Carcinoma Larynx in Children

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Abstract

Carcinoma of the larynx is uncommon in children 15 years of age or younger. Because of low index of suspicion, the diagnosis of childhood laryngeal carcinoma is often delayed. Other factors contributing to its delayed diagnosis are the similarity of its early symptoms to those of other benign, common childhood conditions and the relative difficulty encountered during pediatric laryngeal examination. Treatment of pediatric laryngeal cancers is challenging, because no guidelines are available.

We believe that every single case of childhood laryngeal cancer should be reported, because of paucity of literature on this childhood entity, it is limited only to case reports and reviews. This makes treatment options and outcomes difficult to interpret.

Keywords: Larynx, squamous cell carcinoma, pediatric.

INTRODUCTION

Malignant neoplasms of pediatric larynx are rare. Laryngeal cancers account for less than 0.1% of all head and neck malignancies in childhood. Since 1868, only 71 cases of childhood SCC of larynx have been reported. We present a case of squamous cell carcinoma (SCC) in a 15-year-old female, along with review of literature, emphasizing on etiological factors and various treatment options available, and their outcomes.

CASE REPORT

A 15-year-old female was referred to our institution, with a history of hoarseness of four months duration. She was earlier seen at primary center, where direct laryngoscopy (DL) and biopsy was done. Biopsy was SCC of left vocal cord. The histopathology review at our institute confirmed the diagnosis (Fig. 1).

There was no history of preceding upper respiratory tract infection (URTI), voice abuse, tobacco use in any form, or previous radiation exposure. There was no family history of malignancy. Indirect laryngoscopy revealed irregular left vocal cord with normal mobility. There was no palpable neck node. On microlaryngoscopy examination, medial surface of left vocal cord was irregular, anterior and posterior commissure were free and there was no supraglottic or subglottic extension. After injecting saline, mucosa upto lamina propria was removed with CO₂ laser using apneic technique. On histopathology examination only

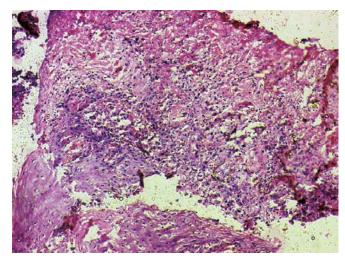


Fig. 1: Photomicrograph showing squamous cell carcinoma with invasion

dysplastic changes were seen. In view of limited disease no adjuvant treatment was advised. One year after diagnosis, she was locoregionally controlled and her speech was good.

DISCUSSION

Carcinoma of laryngopharynx are rare in children. Rehn reported the first case of SCC of the larynx in a child in 1868.³ Since 1868, only 71 cases of childhood SCC of larynx have been reported, including the present case.¹⁻¹⁵

In children, the male preponderance of laryngeal cancer is not as strong as in adults. Of the cases in which gender was indicated, a 2:1 (42/22) male: female ratio was present. The age distribution for the cases in which the exact age

was given: Seven children between one and five years; 17 children between six and ten years; and 45 children between 11 and 15 years. The predominant site of involvement within the larynx was glottis in 28 (78%), supraglottis in 6 (17%) and subglottis in 2 (5%) children. The tumor was confined to the larynx with mobile cords (T1/T2) in 18 cases and fixed cords or exolaryngeal spread (T3/T4) was seen in 17 cases. All childhood primary laryngeal carcinomas reported to date have been squamous, except for one case each of adenocarcinoma, mucoepidermoid carcinoma, adenoid cystic carcinoma and clear cell carcinoma. Metastasis to larynx has also been reported in children. Metastatic cervical nodes at presentation and during follow-up were seen in a single case each. 12,4

The majority of cases reported in children presented with prolonged symptoms of hoarseness or upper airway obstruction. 1-4,6,8 Frequently a delay in the diagnosis was reported with symptoms attributed to vocal changes during puberty, recurrent URTI or vocal abuse. An additional cause is the difficulty in properly examining the larynx in children. A high index of suspicion is necessary to diagnose cancer of the larynx in pediatric subgroup. If relevant symptoms persist or progress in a child, appropriate imaging studies, indirect laryngoscopy or DL examination and biopsy are warranted.

The principal predisposing factor for the development of SCC of the larvnx is irradiation of benign lesions of the head and neck, especially juvenile laryngeal papilloma. 20-22 In the mayoclinic series of 101 cases of laryngeal papilloma, six of 43 (14%) treated with radiation developed SCC of the larynx before 30 years of age, while this does not occur in any of the 58 similar cases treated with surgery alone.²³ Other known risk factors include active and passive smoking and exposure to chemical agents like asbestos. Infection with human papilloma virus (HPV) 18 and 33 has also been associated with childhood laryngeal cancer. 8 A characteristic chromosomal translocation 15:19 has been found in a child with supraglottic cancer. 12 Tobacco use is considered as the most important etiological factor in adult patients. Tobacco use has been considered as the principal risk factor for childhood laryngeal cancer in only two cases. One patient had the history of continuous tobacco chewing for two years 15 and other had the history of smoking half a pack of cigarette per day for three years.²⁴

Treatment of laryngeal carcinoma in children poses many problems. Review of cases reported in literature is not helpful in decision making, as most of these reports have no adequate information regarding staging, treatment and outcome. They have been treated commonly with surgery, radiation therapy (RT) or both and less commonly with chemotherapy (CT). In children and adolescents, radical radiotherapy can result in significant growth retardation of both soft tissue and bone with consequent deformity and dysfunction. Late effects of radiation treatment in children include facial growth retardation, neuroendocrine dysfunction, visual problems, dental abnormalities and hypothyroidism. Late effects at or beyond 10 years from radiotherapy include chondronecrosis, esophageal stenosis, second malignancy and brain hemorrhage. ²⁵

Gindhart et al in their 1980 review of childhood laryngeal SCC found that in 10 of 17 cases treated with RT, surgery, or both, there was no evidence of disease at follow-up intervals ranging from one month to 20 years, and in seven cases the tumor proved fatal. Since 1980, of the 17 cases where the site within the larynx was specified, 13 had glottic primaries and four had supraglottic primaries. Of the 13 cases of glottic primaries, the treatment was cord stripping in two,6 partial laryngectomy in three,4,8,14 total laryngectomy (TL) in two, 10,13 RT in three, 1,7,11 TL plus RT in one¹³ and CT plus RT in two.^{2,3} Though long-term follow-up is not reported for all these cases, local recurrence was seen in only two patients, both of which were successfully salvaged. 1,11 Of the four cases of supraglottic primaries, two died from rapidly progressive disease^{5,12} and two were treated with RT.13,15

It is difficult to formulate treatment guidelines based on these data, but following points can be considered. Supraglottic carcinomas have a rapidly progressive fatal course, therefore, should be treated radically. Glottic carcinomas are associated with a favorable outcome. So in glottic carcinomas, as in our case, laryngeal preservation should be preferred. Until some evidence-based management guidelines are formulated, every case should be treated on its merit.

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