Laryngeal squamous cell carcinoma in a 15-year-old

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ABSTRACT

Laryngeal carcinoma in childhood is exceedingly rare, accounting for less than 0.1% of all head and neck malignancies seen in children. Due to its rarity and non specific nature of symptoms, this malignant condition can be easily missed and treated as other common benign conditions. Early diagnosis is important as it influence the choice of treatment modality. We report a rare case of laryngeal squamous cell carcinoma in a 15-year-old boy.

Keywords: Laryngeal neoplasm, children, squamous cell carcinoma, total laryngectomy

INTRODUCTION

Laryngeal carcinoma in childhood is a rare entity, accounting for less than 0.1% of all head and neck malignancies in children. [1, 2] Rehn reported the first case of squamous cell carcinoma in a thee year old child in 1868 [3] and since then, a total of 71 cases had been documented in the literature up until 2010. [4] We report the case of laryngeal squamous cell carcinoma (SCC) in a 15-year-old boy.

CASE REPORT

A 15-year-old Indian boy presented to a general hospital with progression of hoarseness in the previous five months. In that time, he had been treated as recurrent upper respiratory tract infection before being referred to an otolaryngologist. He was diagnosed with a moderately differentiated SCC of the larynx. Subsequently, he was referred to the oncology clinic in our centre for primary radiotherapy. However, he was noted by the oncologist to have stridor and he was referred immediately to the otorhinolaryngology clinic with impending airway obstruction.

He reported that he had been experiencing dyspnoea for past two weeks before the oncology clinic appointment. He had no history of using tobacco or consuming alcohol, or exposure to second-hand cigarette smoke. There was no history of laryngeal papillomatosis, laryngeal surgery, trauma, intubation, irradiation, carcinogenic chemicals exposure

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or sexual activity. There was no reported history of carcinoma in his family. Indirect laryngoscopic examination revealed an exophytic mass involving the entire right vocal cord, extending to the anterior commisure and the anterior two third of left vocal cord. The mobility of both cords was significantly diminished. Other examinations that included routine blood investigations and chest radiography were all normal. A contrast computed tomography scan showed multiple small lymph nodes in the left supraclavicular and both submandibular and cervical regions.

An emergency tracheostomy was performed. Direct laryngoscopy revealed a transglottic tumour that was seen to involve both ventricles, extending to the subglottic region. A repeat biopsy was taken and the findings confirmed the finding of the earlier report (Figures 1). His disease was classified as $T_4N_{2c}M_0$ moderately differentiated SCC of the larynx.

After considerable discussion with the family and the referring oncologist, it was recommended that he should proceed with treatment. A total laryngectomy and bilateral selective neck dissection were performed without any complications. He recovered uneventfully from the surgery and four weeks later was started on a radical concurrent chemoradiotherapy. This comprised of radiation dose of 60 Gy in 30 fractions delivered to the larynx over a six weeks, followed by 50 Gy in 25 fractions to the neck over five weeks. He also received weekly intravenous Cisplatin. He is on regular surveillance follow up and in the last review has been recurrence free for nearly three years.

DISCUSSION

Laryngeal carcinoma in childhood is difficult to diagnose and be easily mistaken for other benign conditions for several reasons; low index of suspicion, rarity of the disease and difficulty with fibre optic endoscopic examination of larynx in children as the procedure can

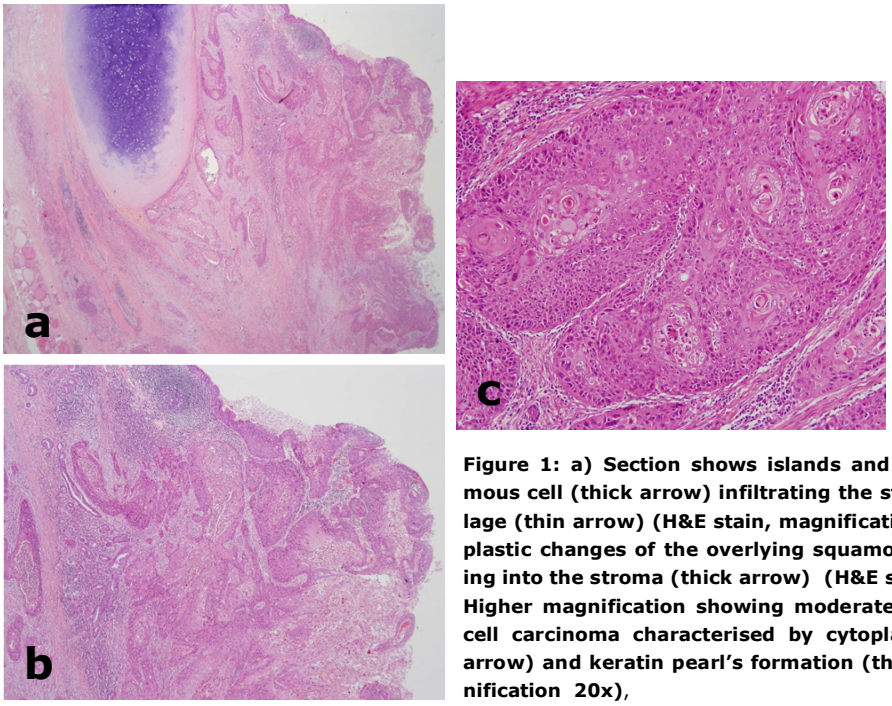

Figure 1: a) Section shows islands and sheets of malignant squamous cell (thick arrow) infiltrating the stroma and spared the cartilage (thin arrow) (H&E stain, magnification 5x), b) presence of dysplastic changes of the overlying squamous epithelium and infiltrating into the stroma (thick arrow) (H&E stain, magnification 10x), c) Higher magnification showing moderately differentiated squamous cell carcinoma characterised by cytoplasmic keratinisation (thick arrow) and keratin pearl's formation (thin arrow) (H&E stain, magnification 20x),

be quite uncomfortable. Hence, they usually diagnosed late with the disease more advanced. [5] Our patient was only diagnosed with laryngeal carcinoma after having hoarseness for five months, and was unfortunately treated as recurrent upper respiratory tract infection. If he had been referred earlier, the diagnosis could have been made in its early stage and he could have been treated with endolaryngeal laser surgery or primary radiotherapy with preservation of larynx. A high index of suspicion is therefore required. As a rule of thumb, persistent hoarseness of more than two weeks regardless of age warrants for referral and a fibre optic endoscopic evaluation of the larynx.

The principal predisposing factor for SCC of the larynx is exposure to radiation to the head and neck region. Majoros *et al*. in their study of 101 cases of juvenile laryngeal papilloma reported that 14% of those treated with radiotherapy had developed SCC of the larynx, whereas none occurred in those treated with surgery alone. [6] Other known risk factors include smoking, exposure to carcinogenic chemicals and family history of malignancy. Our patient did not have any of these risk factors.

It is well established that Human Papilloma Virus (HPV) is associated with the development of laryngeal papilloma. One study showed that HPV-11 was found in all laryngeal and bronchogenic cancers in patients with early onset recurrent respiratory papillomatous. [7] An analysis of 1,435 cases of laryngeal cancer demonstrated positivity for HPV in 24%. [8] Other literature have reported HPV detection in laryngeal carcinoma and the prevalence ranged from 13% to 50%. [9-11] In our case, we did not check for HPV positivity HPV genotyping was not available in our centre.

The management of laryngeal carcinoma in childhood remains a challenge due to the lack of universal guidelines on treatment protocol. Although the tumour tends to behave more aggressively in children, SCC in children is said to respond to treatment similarly to adults cases. [1, 12] However, as we are dealing with a growing adolescent, the psychosocial aspect of the treatment needs to be tackled with due consideration. The decision for total laryngectomy is often difficult to accept by patients and relative. Transglottic carcinoma is associated with high incidence of laryngeal framework invasion and extralaryngeal spread. For this reason, a radical combined approach was adopted in this case. Postoperative concurrent radiotherapy and chemotherapy has been reported to improve the rates of local and regional control as well as disease-free survival in squamous cell carcinoma in head and neck region. [13, 14]

In conclusion, laryngeal carcinoma in childhood is rare and the diagnosis is often delayed. Early recognition is importance as it will influence the choice of treatment modality significantly.

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