

Case Report

Carcinoma larynx in a 10-year-old girl: a rare clinical entity

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Abstract:

Childhood carcinoma larynx is a very rare clinical entity. A girl of 10 year-old presented with persistent and progressive sore throat with dysphagia, change of voice and respiratory distress with stridor which needed tracheostomy and nasogastric tube feeding. Indirect laryngoscopy revealed exophytic growth involving the epiglottis and ary-epiglottic folds, restricted movement of hemilarynx and compromised airway. Biopsies were positive for squamous cell carcinoma. The malnourished cachectic poor patient was referred for receiving radiotherapy. A high index of suspicion is necessary to make the diagnosis since consequences of late diagnosis are very serious. This report underscores the need for prompt evaluation of children with persistent and progressive sore throat with dysphagia.

Key word: Childhood carcinoma larynx, laryngeal carcinoma.

Introduction:

The first documented case of laryngeal cancer in a child was reported by Rehn in 1868 in a 3 year-old child¹. For more than a century thereafter, only 54 cases of laryngeal carcinoma have been reported in children 15 years of age or younger². Only, before this case is being reported, 22 cases in patients 10 year- old or younger and 63 cases in patients 15 year-old or younger have been reported up to date since 1868²⁻¹².

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Nevertheless, physicians must be aware that a small percentage of cases do occur in the pediatric age group since case reports in patients less than 15 years of age, laryngeal cancers accounts for less than 0.1% of all head and neck malignancies^{13, 14, 15}. In a survey of 98 cases of carcinoma of the larynx in patients less than 20 years of age, have about 20 cases in the 12 and under age group³. However, the incidence of female patients is higher in childhood (40% of childhood cases vs. less than 10% of adult cases)⁵, and risk factors, except previous irradiation of papilloma, papillomatosis, malnutrition, are rare. Adolescent carcinomas also appear to be linked to immunologic and genetic factors rather than more common risk factors such as tobacco use, previous radiation, and chemical carcinogens found in adult patients^{16, 17}. Malignant degeneration can take place in juvenile papillomatosis which was found by a study to be more common in children who had had radiotherapy

for papillomatosis; consequently the practice of irradiating these children has been abandoned³. Among the other reported risk factors for laryngeal carcinoma are papillomatosis of the larynx, laryngopharyngeal reflux, human immunodeficiency virus (HIV) infection, immunosuppressive therapy, exposure to drug use during pregnancy, both active and passive smoking, exposure to certain chemicals (e.g. asbestos), alcohol use, poor oral hygiene, and a family history of cancer;¹⁸ so, attempt for HPV viral subtyping, search for p16 and 15:19 translocation with not neglecting the HIV should be made for aetiopathogenesis. Younger patients tend to have a long history of progressive airway obstruction, dysphagia, or dysphonia; but lesions are not recognized until an otolaryngologist is consulted for a fiberoptic airway examination and a delay in diagnosis attributed to the erroneous attribution of some common paediatric respiratory and voice problems and difficult paediatric examination makes the problem more complicated. The scarcity of cases, attempt to preserve anatomy and function of larynx and for avoidance of complications impedes establishment of treatment protocols in children. All authors agree that a primary work up should consist of detailed radiological imaging endoscopy with biopsy, and possible tracheostomy for airway protection and, thereafter, definitive management of squamous cell carcinoma in pediatric patients has varied based on the individual circumstance of the patient^{19, 20}. Prognosis of childhood laryngeal cancer is unclear since the reported survival rates are not tumor stage related.

Case report:

A 10-year-old malnourished girl had attended in the Deptt. of ENT – Head & Neck Surgery, Sher-e-Bangla Medical College Hospital, Barisal, with sore throat, dysphagia, change

of voice and respiratory distress. The sore throat was persistent and progressive in nature for about two months, the dysphagia was also persistent and progressive over one month, progressive hoarseness of voice over two weeks and lastly the respiratory distress over one week associated with stridor for 3 days which necessitated tracheostomy and nasogastric tube feeding.



Figure 1: A 10-year-old tracheostomized girl with carcinoma larynx

The Indirect laryngoscopic examination revealed a globular exophytic growth involving the epiglottis left AE fold and anterior part of the right AE fold. Left vocal cord could not be seen but movement of the left hemilarynx was found restricted with compromised airway. There was no palpable neck node and no systemic metastasis found. The patient was appeared malnourished and cachectic with anemia. Systemic examination did not reveal any noticeable abnormalities. In Direct laryngoscopic examination, the globular exophytic growth in the larynx was assessed and staged as T₃N₀M₀.

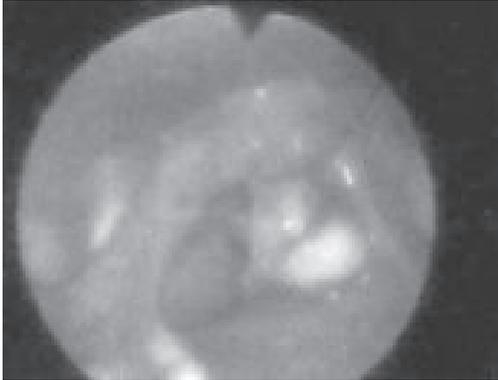


Figure 2: Endoscopic view of laryngeal carcinoma.

Tissue was taken from the growth for histopathological examination and that revealed well differentiated squamous cell carcinoma (SCC). The patient was referred to NICRH, Mohakhali, Dhaka, for oncological management.

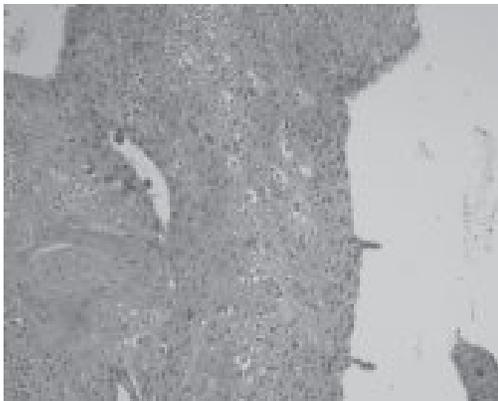


Figure 3: Histopathological section displays feature of squamous cell carcinoma in biopsy from laryngeal lesion.

Due to the personal, financial and communication problem, patient could not afford her treatment. Within two weeks following diagnosis, the girl became more cachectic and ultimately died of cardiopulmonary failure.

Discussion:

Carcinoma larynx is an old-age malignancy. In children, the common laryngeal neoplasm is laryngeal papillomatosis. The most common age of carcinoma larynx is around 6th decade of life²¹. In Bangladesh, mean age at presentation of supraglottic carcinoma is 35-80 years^{22, 23}. 90% of laryngeal carcinoma is squamous cell carcinoma and its different variant²¹. Among the laryngeal carcinoma, supraglottic carcinoma appears earlier in adult population. Supraglottic carcinoma is common laryngeal carcinoma of this subcontinent compared to the western population. This is found more among the younger age group in low socioeconomic group with less male/female ratio in the developing country^{22, 23}.

Clinical manifestations of laryngeal carcinoma in adolescents may include hoarseness or cough, which may be mistaken for common respiratory infections, pre-pubertal voice change, or other benign childhood conditions²⁴. Recurrent respiratory tract infection, or asthma, vocal abuse, difficult examination in children makes the case more aggressive and ultimately fatal^{16, 17}. Due to its rarity, tendency to mimic benign conditions and the relative difficulty of the pediatric laryngeal examination, SCC is not usually considered in the differential diagnosis of persistent hoarseness or cough, which may lead to a delay in diagnosis. Vocal folds are the most common site of involvement by SCC in adolescents, followed by supraglottic and then subglottic locations²⁵.

Etiopathological basis for childhood and adolescence carcinoma larynx is postulated being irradiation for papillomatosis, HPV infection, HPV induced genome mutation, chromosomal translocation^{18, 21}. The relationship of HPV to laryngeal carcinoma is unclear although some studies have attributed laryngeal irradiation as a

predisposing factor in the development of carcinoma. Until the 1970s, malignant degeneration of recurrent respiratory papillomatosis [RRP] was seen in association with patients that underwent radiation therapy for their disease. Spontaneous malignant degeneration of RRP has been reported in the laryngotracheal and bronchioalveolar regions with an incidence rate of 2.3%²⁶. Regarding HPV subtyping, HPV 11 had been linked to the malignant transformation of juvenile-onset RRP²⁷. Malignant transformations of papillomatosis were 14% (6 of 43 irradiated cases) before 30 years of age whereas no transformation was reported in 58 similar cases treated with surgery alone²⁸. Some study relates the risk factors like geographical distribution, malnutrition, and social classes since majority of supraglottic carcinoma cases were from lower socioeconomic group^{22, 23}. The differential diagnosis of laryngeal neoplasm in children includes: papilloma, subglottic hemangiomas, squamous carcinoma, rhabdomyosarcoma, and adenocarcinoma of minor salivary gland.

In this study 10-year-old female child with carcinoma larynx is so far the first reported case in this country. The postulated causes of our case are HPV infection along with factors like malnutrition and effect of secondary smoking or other environmental oncogenic factors. Due to unfamiliarity of this disease among the physician and pediatricians, she was diagnosed in an advanced stage. Aggressiveness of the disease process, malnutrition, lack of medical support, financial constrain, communication inconvenience, she could not get proper and early intervention which caused her unfortunate fatal outcome.

Conclusion:

Childhood laryngeal carcinoma is very rare malignant condition. The etiology of this

disease is not exactly known. Clinical features are aggressive and the outcome is poor since the diagnosis and the treatment is always late. In any suspected cases where some risk factors are associated should be dignified. Although unusual, one should have a high index of suspicion for any hoarseness, cough, or upper airway disease that does not respond to appropriate medical treatment. By reporting this case and highlighting the difficulties in diagnosis and treatment we hope to increase clinical awareness and thus lead to an improved outcome. Accurate and early diagnosis of the childhood laryngeal carcinoma with prompt aggressive treatment is essential.

References:

1. Cunningham MJ, McGuirt WF and Myers EN, Malignant tumors of the head and neck. In: CD. Bluestone, SE. Stool and MA. Kenna, Editors, Pediatric otolaryngology (3rd ed.), Saunders, Philadelphia, 1996; 1557.
2. Gindhart TD, Johnston WH and Chism SE et al. Carcinoma of the larynx in childhood. *Cancer* 1980; 46: 1683–1687.
3. Ohlms LA, McGill T and Healy GB. Malignant laryngeal tumors in children: A 15-year experience with four patients, *Ann Otol Rhinol Laryngol* 1994; 103: 686.
4. Rehn H. Cancer of the vestibule and vocal cords in a 3-year-old boy. *Arch Pathol Anat Physiol Klin Med* 1868; 43: 129–133.
5. Zuckerman JD, Shehata BM, Hill CH & Sobol SE. (2009). Invasive laryngeal carcinoma in a 15-year-old boy with neglected papillomatosis. *The Internet Journal of Otorhinolaryngology* 2009; 8: 32 – 5.

6. Nsamba C, Marrocco G. Carcinoma of the Larynx in a Child. *J Laryngol Otol* 1989; 93: 89-92.
7. Ossoff RH, Tucker GF and Norris CM. Carcinoma of the larynx in a 11-year-old boy. *Otolaryngol Head Neck Surg* 1980; 88 (2): 142–145. View Record in Scopus | Cited By in Scopus (10).
8. Orton HB. Carcinoma of the larynx. Clinical report of a case - age 13 1/2 years. *Laryngoscope* 1947; 57: 299–303.
9. Laurian N, Sadov R, Strauss M, Kessler E. Laryngeal carcinoma in childhood. Report of a case and review of the literature. *Laryngoscope* 1984; 94(51): 684-7.
10. Zalzal GH, Cotton RT, Bove K. Carcinoma of the larynx in a child. *Int J Pediatr Otorhinolaryngol* 1987; 13(2): 219-25.
11. Singh W and Kaur A. Laryngeal carcinoma in a 6 year old with a review of the literature, *J Laryngol Otol* 1987; 101 (9): 957–958.
12. Simon M, Kahn T, Schneider A and Pirsig W. Laryngeal carcinoma in a 12-year-old child. Association with human papilloma virus 18 and 33, *Arch Otolaryngol Head Neck Surg* 1994; 120: 277–282. View Record in Scopus | Cited By in Scopus (24)
13. Rastogi M, Srivastava M, Bhatt MLB, et al. Laryngeal carcinoma in a 13-year-old child. *Oral Oncol Extra* 2005; 41: 207–10.
14. Jones AC. Carcinoma of the larynx in a girl aged 15. *Arch Otol* 1942; 35: 426–430.
15. McGuirt WF, Little JP. Laryngeal cancer in children and adolescents, *Otolaryngology Clinics of North America* 1997; 30: 207-214.
16. Faragó L. & Nagy A. A rare case of malignant degeneration of a childhood laryngeal papilloma. *Mschr Ohrenheilk* 1966; 100: 131–135.
17. Rutt AL, Hawkshaw MJ, Sataloff RT. Laryngeal cancer in patients younger than 30 years: a review of 99 cases. *Ear Nose Throat J* 2010; 89(4): 189-92.
18. Ossoff RH, Tucker GF, Norris CM. Carcinoma of the larynx in an 11 year old boy with late cervical metastasis: report of a case with ten year follow-up, *Otolaryngology Head and Neck Surgery* 1980; 88: 142-145.
19. Brasilino de Carvalho et al. Head and Neck Squamous Cell Carcinoma in Childhood. *Medical and Pediatric Oncology* 1998; 31: 96-99.
20. Birchall MA and Pope L. Tumours of the larynx. *Scott-Brown's Otorhinolaryngology, Head and Neck Surgery*, 7th edition, editor: Michael Gleeson, Edward Arnold. 2008; 194: 2598-2622.
21. Walkinson JC, Gaze MN, Wilson JA. Tumours of the larynx. *Stell and Maran's, Head and Neck Surgery*, 4th ed; *Butlerworth Heinemann*, Oxford 2000; 15: 233-273.
22. Hossain MA, Ahmed SMTU, Alam MM, Tarafder KH, Humayun AHMP. Presenting features of supraglottic carcinoma of larynx. *Bangladesh J Otorhinolaryngol* 2010; 16: 106-112.
23. McDermott A, Raj P, Glaholm J, et al. De novo laryngeal carcinoma in childhood. *J Laryngol Otol*. 2000; 114(4): 293–5.
24. Kitcher ED, Yarny J, Gyasi RK & Cheyuo C. Laryngeal Cancer at the Korle

- Bu Teaching Hospital Accra Ghana. Ghana Medical Journal 2006; 40: 45-49.
25. Gaylis B, Hayden RE. Recurrent respiratory papillomatosis: progression to invasion and malignancy. American Journal of Otolaryngology 1991;12:104-112.
26. Moore CE, Wiatrak BJ, McClathey KD, Koopman CF, Thomas GR, Bradford CR, Carey TE. High-risk human papillomavirus types and squamous cell carcinoma in patients with respiratory papilloma. Otolaryngology Head and Neck Surgery 1999; 120(5): 698-705.
27. French C, Kutok J, Faquin W, et al. Midline carcinoma of children and young adults with NUT rearrangement. J Clin Oncol 2004; 22: 4135-9.
28. Majoros M, Devine KD and Parkhill EM. Malignant transformation of benign laryngeal papilloma in children after radiation therapy. Surg Clin North Am 1963; 43: 1049-1061.