Long term survival of locally advanced carcinoma larynx in an 8-year-old child

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ABSTRACT

Paediatric laryngeal squamous cell carcinomas (SCC) are described in a very small percentage of children. These tumors are presented in the advanced stage because of lack of clinical suspicion. The treatment of paediatric laryngeal SCC is not well-defined. We report a successfully treated locally advanced glottic SCC in an 8-year-old child.

Key words: Chemoradiation, infection, hypothyroidism, laryngeal tumor, radiotherapy

INTRODUCTION

Carcinoma of larynx is a disease that commonly occurs in adults with an incidence of 1.3-8.8% per lakh population in India as stated by Indian Council of Medical Research (ICMR), data provided in 1992. Amongst the paediatric population, incidence of head and neck cancers is about 0.25% with male preponderance and an incidence ratio of 1.78:1. Majority of these laryngeal malignancies were lymphoma, rhabdomyosarcoma, and nasopharyngeal carcinoma.[1] Pediatric laryngeal squamous cell carcinoma as such has not been described much in literature. We report a long time survivor of squamous cell carcinoma larynx, in an 8-year-old girl, successfully treated with radiotherapy and chemotherapy at our institute.

CASE REPORT

An 8-year-old girl presented in May 2003 to our hospital with history of difficulty in breathing and change in character of voice of one week duration, with a diagnosis of suspected neoplasm outside. History was not suggestive of voice abuse or exposure to passive smoking. There was no previous history of benign papillomas. On examination, a mass lesion in the right aryepiglottic fold with fixity to the vocal cord was noted. Contrast computer tomography (CT) scan of the head and neck showed right vocal cord lesion [Figure 1a and b]. She underwent emergency tracheostomy for progressive breathing difficulty. Biopsy of mass lesion with immunohistochemistry (IHC) was reported as poorly differentiated carcinoma, cytokeratin positive [Figure 2a and b] with Ki-67 of 20%. It was diagnosed as T3 lesion with no lymph nodes in the neck and with a final clinical stage of T3N0M0 stage III squamous cell carcinoma of glottis. Retrospectively human papillomavirus (HPV) testing for HPV 6,11,16,18,31 and 33 were done using polymerase chain reaction and were negative. She was planned for neoadjuvant chemotherapy followed by concurrent chemoradiation and chemotherapy with the same regimen following chemoradiation. She received 3 cycles of cisplatin-80 mg/m² on day 1 and 5-fluorouracil 750 mg/m² for 3 days; with partial response. Following second cycle she developed non-neutropenic fever with mucositis, which was managed conservatively. The remaining part of chemotherapy was uneventful. Second phase of treatment with radiotherapy of 46 gray of telecobalt 60, was administered along with weekly Cisplatin chemotherapy. Boost radiotherapy of 10 gray was also given to left side of the neck. Post-concurrent chemoradiotherapy, Cisplatin, and 5-fluoro uracil were given for three cycles. After 8 months, she developed hypothyroidism and is on thyroxine replacement therapy since then and doing well in her studies. At present, the patient is on regular follow-up without any problems except for a tracheotomy scar.

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DISCUSSION

The incidence of laryngeal cancer among children is very rare and accounts for < 1% of paediatric tumors.[2] There are only 21 cases excluding the present case described among children less than 10 years and none from India, to the best of our knowledge.[3-5] It requires a very high index of suspicion among children and adolescents as the symptoms are usually attributed to a respiratory tract infection or change in voice associated with puberty.[6] Though benign lesions like papillomas form majority of laryngeal lesions, it is a diagnosis of exclusion based on macroscopic features like polypoidal growth, friability and also requires confirmation with biopsy and immunohistochemistry, as it is a rare condition. Squamous cell carcinoma (SCC) larynx in adults is associated with many risk factors like smoking, HPV virus infection etc.: However, among children, the most important recognised risk factor is radiation therapy for benign papillomas followed by passive or active smoking and familial history as well as HPV associated papillomas transforming into SCC.[7,8] It is described that papillomas which recur or the ones that progresses to a squamous cell carcinomas are usually associated with HPV type 6,11 genotypes.[9] However, our patient was negative for HPV 6, 11, 13, 16, 31, 33. The treatment of paediatric SCC is not well-defined because of rarity of the tumor and, as such, evidence is confined to only case reports and case series. All three modalities of treatment like surgery, radiotherapy, and rarely chemotherapy have been attempted in various scenarios. In glottic tumors, it is important to preserve the voice as well as balance the long term side effects of radiotherapy. In a review of 13 cases of glottic tumors; patients were treated either with single modality of treatment like surgery (cord stripping, partial laryngectomy, total laryngectomy), radiotherapy or combined modalities like surgery and radiotherapy as well as concurrent chemoradiotherapy. These studies showed encouraging results with only two patients showing local recurrence following only radiotherapy treatment in their short follow-up.[9] The standard treatment for adults with T3/T4 tumors is laryngectomy with post-operative radiotherapy or chemoradiotherapy. There is increasing interest towards larynx conserving treatment modalities in patients with locally advanced carcinoma larynx.[9,10] Our patient was just 8-years-old when she was diagnosed and did not agree for voice losing surgery like laryngectomy and hence opted for chemoradiotherapy. She received 40 Gy with concurrent chemotherapy with cisplatin and post-operative chemotherapy. After nearly 10 years of follow-up, she is doing well with thyroxine replacement for hypothyroidism as a result of neck irradiation.

CONCLUSION

It is very rare to deal with tumors like SCC of glottis in young children. Symptoms can be easily overlooked as it is not suspected, leading to delay in diagnosis and poor outcome. Key to early diagnosis lies in the evaluation of symptoms like persistent hoarseness or cough with difficulty in breathing with laryngoscopy and biopsy of any suspicious lesion than treating conservatively. Modalities like minimally invasive surgeries and chemoradiation which preserve the integrity of larynx and hence speech and swallowing as well as outlook of young patients like ours should be considered whenever possible. Regular follow-up of the patient is very important to identify any immediate as well as late complication arising out of treatment.

REFERENCES

Viveka, et al.: Larynx carcinomas in an 8-year old child


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