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Unusual presentation of complete tracheal rings in a 15 year old trauma patient

B. S. Schmidt[1], E. J. Herschmiller[2], R. J. Jarrah[1], T. A. Nakagawa[1]

[1]Department of Anesthesiology (Section on Pediatric Critical Care) and Surgery, Wake Forest School of Medicine, Winston-Salem, North Carolina, USA
[2]Department of Anesthesiology, New York Presbyterian/Columbia, New York, USA

Corresponding author: [1]T. A. Nakagawa, Department of Anesthesiology (Section on Pediatric Critical Care) and Surgery, Wake Forest School of Medicine, Winston-Salem, North Carolina, USA. Email: tnakagaw@wakehealth.edu

Key points

Complete tracheal rings may be encountered in the spectrum of congenital tracheal stenosis and is usually diagnosed in early childhood. In rare cases they can remain undiagnosed throughout childhood and manifest as an unexpectedly difficult airway requiring intubation with a much smaller endotracheal tube than would be expected for the patient's size and age

Abstract

Complete tracheal rings may be encountered in the spectrum of congenital tracheal stenosis, and is usually diagnosed in early childhood. We present an unusual case of a 15-year-old trauma patient with progressive respiratory failure and an unanticipated difficult airway during intubation. The patient progressed to cardiopulmonary arrest and required extracorporeal membrane oxygenation for respiratory support. Bronchoscopy revealed complete tracheal rings impeding passage of an appropriately sized endotracheal tube into the airway. A much smaller endotracheal tube was required to intubate this patient's trachea.

Keywords: Intubation, intratracheal, tracheal stenosis, extracorporeal membrane oxygenation, bronchoscopy, tracheal abnormalities.

Introduction

Complete tracheal rings are a finding that may be encountered in the spectrum of congenital tracheal stenosis. Disproportionate growth of the tracheal cartilage related to the posterior tracheal pars membrane, or from a defect in the cervical splanchnic mesenchyme is thought to be a cause for development of this abnormality (1). Three anatomic classifications were described by Cantrell and Guild (figure 1). Type I is characterized by generalized hypoplasia. Type II is characterized by a funnel type stenosis and a gradual taper over the length of the trachea. Type III is a segmental stenosis with no more than 2-3 cartilage rings involved (2).

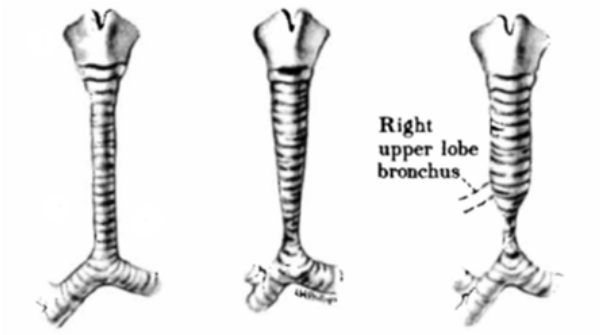

Fig. 1. Three main classifications of congenital tracheal stenosis as classified by Guild et al. (2).

Other types of tracheal stenosis have been described, including a "corkscrew" type of stenosis of the distal trachea (3). Concentric tracheal rings are a common characteristic of each of the many types of congenital tracheal stenosis. An abnormal origin of the right upper lobe bronchus arising directly from the trachea (bronchus suis or "pig bronchus") is seen in up to 20% of cases.

We present an unusual case of a 15-year-old trauma patient with progressive respiratory failure and an unanticipated difficult airway during intubation due to undiagnosed complete tracheal rings. We received parental permission to publish this case report. Further, the Wake Forest Institutional Review Board waives the need for consent for case reports provided they comply with HIPAA regulations.

Case report

A 15 year old male with a history of hypothyroidism and Scheuermann's kyphosis presented to our pediatric trauma center after suffering an all-terrain vehicle accident. Injuries included: three-column fracture of his spine with cord transection at the level of T8-T9, pulmonary contusions, and rib fractures adjacent to the spinal fractures. The patient was admitted to the pediatric intensive care unit (PICU) for blood pressure management and neurologic monitoring, with no respiratory compromise. He was taken to the operating room for laminectomy, decompression of the spinal cord, and posterior spinal fusion. He was easily ventilated using a bag mask during induction. The patient's cervical collar was removed, and inline cervical stabilization was maintained while an asleep elective fiberoptic intubation was performed. A 7.5 cuffed endotracheal tube (ETT) was placed without difficulty, and secured at 23 cm. Endotracheal tube position was confirmed by auscultation and continuous end-tidal carbon dioxide ($ETCO_2$) monitoring. He was placed in the prone position for the posterior spinal fusion. Postoperatively, he was extubated and returned to the PICU with no cardiorespiratory issues. On postoperative day 3 he developed dyspnea with acute rapidly progressive hypoxia. Oxygen saturations decreased to the 70s, and did not improve despite aggressive pulmonary toilet and institution of high-flow nasal cannula oxygen therapy. Due to persistent hypoxemia and impending respiratory failure, rapid sequence induction was performed with etomidate and succinylcholine to electively secure his airway. The initial intubation attempt was unsuccessful; the provider was able to pass a 7.5-mm ETT through the glottic opening; however, more distal (subglottic) resistance resulted in herniation of the ETT back into the laryngeal vestibule. A second intubation attempt was successful with placement of the 7.5 cuffed ETT in the airway confirmed with a colorimetric $ETCO_2$ detector. However, oxygenation saturations failed to improve. Direct laryngoscopy was performed to evaluate ETT position. This examination revealed a Grade 2 view of the ETT passing under the epiglottis and through the vocal cords. Because of persistent desaturation, the ETT was removed, and bag-mask-ventilation was reinitiated, with mild improvement in oxygen saturations. Reintubation with a 7.5 cuffed ETT resulted in color change on the $ETCO_2$ detector when ventilation was initiated. However, despite aggressive manual ventilation with 100% oxygen, saturations did not improve and the patient suffered a bradycardic arrest. Pediatric Advanced Life Support (PALS) measures were initiated. Bilateral breath sounds were minimally audible with intermittent oxygen saturations in the 50s.

Video laryngoscopy was performed with a McGrath size 4 laryngoscope to confirm position of the ETT due to the difficult intubation. The ETT tip was visualized sitting outside the glottic opening in the laryngeal vestibule. The ETT was removed and bag mask ventilation resumed. Repeat direct laryngoscopy (DL) with the same laryngoscope provided a Grade 1 view and a 7.0 cuffed ETT was visualized passing through the vocal cords. Oxygen saturations improved to the mid-60s with manual ventilation and return of spontaneous circulation

(ROSC) occurred after 7 min of cardiopulmonary resuscitation.

Chest radiograph revealed opacification of the entire left chest, concerning for hemothorax, and a chest tube was placed with evacuation of 800 ml of bloody output. This did not appreciably improve saturations and a flexible bronchoscopy was performed to evaluate for mucus plugs or kinking of the ETT. Despite poor visualization of the airway, it appeared that the ETT was unobstructed. Persistent hypoxia despite aggressive airway maneuvers resulted in a decision to pursue extracorporeal membrane oxygenation (ECMO) support. Percutaneous cannulation of the right internal jugular vein and right common femoral vein was performed without complication. Veno-venous (VV) ECMO support was initiated and oxygen saturations quickly improved.

Further attempts to advance the ETT were unsuccessful. Flexible bronchoscopic evaluation was repeated showing the ETT positioned well above the carina (figure 2). Complete tracheal rings were found to comprise the lower two-thirds of the airway (figure 3). The tracheal rings caused a long segment of tracheal stenosis which impeded further advancement of the ETT.

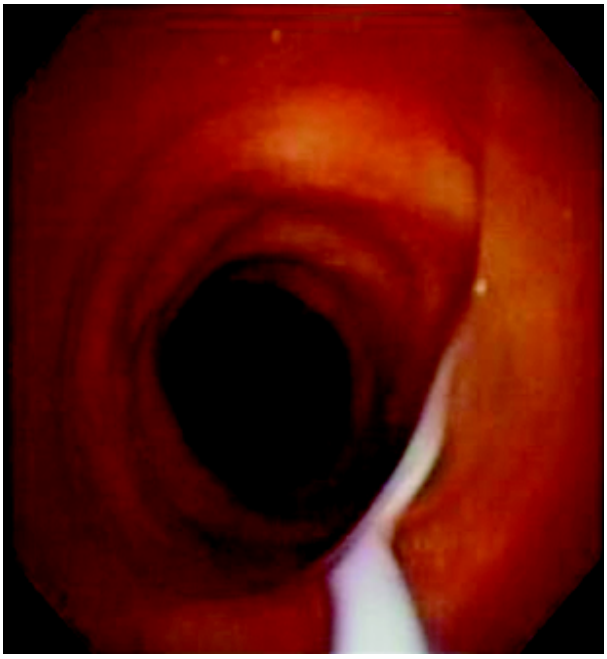

Fig. 2. Tip of 7.5 French endotracheal tube at point of maximal advancement, well above the carina.

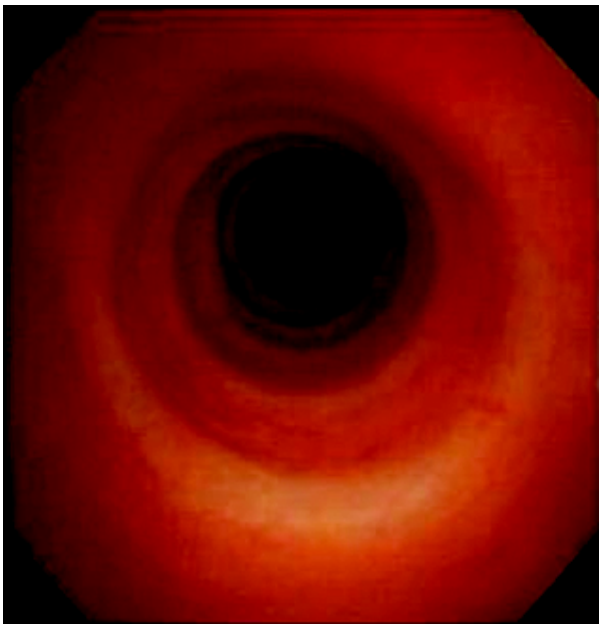

Fig. 3. Concentric tracheal rings observed more clearly after removal of the endotracheal tube while fully supported on extracorporeal membrane oxygenation.

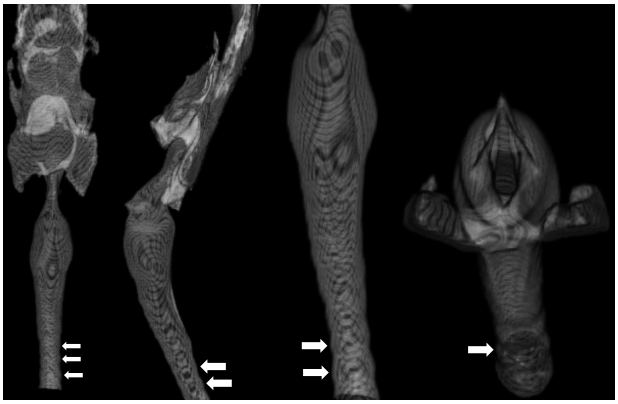

Fig. 4. Three-dimensional reconstruction of the patient's upper airway and proximal tracheal from computed-tomographic images, demonstrating funnel-like narrowing of trachea with a subtle appearance of concentric cartilaginous rings.

A 5.0 uncuffed ETT was placed under direct bronchoscopic visualization and advanced 1-2 cm above the carina. No air leak was noted when this smaller tube was placed in the airway. Review of the initial computed tomography (CT) scan revealed subtle evidence of complete tracheal rings in the distal trachea. These findings were more noticeable after advanced three-dimensional reconstructions were created (figure 4). Repeat chest CT scan did not reveal any intra- or extrathoracic causes for the sudden decompensation or the hemothorax. Cardiac work-up did not reveal any congenital anomalies, ab-

normal ventricular function, or pulmonary arterial sling. The patient was weaned from VV-ECMO to minimal mechanical ventilator support over the ensuing two days.

Unfortunately, neurologic examination revealed that the patient was in a persistent vegetative state and unresponsive despite weaning sedation. Head imaging revealed evidence of global anoxic injury, and a grim prognosis for meaningful neurologic recovery was conveyed to his family.

After further discussion, supportive medical therapies were terminated and the patient expired on hospital day 11. Request for autopsy was declined.

Discussion

Congenital tracheal stenosis is frequently a diagnosis of infancy. The hallmark symptom is biphasic stridor which can be exacerbated by an upper respiratory infection. Diagnosis can also be made at the time of operation for an unrelated issue, as intubation can be difficult (4) or even impossible, requiring the use of laryngeal mask airway or advanced invasive support such as ECMO. Typically, complete tracheal rings are diagnosed and monitored over time by bronchoscopy. Boiselle et al. suggest that paired end-inspiratory dynamic expiratory CT techniques with 3-D reconstruction may provide just as much information as bronchoscopy and can be used as an adjunct (5). In young children, however, dynamic expiratory CT techniques may not be reliable since they require patient cooperation and compliance with breathing instructions. Definitive management of complete tracheal rings will require surgical intervention in the majority of patients. Slide tracheoplasty is the treatment of choice (6). When an open approach is not safe or advisable, endoscopic approaches may be considered. Laser division allows a controlled separation of the complete rings along the posterior wall of the trachea (7). Balloon dilatation under fluoroscopy has been used to successfully divide the posterior aspect of the complete cartilaginous rings (8). Both techniques often involve the postoperative placement of a tracheal stent, and usually require repeat interventions to achieve a sufficiently wide airway (9). Pediatric cardiothoracic surgery and ECMO support should be available in the event of airway compromise.

Tracheal rings are associated with vascular anomalies, most commonly a pulmonary sling (9). Other anomalies have been reported including tracheoesophageal fistula, esophageal atresia, VATER/VACTERL syndromes, cardiac abnormalities, Pfeiffer's syndrome (10), and scoliosis (1).

Our case is somewhat unique since this patient was asymptomatic during childhood. This child never demonstrated stridor and was able to maintain an active lifestyle without breathing problems or evidence of airway obstruction. Importantly, our case illustrates a rare cause of an unexpected difficult airway. Prior documentation of intubation did not suggest a difficult airway; however, fiberoptic intubation was used to provide cervical spine protection. There was nothing to suggest tracheal stenosis during previous airway endoscopy. No problems with increased airway pressures, oxygenation, or ventilation were documented in the intraoperative anesthesia record. We suspect intubation during the first surgery appeared unremarkable because the 7.5 cuffed ETT with the balloon inflated compressed the vocal cords, preventing movement of the ETT, which was likely positioned at the level of the thoracic inlet.

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