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De novo laryngeal carcinoma in childhood

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Abstract
Numerous factors contribute towards a late diagnosis of laryngeal malignancy in childhood. These include its rarity, the similarity of its early symptoms to those of other benign, common childhood conditions as well as the relative difficulty encountered during paediatric laryngeal examination.

We believe that these cases are of sufficient interest when they occur to warrant reporting since the consequences of late diagnosis in these young patients can be serious.

We present a case of an 11-year-old boy with advanced squamous cell carcinoma of the larynx ($T_3N_0M_0$), who was successfully treated with primary total laryngectomy and bilateral selective neck dissections to avoid the potential additional morbidity of radical radiotherapy.

Key words: Larynx; Carcinoma, squamous cell; Child

Case report

An 11-year-old Caucasian boy had presented to another ENT department with a history of dysphonia of two months duration. He was previously noted to be very fit and well. There was no history of preceding upper respiratory tract infection, laryngeal papillomatosis, active or passive smoking, or radiation exposure.

Initial ear nose and throat examination including indirect laryngoscopy was recorded as normal and the patient was subsequently sent for speech therapy. Six months later, he was still dysphonic and soon thereafter he developed stridor. Laryngoscopy revealed an abnormal left hemilarynx with a fixed left vocal fold. An urgent computed tomography (CT) scan confirmed a small transglottic tumour 3 cm^3 in volume with no involved lymph nodes. The patient and his family returned to the UK for further management.

Staging panendoscopy and biopsy showed fixation of the left hemilarynx with a large submucosal swelling extending from the ventricular band and into the subglottis. The left vocal fold was hardly recognizable.

The histopathological diagnosis revealed a well-differentiated keratinizing infiltrating squamous carcinoma.

A subsequent magnetic resonance image (MRI) scan delineated the CT findings more clearly, in particular a predominantly left-sided laryngeal mass which extended around the posterior wall to the right side of the larynx and for 1 cm into the subglottic region (Figure 1). In addition, no significantly enlarged nodal groups were identified. Preoperatively he was therefore staged as $T_3N_0M_0G_1$ squamous cell carcinoma of the larynx. After considerable discussion with the paediatric oncology team the patient underwent total laryngectomy with preservation of pyriform fossa mucosa on the right but not on the left, along with bilateral anterolateral (levels 2–4) selective neck dissections, size 4 provox valve insertion, cricopharyngeal myotomy and left hemithyroidectomy. The pharyngeal

Fig. 1
Macroscopic view of total laryngectomy specimen showing the left-sided transglottic lesion.

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remnant was closed with a vertical double layer anastomosis including mucosa in the first layer and constrictor muscles in the second layer. Inferiorly the trachea was divided 1 cm below the tumour through the fourth ring anteriorly and second ring posteriorly. A fine bore nasogastric tube was passed through the provox valve for feeding.

Post-operatively he made an excellent recovery and started feeding orally by day seven and taking a normal diet by day 10. He was discharged home with a stoma button in situ on day 14, and achieved good quality valved speech by eight weeks.

The surgical specimen confirmed a low volume left transglottic tumour ($1.3 \times 1 \times 0.6$ cm) with clear resection margins. The lymph nodes were clear of tumour. Positive immunohistochemical staining for p53 and Ki67 was found throughout the specimen.

Discussion

The first documented case of laryngeal carcinoma in childhood was published in 1968.[1] Since then there have only been 58 such cases reported in the literature. Of these, all were squamous with one exception, an adenocarcinoma arising from a minor salivary gland within the larynx.[2]

Of the reported cases where sex was indicated, there appeared to be a male preponderance of 3:2, in contrast with a 4:1 sex ratio found in adult patients.[3,4]

Furthermore, the age distribution of cases revealed a rising age dependent incidence: seven tumours were found in the one to five year age group, 14 in the six to 10 year group, and 36 in the 11–15 year olds. Finally there were two cases where age was not documented.[1,5-8]

The principal known aetiological factor for laryngeal carcinoma in young people is radiation, namely irradiation for bening lesions such as juvenile papillomatosis.[3,4,9,10]

In 1963 at the Mayo Clinic, a series of 101 laryngeal papillomatosis cases were studied. Forty-three had been irradiated. Of these, six (14 per cent) had subsequently developed squamous carcinoma of the larynx before the age of 30 years.[11]

In adult patients, smoking is by far the most important aetiological factor. In only one of the 58 previous cases was active smoking identified as the principal risk factor.[5] This 13-year-old boy had smoked half a packet of cigarettes a day for the previous three years. Parental smoking was considered to be a risk factor in one case.[12]

The clinical presentation of all the cases including our patient was upper airway obstruction, dysphonia and cough. These symptoms are commonly attributed to infection or prepubertal voice change in childhood and can lead to delay in diagnosis.[13]

In paediatric practice, there are additional difficulties. Firstly, examination of the larynx often necessitates examination under general anaesthesia and secondly, co-existing juvenile papillomatosis may be misleading.

In cases where tumour site and extent were recorded, including our patient, the vocal folds were the most common site of involvement. In 23 cases, the folds were the only or principal site of involvement and in 11 cases the vocal fold was noted to be fixed.[3] Four children had supraglottic neoplasms[14–16] and in just one case was the tumour found to be subglottic.[17]

Inflammatory cervical lymphadenopathy was frequently described but in no case was there histological documentation of metastatic disease.[3]

The differential diagnosis of laryngeal neoplasm in children includes laryngeal papillomatosis, squamous carcinoma, rhabdomyosarcoma[18] subglottic haemangioma[19] and adenocarcinoma arising from minor salivary glands.

Treatment of laryngeal carcinoma in children poses many problems. With children, even more than adults, there is a strong emotional desire to avoid mutilating surgery and preserve laryngeal function. However, in children and adolescents, radical radiotherapy can result in significant growth retardation of both soft tissue and bone with consequent deformity and dysfunction. Asymmetrical irradiation to the neck can result in significant kyphoscoliosis in later life. Furthermore such patients may incur a significant lifetime risk of radiation-induced second malignancy.[20] Owing to the rarity of epithelial tumours in children, there are few published reports and hence a paucity of data; however it would appear that following laryngectomy, functional restoration in children may be superior to that achieved in adults.[4]

A literature review is not very helpful in the decision-making process because case numbers are small and many reports omit staging information, treatment details and outcomes. An additional source of difficulties is that paediatric oncological services have little experience of the management of head and neck squamous cell carcinoma, while adult head and neck services are unsuitable for supporting children.

In our case, the decision to perform a total laryngectomy rather than to irradiate was difficult. Both adult and paediatric oncologists and surgeons were involved.

After much consultation the team arrived at the conclusion that ablation of this tumour bulk by radiotherapy alone would not be guaranteed and that its effects on neck growth and voice had the potential to become more mutilating than a total laryngectomy. Moreover, the operative field would be less compromised if reconstruction, for example, laryngeal transplantation should become possible in the future. A partial laryngectomy was not thought to be an appropriate treatment option.

The setting chosen for the surgery was a major tertiary paediatric centre where paediatric nursing, the psychological expertise of the paediatric oncology team and parental support were available. Laryngectomy and vocal rehabilitation expertise were provided by the specialist nurse and speech therapist from the adult head and neck centre.

Conclusion

Malignant tumours of the larynx are rare in children. Delay in the diagnosis is common because the presenting symptoms may be mistaken for benign laryngeal or inflammatory airway disease.

Regardless of age, the clinical and pathological appearances of carcinoma of the larynx are similar, although examination is more difficult in paediatric patients. The current literature is unhelpful in treatment planning and treatment should therefore be individualized using the expertise of both adult and paediatric oncology teams.

Finally, 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.

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