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CLINICAL RECORD

Congenital duplication of the larynx

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Abstract

Introduction: The larynx is an intricate structure serving three important functions in humans: it protects the lower respiratory airway, facilitates respiration and helps produce sound through a key role in phonation.

Objective: We report the first published finding of congenital duplication of the larynx in a patient with previously cleared squamous cell carcinoma of the neck and a new diagnosis of squamous cell carcinoma of the lung.

Case report: We describe the incidental finding of duplication of the larynx in a 62-year-old man with previously completely cleared squamous cell carcinoma of the neck, who presented with worsening dyspnoea. We also provide a brief overview of other published cases in which duplication of the vocal folds and epiglottis has been reported.

Results: Our patient experienced no symptoms related to this incidental finding of congenital duplication of the larynx.

Conclusion: The first case of congenital duplication of the larynx is currently of academic interest only; however, the possible association with squamous cell carcinoma is postulated to raise awareness in clinicians who may observe further cases in the future.

Key words: Congenital; Larynx; Bronchoscopy; Carcinoma, Squamous Cell

Introduction

The larynx is an intricate structure and serves three important functions in humans: it protects the lower airway by closing abruptly on mechanical stimulation, thereby halting respiration and preventing the entry of foreign matter into the airway; facilitates respiration through the control of airflow during breathing; and helps produce sound through a key role in phonation.

The larynx develops from the fourth and fifth branchial arches as the laryngotracheal groove, a median longitudinal groove in the ventral wall of the pharynx. It develops from the endodermal lining and adjacent mesenchyme of the foregut. At the fourth and fifth week of gestation, the laryngotracheal groove has deepened and the tracheoesophageal folds fuse in the midline to form the tracheoesophageal septum, leading to the separation of the tracheal airway lumen from the opening of the oesophagus.

This primitive laryngeal aditus develops from a vertical, slit-like aperture into a T-shape through the development of three tissue masses or eminences. The hypobranchial eminence, which first appears on the third week of development, later becomes the epiglottis, while the sides of the 'T' become two arytenoid swellings. The arytenoid swellings are separated by an interarytenoid notch, which later becomes obliterated. Between the fifth and seventh week of gestation, these masses have grown to obliterate the laryngeal aditus, which is recanalised by the tenth week.

Case report

A 62-year-old man with previously completely cleared squamous cell carcinoma (SCC) of the neck presented with worsening dyspnoea. A computed tomography scan revealed a 4-cm right lung mass, which was found to be SCC on histological examination. During bronchoscopy to obtain histology, an incidental finding of duplication of the larynx was made (Figure 1). The true vocal folds were functionally normal while the vestigial secondary vocal folds were non-functioning. On questioning following the procedure, the patient did not complain of any change of voice, recurrent aspirations or other laryngeal symptoms. A duplication of the larynx during development was reported as a novel finding. We tentatively suggest that this finding may provide a possible predisposition for or association with squamous cell malignancies.

- **Duplication of the larynx is a novel finding**
- **Our patient presented with no laryngeal symptoms and our finding was made incidentally on bronchoscopy**
- **Duplication of the larynx may be associated with squamous cell carcinoma**

Discussion

Frank and Malev reported congenital, unilateral duplication of the vocal folds and documented five further cases from foreign literature.[1] Wittig *et al*. reported duplication of the epiglottis in Weyer's acrofacial dysostosis, a rare disorder affecting the development of teeth, nails and bones.[2] This case represents the first reported instance of duplication of the larynx in the published literature. The occurrence of two unrelated squamous cell malignancies in this patient raises the possibility that congenital duplication of the

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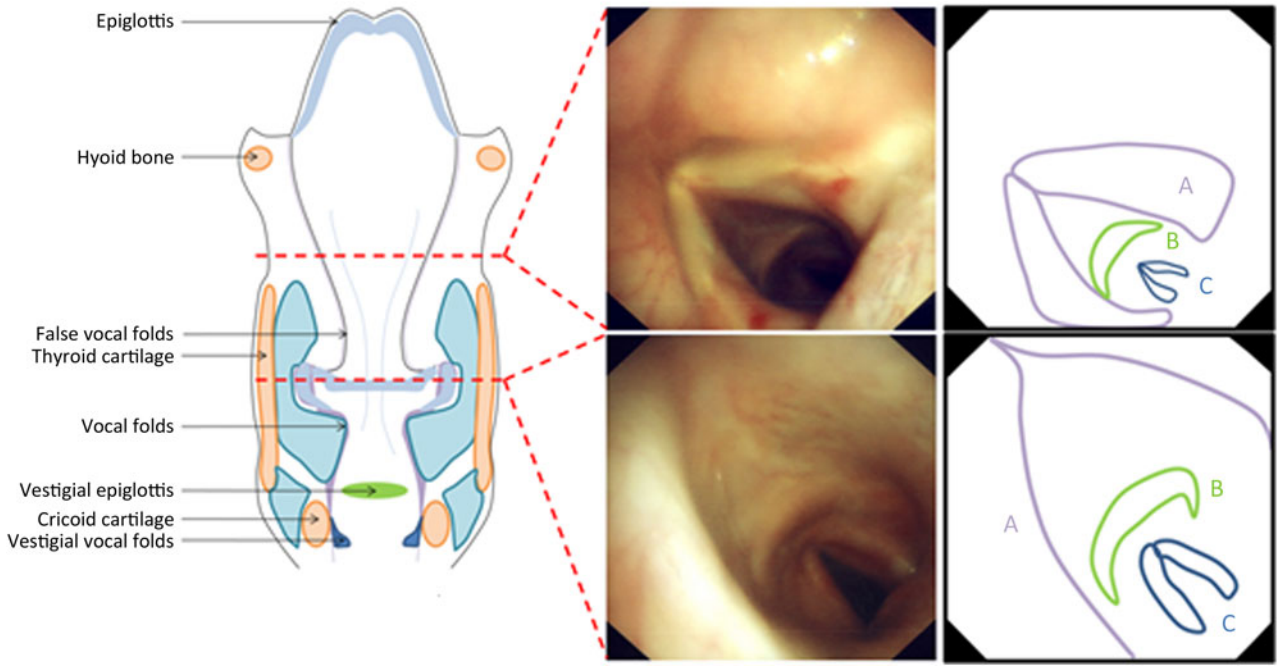

FIG. 1

Photographs taken at bronchoscopy demonstrating duplication of the larynx; the diagrams to the left and right of the photographs show the level at which duplication was observed. A = true vocal folds; B = vestigial epiglottis; C = vestigial vocal folds

larynx may represent an association with this tumour type. However, it is clear that in an isolated case no firm conclusions can be drawn, but in the interest of scientific endeavour and curiosity this point is highlighted for clinicians who may progress to observe further cases in the future.

References

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