International Journal of Pediatric Otorhinolaryngology 111 (2018) 1–6

Contents lists available at ScienceDirect

International Journal of Pediatric Otorhinolaryngology

journal homepage: www.elsevier.com/locate/ijporl

Case Report

Congenital complete absence of tracheal rings with trifurcate carina: Case report of a rare clinical and endoscopic presentation

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ARTICLE INFO

Keywords:
Airway
Congenital anomalies
Bronchoscopy
Tracheomalacia
Tracheal obstruction
Pediatric stridor

ABSTRACT

We describe the case of a child with isolated absence of cartilaginous tracheal rings and a trifurcate carina. At 6 months of age, the patient presented to our multidisciplinary airway clinic with stridor and recurrent severe respiratory infections requiring hospitalization. Radiographs showed airway narrowing. Exam demonstrated biphasic stridor. Flexible fiberoptic laryngoscopy demonstrated only mild laryngomalacia. Operative bronchoscopy demonstrated severe tracheomalacia with absence of any visible tracheal rings and a trifurcate carina. Subsequent CT imaging corroborated these findings and did not demonstrate any other major abnormality. The patient did not require operative intervention and his subsequent course was uncomplicated.

1. Introduction

Tracheomalacia is the second most common etiology of stridor behind laryngomalacia in the pediatric population [1]. Approximately 1 in 2600 live births are affected by airway malacia involving the trachea and/or the main bronchi [2]. Although the majority of patients with tracheomalacia improve with growth and age, some patients with severe symptoms require operative intervention. We present a rare case of severe tracheomalacia caused by full-length absence of tracheal rings.

The C-shaped cartilaginous tracheal rings range in number from 18 to 22 and develop from ventrally located mesenchymal cells after differentiation into chondrocytes during the 10th to 12th week of gestation [3,4]. Malformations in tracheal development are frequently associated with foregut anomalies such as tracheoesophageal fistula and esophageal atresia.

Tracheal obstruction may be categorized as congenital or acquired, further as extrinsic or intrinsic. Extrinsic etiologies of tracheal obstruction include cervical, thoracic or mediastinal masses, and abnormalities of the major vessels that compress the airway. Intrinsic tracheal obstruction may be due to stenosis – iatrogenic stenosis, complete tracheal rings, or sleeve trachea – or severe tracheomalacia. Congenitally absent or deficient tracheal ring cartilage is an exceedingly rare phenomenon that historically has been associated with other major malformations, and has been reported in isolated case reports and small series [5–9].

1.1. Case report

A 6 month old boy presented to our multidisciplinary airway clinic for evaluation of stridor and respiratory distress with crying or feeding. He had a single apneic/cyanotic episode, and had several prior hospitalizations for respiratory illnesses. He was otherwise healthy with no known medical history. A prior chest radiograph demonstrated tracheal narrowing, generating concern for obstruction (Fig. 1). On physical exam the child was noted to be breathing comfortably, but had audible biphasic stridor.

Clinic flexible fiberoptic laryngoscopy demonstrated mild type 1 laryngomalacia with slightly foreshortened aryepiglottic folds. This was not obstructive and could not account for the patient's stridor or history of respiratory distress, and the decision was made to take the patient to the operating room for bronchoscopy.

Direct laryngoscopy/bronchoscopy was performed under general anesthesia, using a spontaneous breathing anesthetic technique (Figs. 2–5). This exam demonstrated a normal glottis, and a normal and patent subglottis. Endoscopy of the trachea, however, demonstrated full length, severe tracheomalacia. There were no identifiable tracheal rings. With absent tracheal rings and nearly full collapse of the airway with respiration, the trachea had an esophageal appearance (Fig. 4). Marked external pulsations were observed, raising concern for anomalies of the major vessels. A trifurcate carina was additionally noted (Fig. 5).

Given the severity of the findings – absent tracheal rings and pulsatile compression – the patient was intubated and taken to the CT scanner in order to evaluate the possibility of associated congenital

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https://doi.org/10.1016/j.ijporl.2018.05.009
Received 20 November 2017; Received in revised form 9 May 2018; Accepted 9 May 2018

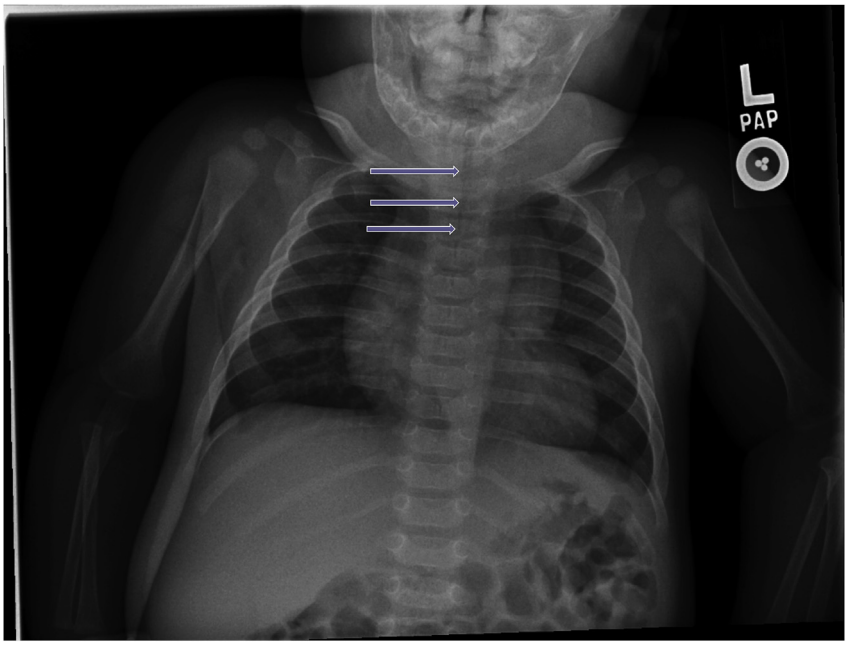

Fig. 1. AP Chest X-ray taken in Emergency Department during an episode of worsened respiratory distress, demonstrating diffusely narrow airway (arrows).

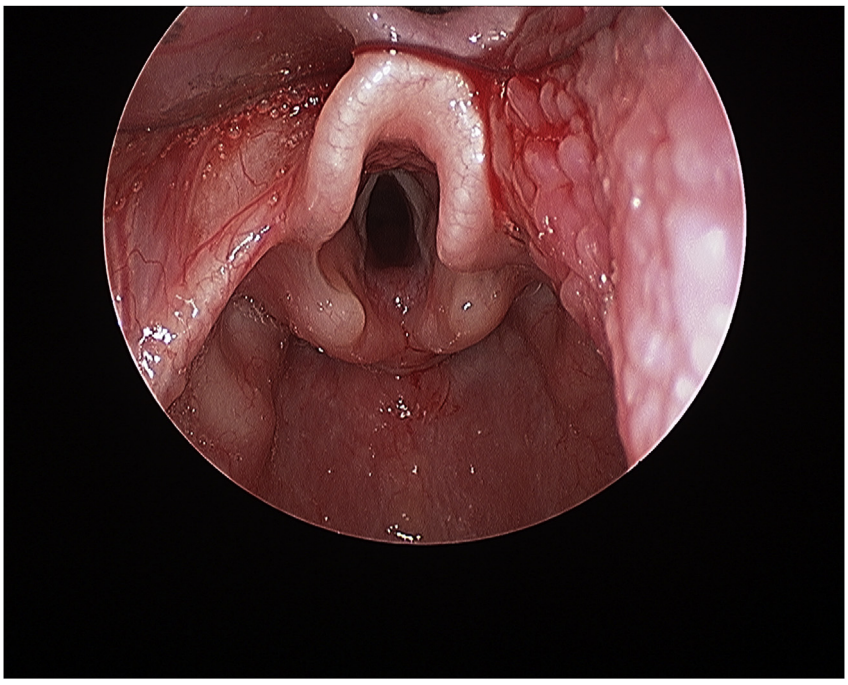

Fig. 2. Direct laryngoscopy reveals minimal to normal findings of supraglottis, and normal appearing true vocal folds.

anomalies, particularly major vessel congenital anomalies. CT did not demonstrate any other abnormalities, but did corroborate the absence of tracheal rings and a trifurcate carina (Fig. 6).

The patient's hospital course was uncomplicated. He was extubated on postoperative day one, and discharged the following day. Esophagram was without evidence of atresia or tracheoesophageal fistula. The patient underwent uncomplicated repeat bronchoscopy 8 weeks later which demonstrated unchanged findings. Over two years of follow-up, his quiet stridor has persisted but he has had no further apneic/cyanotic episodes, and is growing and developing normally. He remains in the upper percentiles of his growth curve, and participates fully in normal activity with no limitations. He has had no further difficulty with feeding. He had one further admission for respiratory distress due to RSV infection at age 2, and required home supplemental oxygen for one week before to returning to his baseline health. No major intervention was planned, and we recommended to the parents of the child that he continue to follow up with pediatric otolaryngology with repeat operative endoscopy as indicated.

2. Discussion

Absent tracheal rings is an extremely rare finding. We are able to identify eight cases reported in the literature (Table 1). All reports describe segmental tracheal collapse from 1 to 3.5cm in length. The majority of these cases were accompanied by other major developmental anomalies, particularly esophageal atresia and