

Primary Non Hodgkin's Lymphoma of Glottis: A Diagnostic Dilemma

Shashikant Anil Pol¹, Aditi Mahajan^{2*}, Reetu Kundu³, Surinder K Singhal⁴ and Nitin Gupta⁵

¹Assistant Professor, Department of ENT, JIPMER, Puducherry, India

²Senior Resident, Department of ENT, GMCH, Chandigarh, India

³Assistant Professor, Department of Pathology, GMCH, Chandigarh, India

⁴Associate professor, Head of Department ENT, GMCH, Chandigarh, India

⁵Associate professor, Department of ENT, GMCH, Chandigarh, India

***Corresponding Author:** Aditi Mahajan, Senior Resident, Department of ENT, GMCH, Chandigarh, India.

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Abstract

Primary Non-Hodgkin's 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. Early symptoms are nonspecific and subtle, so confirmation of the diagnosis is difficult thereby leading to a delay in management. We hereby present a case of a 30 year old male with Non Hodgkin's lymphoma of larynx who presented with hoarseness as the only complaint. In this case, intra operative assessment and histological interpretation was critical, and formed the basis of correct management in the form of radiotherapy.

Keywords: Non Hodgkin Lymphoma; Microlaryngeal Surgery; PET Scan; IMRT

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]. Sometimes rapid enlargement of submucosal masses can result in relatively acute airway compromise. Initial management should include airway stabilization and a biopsy specimen. Various combinations of intravenous steroids, chemotherapy and external beam radiation therapy have been used successfully in lesions compromising the airway [2]. In laryngeal lymphoma, radiation therapy may be the only therapeutic modality necessary to achieve an excellent prognosis [3,4].

Case Report

A 30 year old non smoker male patient presented with hoarseness since 2 years, which was insidious onset, gradually progressive and persistent in nature non responding to medical and speech therapy. There was no history of voice abuse and any respiratory difficulty.

On indirect laryngoscopy, a cystic swelling was noticed which was involving the right true vocal cord from anterior one third to vocal process of arytenoid posteriorly. Both vocal cords were mobile and glottic chink was adequate. This finding was confirmed

with ninety degree rigid endoscope and a diagnosis of benign cystic lesion of larynx was made (Figure 1). Patient was planned for excision of cystic lesion under general anaesthesia. CT scan of neck was not performed considering the benign nature of the lesion.

Figure 1: Endoscopic image of cystic lesion.

After all investigations for preanaesthetic check up, patient's written and informed consent was taken for microlaryngeal surgery. Patient was taken under general anaesthesia with orotracheal intubation. Microscopic findings showed a single cystic swelling which was extending anteriorly from anterior one third of right true vocal cord to the vocal process of right arytenoid posteriorly. Superiorly it was extending from free upper edge of right true vocal

cord and inferiorly going till subglottis. Medially it was involving almost half of the glottis and laterally it was attached to the medial border of right true vocal cord not infiltrating it. Whole cystic lesion was excised with cold instruments and sent for histopathological examination. Patient was extubated without any difficulty.

The histopathology report showed sheets of atypical lymphoid cells beneath the stratified squamous mucosal epithelium. On immunohistochemistry the atypical lymphoid cells are positive for CD 20 and are negative for CD 3 and chromogranin. So final report was Non Hodgkin's lymphoma low grade B cell type (Figure 2).

Figure 2: Endoscopic image after excision.

PET scan was done and it showed FDG avid soft tissue thickening in bilateral true vocal cords. No PET evidence of abnormal FDG avid hypermetabolism elsewhere in the body.

Patient underwent local radical radiotherapy to neck 40 Gy/20 fractions by IMRT. Patient was followed till four years with no disease clinically (Figure 3) and on repeat PET scan.

Figure 3: HPE shows sheets of atypical lymphoid cells beneath the stratified squamous mucosal epithelium (H&E, x100). Inset shows neoplastic lymphoid cells with membranous positivity for CD20 immunostain (IHC, x400).

Discussion

Primary hematologic laryngeal tumors account for less than 1% of laryngeal tumors [5,6]. Lymphoma of larynx are mainly NHL and are most commonly located in the supraglottic region, as this area of the larynx contains follicular lymphoid tissue.

Non-Hodgkin lymphoma (NHL) is the second most common primary hemopoietic tumor after plasmacytoma. Fewer than 100 reported cases of primary NHL of larynx are reported in the general literature [7,8]. NHL of the head and neck usually arises in the extranodal lymphatic system of Waldeyer's ring. Involvement of extranodal extralymphatic sites is less common, and involve the sinonasal tract, salivary glands, thyroid and orbit more often than the larynx. Laryngeal NHL is seen in patients of varying age ranging from 4 to 81 years, although the mean age is 58 years and occurrence in children is extremely rare [7,9]. Our patient was 30 years of age. Sex-ratio has also been reported to be variable in different series [11,12]. Symptoms of NHL are site-specific and thus have manifestations similar to those produced by carcinomas in the same location. These include hoarseness, dysphagia, and cervical lymphadenopathy [14-16]. Hoarseness was the only complaint present in our patient. Systemic symptomatology is unusual, since laryngeal lymphomas tend to remain localized for prolonged periods.

Histologically, primary laryngeal lymphoma originate more commonly from B-cell, though some T-cell and NK- cell lymphomas may be found. The salient feature of lymphomas is that these are considered as clonal proliferation of lymphocytes arrested at different stages of differentiation. Immunohistochemistry with various antibodies is essential to reach the specific lineage and the developmental stage. A panel of markers is used, which includes leukocyte common antigen (LCA), B-cell markers (CD20 and CD79a), T-cell markers (CD3 and CD5) and other markers like CD23, bcl-2 and CD10 [13].

PET-CT, in particular, is finding an important place for radiological staging in laryngeal lymphoma, both for low-grade and high-grade ones [17]. The main modalities of treatment is radiotherapy alone or in combination with chemotherapy [18]. Combined chemoradiotherapy, seems to be the preferred modality of treatment, especially for high-grade lymphomas, and provides an excellent outcome [19,20].

Conclusion

- Clinically Primary Non Hodgkin's lymphoma of larynx is very difficult to differentiate from benign lesions of larynx.

- It can present with variable symptoms and signs depending upon the site involved.
- The intraoperative findings, detailed histopathological examination and immunohistochemistry forms the basis of diagnosis and thus successful treatment with fairly good prognosis.

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