

Laryngeal NK/T-cell Lymphoma: A Report of a Rare Case with Unusual Location

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ABSTRACT

Extranodal NK/T-cell lymphoma (ENKL) is a rare type of non-Hodgkin lymphoma, and involvement of the larynx is an even rarer entity. We present a case of 23-year-old woman presenting with several weeks of respiratory distress and hoarseness. Bronchoscopy showed severely stenosed glottis and subglottis with multiple 1–2 mm pink-colored nodules over the false vocal cords and vallecula. Histology revealed unremarkable squamous mucosa with underlying diffuse proliferation of atypical lymphoid cells in a necrotic background. The atypical lymphoid cells were positive for CD2, CD3, CD56, CD30, and EBER with high Ki-67 proliferative activity. The overall findings were consistent with extranodal NK/T cell lymphoma of the larynx. The diagnosis of ENKL can be challenging due to the non-specific clinical presentation and relatively unusual localization. In the setting of inflammation of mucosa, neoplastic NK/T cells may be masked by secondary reactive inflammatory cells or intense necrosis. Therefore, a diagnosis of ENKL is frequently dependent on a series of immunohistochemical and molecular studies.

Key words: Angiocentric, extranodal NK/T-cell lymphoma, larynx, lymphoma, non-hodgkin lymphomas

INTRODUCTION

Extranodal NK/T-cell lymphoma (ENKL) is an aggressive angiocentric and angiodestructive tumor associated with prominent necrosis. ENKL affects people worldwide; however, it is more prevalent in Asia, Central, and South America.^[1] ENKL predominantly involves extranodal sites, particularly the upper aerodigestive tract.^[1,2] Epstein–Barr virus (EBV) is found in nearly 100% of the cases.^[2] Common non-Hodgkin lymphomas of the larynx are diffuse large B-cell lymphoma, marginal zone B-cell lymphoma, and extramedullary plasmacytoma.^[3] Localization of ENKL to the larynx is very rare, and to our knowledge, only 31 cases of laryngeal NK/T-cell lymphoma have been reported in the literature.^[4] Due to the uncommon location, vague symptoms, and non-specific gross morphology, the

diagnosis of ENKL can be difficult. Herein, we present a case of ENKL involving the subglottic area.

CASE REPORT

The patient is a 23-year-old woman who presented with several weeks of respiratory distress and hoarseness. Flexible bronchoscopy showed swollen vocal cords and arytenoids with severely stenosed glottis and subglottis and heaped up irregular, easily friable tissue. In addition, it showed multiple 1–2 mm pink-colored nodules over the false vocal cords and vallecula. Rigid bronchoscopy was performed with excision of some of the glottic and subglottic tissue. Microscopic examination of the specimen revealed unremarkable squamous mucosa with underlying diffuse proliferation of atypical lymphoid cells with angiocentric and

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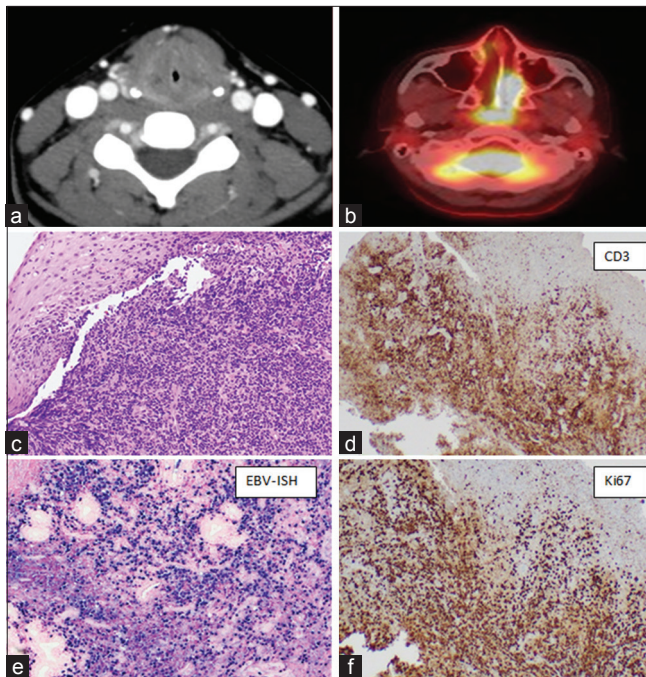


Figure 1: Computed tomography scan shows subglottic stenosis; PET scan shows the left nasal mucosa thickening; microscopic section of subglottic biopsy demonstrates squamous mucosa with atypical lymphocytic tissue (c); immunostains show that atypical lymphocytes are positive for CD2, CD3 (d), CD7, CD30, CD56, and Epstein-Barr virus-in situ hybridization (e); Ki-67 highlights high proliferative index; and (f). Immunoprofile confirms the diagnosis

angiodestructive growth pattern in a necrotic background. The atypical lymphoid cells were positive for CD2, CD3, CD56, CD30, and EBV with a high Ki-67 proliferative index. The overall findings were consistent with extranodal NK/T cell lymphoma [Figure 1].

DISCUSSION

ENKL, nasal-type is aggressive lymphoma arising from NK-cell or T-cell lineage. ENKL almost always presents as an extranodal tumor, but lymph node involvement can occur secondarily. These tumors have predilection for involvement of aerodigestive tract (nasal cavity, nasopharynx, paranasal sinuses, and palate) but rarely could occur at other sites including gastrointestinal tract, skin, and testis.^[5-9] The pathogenesis of these tumors is unknown yet, but their strong association with EBV infection is suggestive of an oncogenic role of the virus.^[10] ENKL of the nasopharynx usually presents with nasal mass and obstruction. Furthermore, due to the angiodestructive nature of the disease, they can develop extensive destruction of the midfacial structures and hard palate perforation; however, depending on the site of involvement, they can present differently.^[11]

Lymphomas of the larynx are very uncommon and comprise only 1% of all laryngeal neoplasm. They are mostly non-Hodgkin's B-cell lymphoma, and the primary NK/T-cell lymphoma of the larynx is extremely rare.^[12,13] Laryngeal lymphomas usually present as non-ulcerated polypoid masses, with most cases showing involvement of the epiglottis and aryepiglottic folds; however, they can involve other locations such as subglottis and vocal cords as well.^[14,15] Diagnosis of these tumors is usually challenging because of their non-specific clinical presentation and gross morphology. Furthermore, the histopathologic examination can be difficult due to reactive inflammation and intensive necrosis that can mask the tumor and make the diagnosis very challenging in the small biopsy specimen. Therefore, knowledge about the rare sites of involvement and application of the ancillary test is needed for an accurate diagnosis.

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