Laryngeal Lymphoma in a Child – Case Report and Review of Literature
Anupriya Ayyaswamy¹, Prasanna Kumar Saravanam¹, Sneha Latha³, Sandhya Sundaram⁴

Abstract
Introduction:
Head and neck is the second most common region for lymphomas. Extranodal lymphomas of the larynx are rare in the pediatric population. Non Hodgkin Lymphoma (NHL) of the larynx is common in the supraglottic region as its rich in lymphoid tissue. They may present with dysphagia, dysphonia, snoring and progressive respiratory distress. Early visualization of the larynx is essential in such cases for appropriate diagnosis to improve the survival rates.

Case Report:
We present a case of 9 year old boy who presented with a change in voice, snoring and feeding difficulties for one year. Video laryngoscopy revealed globular mass arising from the epiglottis. He underwent excision biopsy and by immunohistochemistry was diagnosed to have diffuse large B cell lymphoma. He was treated with chemotherapy and the child is clinically well in the follow-up, 1 year after the completion of therapy.

Conclusions:
Although primary lymphomas of the larynx in children are rare, a high index of clinical suspicion is warranted to avoid diagnostic delays to initiate appropriate management to have better outcomes.

Keywords:
Child, Dysphonia, Lymphoma, Larynx, Snoring.

Received date: 10 May 2021
Accepted date: 31 May 2022

*Please cite this article; Ayyaswamy A, Saravanam PK, Latha S, Sundaram S. Laryngeal Lymphoma in a Child – Case Report and Review of Literature. Iran J Otorhinolaryngol. 2022:34(6):337-341. Doi:10.22038/IJORL.2022.57663.2984

¹Department of ENT, Head and Neck Surgery, Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai
²Division of Pediatric Hemato Oncology, Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai.
³Department of Pathology, Sri Ramachandra Institute of Higher Education and Research, Porur, Chennai.
*Corresponding Author:
Sri Ramachandra Institute of Higher Education and Research. Porur, Chennai 600 116.
E-mail: drmslatha@yahoo.com
Introduction

Primary extranodal lymphoma of the head and neck account for 2.5% of all lymphomas (1). Laryngeal lymphomas are very rare, accounting for 1% of laryngeal malignancies and are mostly reported in the adult population (2). Only very few cases of epiglottis as the primary site of laryngeal lymphoma are reported in the pediatric age group. We report a case of 9 year old boy who was diagnosed with primary B cell lymphoma of the epiglottis, treated with surgery and chemotherapy and is currently well on follow-up.

Case Report

A 9 year old boy presented with complaints of difficulty in swallowing, change in voice and snoring for one year associated with a weight loss of 4 kgs during this period. The child did not have any complaints of breathing difficulty, fever or night sweats. On examination the oral cavity was normal. There was no cervical lymphadenopathy. Indirect Laryngoscopy showed a single globular mass seen occupying the base of the tongue and epiglottis. Bilateral vocal cords could not be visualized. Video laryngoscopy done showed a single globular mass with irregular surface arising from epiglottis extending till vallecula and base of tongue (Figure 1).

Fibre Optic Laryngoscopy showed bilateral vocal cords were mobile, pyriform fossa free on both sides. Contrast-enhanced computed tomography (CECT) neck was done to look for the extent of the lesion which showed hyperdense lesion measuring 2.8 x3.7 x3.5cms involving epiglottis extending into vallecula obstructing the airway (Figure 2).

The patient underwent coblation assisted complete excision of the growth by ENT surgeons. Histopathological examination showed fragment tissues lined by stratified squamous epithelium with underlying subepithelium showing sheets of a monomorphic population of lymphoid cells infiltrating into the mucosal and adjacent cartilaginous tissue (Figure 3).

By immunohistochemistry, the cells were diffusely positive for CD20, CD45 (fig 4a and...
Laryngeal Lymphoma in Children

4b) which was confirmative of B cell lymphoma. As part of the metastatic evaluation, whole-body PET CT and bone marrow biopsy done were normal. CSF analysis done to rule out CNS involvement was also normal.

He was risk stratified as Group B disease and treated with chemotherapy as per LMP 96 protocol. He received COP (Cyclophosphamide, vincristine, prednisone), COPADM 1 and 2, (cyclophosphamide, vincristine, prednisone, doxorubicin, methotrexate) CYM1 & 2 (cytarabine, methotrexate) and intrathecal methotrexate and cytarabine.

PET CT and video laryngoscopy done after completing chemo-therapy showed no evidence of recurrent or residual tumor. He was on regular follow up with pediatric oncology and ENT. A repeat video laryngoscopy done on follow up after 1 year of diagnosis showed normal findings (Figure 5).

Discussion

Head and neck is the second most common site for primary extranodal non-Hodgkin lymphoma, followed by gastrointestinal tract (1). NHL of the head and neck usually arises from the waldeyer’s ring. The other less commonly involved extranodal sites are sinonasal tract, salivary glands, thyroid and orbit. Laryngeal carcinomas account for <0.1% of all head and neck malignancies of childhood (3). Burkitt lymphoma, lymphoblastic lymphoma, diffuse large B cell lymphoma and anaplastic large cell lymphoma are the commoner NHL noted in children and adolescent age groups. Rhabdomyosarcoma and squamous cell carcinomas are the most common laryngeal tumors noted in pediatric patients (4). NHL of the larynx is very rare and B cell phenotype is more predominant accounting for more than 70% of cases (5). Among the NHL subtypes, large B cell Lymphoma and mucosa associated lymphoid tissue type marginal zone lymphomas are the most common primary laryngeal neoplasms (6).

In laryngeal neoplasms, the most commonly involved site is the supraglottic region as its rich in follicular lymphoid tissue (7). A review of primary laryngeal lymphomas from 1996 -2008 cites an incidence of 47% in the supraglottic region, 25% in the glottic area and rest in subglottic or transglottic regions (8).

Symptoms of laryngeal lymphoma include dysphonia, dysphagia, hoarseness of voice, cervical lymphadenopathy, cough, dyspnoea and occasionally systemic features like fever and weight loss might be present (Table 1) (9).
Table 1: The pediatric cases with laryngeal NHL is summarised below

| Study                  | Site of Lymphoma | Age/Gender | Clinical Features                  | Diagnosis                                      | Management                                      | Follow-Up                               |
|------------------------|------------------|------------|------------------------------------|-----------------------------------------------|------------------------------------------------|------------------------------------------|
| Cohen et al 1987       | Supraglottis     | 4 years, 7| Croup / Laryngitis - 1 month       | Malignant small cell neoplasm                  | Chemotherapy and radiation                       | 2 years free of disease                  |
|                        | Supra-glottis and Sub-glottis | 9 years 3 months/F | Difficulty in breathing             | Malignant NHL of Larynx                        | Chemotherapy and radiation                       | 15 years (2 years free of disease)      |
| Palenzuela et al 2002  | Supraglottis, glottis, subglottis | 15/M       | progressive respiratory distress, profuse night sweats | EBV related B cell lymphoma                    | chemotherapy radiotherapy, tumor debulking by laser | died during treatment due to lung infection |
| Rodriguez H et al 2014 | Epiglottic invasion and Glottic narrowing | 8 years/ M | Dysphony and progressive respiratory failure x 3 years | Lymphoblastic T cell Lymphoma                  | Chemotherapy                                   | Child died 16 months after diagnosis due to Septic Shock during 2nd line chemotherapy |
| Amanda Martin et al 2017 | Supraglottis | 14/F       | Dysphagia x few months Weight loss – 14lbs | Mature B cell Lymphoma                         | Surgery and chemotherapy                         | 1 month free of disease                  |
| Paloma et al 2019      | Right pyriform fossa | 13/M       | Rapidly enlarging right neck mass  | Diffuse large B Cell Lymphoma                  | Surgery and chemotherapy                         | Not available                           |

Laryngeal lymphomas remain localized for a longer duration without progression of symptoms, and a low index of suspicion in children leads to diagnostic delays. The symptoms are generally attributed to prepubertal voice changes or respiratory infections. For a child with laryngeal mass, the differential diagnosis to be considered are cystic lesions, recurrent respiratory papillomatosis, and laryngeal malignancies. In the presence of a submucosal mass centered in the supraglottis in imaging studies, NHL should be considered if there is a superior extension to the oropharynx or nasopharynx (10).

The biopsy of the lesion is essential to determine the pathology and guide treatment options. Chemotherapy is the treatment of choice. Diffuse Large B Cell Lymphoma is treated like Burkitt lymphoma. Surgery is recommended in cases of extensive airway compromise (11). Though children have more aggressive forms of non-Hodgkin’s lymphoma, with adequate and appropriate treatment five-year survival rate is high up to 5 years in 90% of children (12).

Conclusion

Though laryngeal neoplasms are rare in children, symptoms like voice change, dysphagia, and dyspnoea need extensive evaluation with a direct laryngoscope to rule out laryngeal masses. As the treatment outcomes are better with chemotherapy, early diagnosis is essential in such cases.

References

1. A. A. Ezzat, E. M. Ibrahim, A. N. El Weshi, Y M Khafaga, M AlJurf, J M Martin et al. Localized non-Hodgkin’s lymphoma of Waldeyer’s ring: clinical features, management, and prognosis of 130 adult patients. Head and Neck.2001;23:547–58.
2. J. B. Epstein, J.D. Epstein, N.D. Le, M.Gorsky. Characteristics of oral and paroraal malignant lymphoma: a population based review of 361 cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2001; 92:519–25.
3. McGuirt JWF, Little JP. Laryngeal cancer in children and adolescents. Otolaryngol Clin North Am. 1997;30:207–14.
4. Manish J, Pankaj C, Prathamesh P, Devendra C, D’Cruz Anil PG. Carcinoma larynx in children. Int J Head Neck Surg 2010;1:49-51.
Laryngeal Lymphoma in Children

5. Kato S, Sakura M, Takooda S, Sakurai M, Izumo T. Primary non-Hodgkin’s lymphoma of the larynx. J Laryngol Otol 1997; 111:571–74.
6. Horny HP, Kaiserling E. Involvement of the larynx by hemopoietic neoplasms. An investigation of autopsy cases and review of the literature. Pathol Res Pract 1995; 191:130–38.
7. Horny HP, Ferlito A, Carbone A. Laryngeal lymphoma derived from mucosa-associated lymphoid tissue. Ann Otol Rhinol Laryngol 1996; 105: 577–83.
8. Konstantinos M, John G, John C, Ioannis K, Victor V, Angelos N. Primary Laryngeal Lymphoma: Report Of 3 Cases And Review Of The Literature. Head Neck.2010; 32: 541-9.
9. J. X. Salazar Guilarte, M. Sancho Mestre, and J. R. Gras Albert. Laryngeal manifestation of B-cell non-Hodgkin lymphoma. Acta Otorrinolaringol Esp. 2012;63:485–87.
10. Ann D. King, Edmund H.Y. Yuen, Kenny I.K. Lei, Anil T. Ahuja, Andrew van Hasselt. Non-Hodgkin Lymphoma of the Larynx: CT and MR Imaging Findings. AJNR Am J Neuroradiol 2004; 25:12-15.
11. V. Paleri, F.W. Stafford, and M. S. Sammut. Laser debulking in malignant upper airway obstruction. Head Neck.2005; 27:296–301.
12. Kelly K, Brukhardt B, Bollard C. Malignant lymphomas in childhood. In: Hoffman R, Benz EJ, Silberstein LE, Heslop HE, Weitz JI, Anastasi J, Salama M, eds. Hematology: Basic Principles and Practice. 7th ed. Philadelphia, PA: Elsevier; 2018: 1330-42.