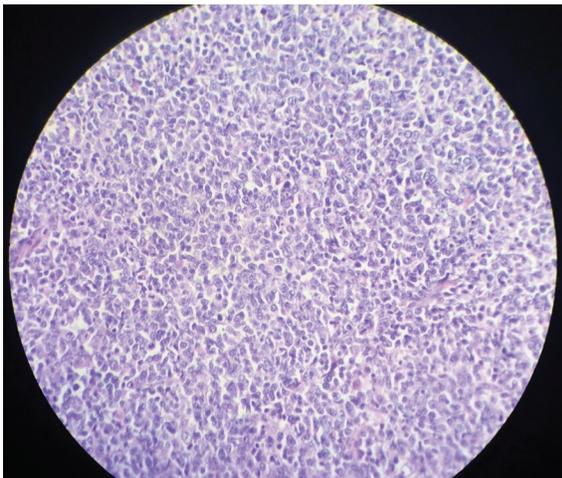
To the best of our knowledge, a rare entity such as DLBCL of unilateral tonsil of 8 cm size has not been previously reported. A massive unilateral enlargement, as seen in our patient, requires a high degree of suspicion toward malignancy and hence histological



**Fig. 1:** Clinical examination showed 5 × 8 cm left tonsillar mass pushing the left anterior tonsillar pillar



**Fig. 2:** CT scan showing left well-defined globular soft tissue density along the left posterior pharyngeal wall



**Fig. 3:** High-power 200× microscopic image showing complete replacement of tonsillar architecture by tumor tissue. Sheets of monomorphic large cells were seen having vesicular nuclei and nucleoli with atypical mitotic figures

examination becomes of utmost importance to differentiate inflammatory from malignant enlargements.

Most tonsillar lymphomas published are that of B-cell type, and the most common histologic type has been reported to be DLBCL19. DLBCL is an aggressive malignancy with a characteristic histological feature of diffuse large lymphocytic growth with nuclear size equal to or more than a normal macrophage nuclear size or over double the normal lymphocytic size.<sup>6</sup>

It is recently established that DLBCL may arise due to a transformation from an underlying low-grade lymphoproliferative condition.<sup>7</sup> Based on histological study, differentials for DLBCL can include undifferentiated carcinomas, sarcomas, plasmacytomas, and malignant melanomas. Therefore, immunohistochemistry is crucial for a pinpointed singular diagnosis. These tumor cells react positively to CD20, PAX5, CD10, BCL6, and BCL2 with some being CD30 positive.<sup>8</sup> While they are strongly negative for epithelial markers (such as cytokeratins), CD3 (T-cell lymphoma), CD138 (plasmacytoma), desmin (rhabdomyosarcoma), myeloperoxidase (granulocytic origin), and S100 (melanoma and neurogenic sarcoma), and hence in case of suspicion, these tests must be done whenever possible. Therapeutics of the disease that may span from chemotherapy alone/radiotherapy alone to combination of both therapies is mostly preferred. A number of prognostic factors have been given with none proved on evidence-based studies due to the gross underreporting of this independent entity. However, presenting masses of size above 7 cm had a significantly poorer outcome as compared with those <7 cm in many studies.<sup>9–12</sup> A 5-year survival rate of 65–85% for patients with early-stage presentation has been reported.<sup>12</sup> Another important prognostic factor considered for primary extranodal lymphomas is their location.<sup>13</sup> With these in mind, the poor prognosis of this case was explained to the patient and prompt treatment with chemoradiation was planned.

## CONCLUSION

Independent origin of localized DLBCL presenting as an enlarged ipsilateral tonsillar mass is a rare entity. Thorough workup with histological and immunohistochemical studies is necessary for appropriate diagnosis. Large bulky size at presentation may denote poor prognosis. A combined treatment consisting of chemotherapy and radiotherapy leads to a satisfactory outcome in patients with this uncommon neoplasm.

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