Lymphoma of larynx presented with hoarseness: Case report

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Abstract
Primary non-Hodgkin lymphoma of the larynx is rare, accounting for less than 1% of all laryngeal neoplasms. Fewer than 100 cases have been reported in the literature and to our knowledge this may be the first reported case of laryngeal non Hodgkin lymphoma presented with hoarseness, aortic aneurysm, hepatitis B and skin rashes.

Keywords: Lymphoma, Larynx, Subglottic mass, Hepatitis B.


Introduction
Primary non-Hodgkin lymphoma of the larynx is rare, accounting for less than 1% of all laryngeal neoplasms. Fewer than 100 cases have been reported in the literature (1).

Case report
A 26-year-old man, with a known case of hepatitis B, was referred to our hospital from the ENT clinic after a severe respiratory distress and hoarseness, which had occurred after an episode of upper respiratory tract infection (URTI). The condition was started 2 weeks earlier. His symptoms were started and exacerbated following URTI. He was treated with Cefixime, Theophylline and Salbutamule (inhaler) but the symptoms did not resolve. He did not have dysphagia or odynophagia, nor did carry any history of asthma, allergy, smoking or illegal substance abuse. His father had died of end stage stomach cancer about a year ago and his brother had vasculitis (who is under the treatment with corticosteroids).

Physical examination revealed fever (38.5°C). Examination of his mouth and oropharynx showed erythema and tonsillar hypertrophy without exudates in his oropharynx. He had an obvious inspiratory and expiratory stridor and overt respiratory distress. Auscultation of lung revealed diffused wheezing in his lungs with a 3/6 systolic murmur in his left sternal border and pulmar area. There was no evidence of peripheral edema. Indirect laryngoscopic examination showed papillomatose lesion in the 1/3 posterior portion of true vocal cords. There were maculopopular rashes on his chest wall, upper back, neck, face and extremities (Fig 1).

Chest x-ray showed an increase in bronchovascular markings in middle and upper lobes of right lung. Lateral head and neck radiography revealed a mass in his subglottic area (Fig. 2-4). Chest CT–scan showed tubular bronchectatic changes at right lobe associated with pulmonary inflammation. Fibrotic band was seen at basal portion of both lungs with no pleural effusion. There was an aneurismal dilatation of aorta with maximum diameter of 50×48 mm in the...
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ascending part and about 49×45 mm in the descending part. The ultrasound echocardiography confirmed the aortic aneurysm. Venereal disease research laboratory (VDRL), fluorescent treponemal antibody-absorption test (FT-ABS), antinuclear antibody (ANA), Cytoplasmic antineutrophil cytoplasmic antibodies (C-ANCA) and Perinuclear Anti-Neutrophil Cytoplasmic Antibodies (P-ANCA) were negative and angiotensin-converting enzyme (ACE) levels fallen within normal range. His rheumatoid factor was 3+ and C reactive protein 1+. His ESR level in the first hour was 54 mm/hr. Emergency tracheotomy and mass debulking was done for emergency air way management. Direct laryngoscopy showed a 1.5 cm mass in subglottic area. Specimens were taken for culture and pathologic diagnosis. The throat cultures were negative for Mycobacterium tuberculosis and fungus. The specimen also contained multiple pieces of creamy-brownish soft tissue. The microscopic evaluation showed tissue composed of large cells with hyperchromatic round to ovaloid nuclei, prominent nuclei and irregular border and eosinophilic cytoplasm arranged as sheets. The immunohistochemistry report was positive for leukocyte common antigen (LCA), antigen KI-67 (60% of tumoral nuclei) and negative for CD3, CD20, CD43, CD30. The pathologic diagnosis was diffuse large cell lymphoma (High grade) and immunohistochemistry test for CD79 was positive which determined the B cell type. He had no symptom afterward.

**Discussion**

Primary non-Hodgkin’s lymphoma (NHL) of the larynx is rare. Fewer than 100 cases were reported in the literature. More commonly, the larynx is involved secondarily after the lymphoma of regional lymph nodes. Laryngeal lymphomas are usually sub mucosal masses, without mucosal ulceration and are most commonly found in
supraglottis especially in the aryepiglottic folds. Invasion of the subglottic larynx and trachea by lymphoma is an uncommon problem which can cause severe airway obstruction and requires multidisciplinary approach (2).

Our patient presented with an ulcerative mass of the subglottic region that is an unusual primary site for laryngeal lymphoma. Hence the site of the lesion cannot definitely rule out the diagnosis. The relationship between the aortic aneurism, lymphoma and hepatitis (in the same patient) has not been shown. There may be an increased risk of aortic aneurism in some vasculitis disorders (3). Some studies showed higher incidence of hepatitis B virus in B-cell non-Hodgkin lymphoma (NHL) patients (4,5), but we did not find any evidence of vasculitis in our patient despite his positive family history (7). Also there is an increased risk of lymphoma in the hepatitis patients (4,5), but there has not been a report of B cell lymphoma, hepatitis B, and aortic aneurism in the same patient in the literature. Opposed to the common treatment regimen in such patients, our patient responded well to a single chemotherapic regimen and did not need another laryngoscopic intervention.

**Conclusion**

There are not any characteristic findings to distinguish laryngeal lymphoma from other neoplasms without pathologic study; but when there are some systemic findings in a patient that physician should consider them as rare conditions of differential diagnosis. However after establishing the diagnosis, treatment of these patients must be started with standard regimens, good prognosis and complete remission of the symptoms (7). The only specific treatment we performed on our patient was tracheotomy due to respiratory difficulty in the first presentation of the patient, which improved after chemotherapy. The patient was symptom free after one year of follow up.

**References**