

CASE REPORT

Preterm Infant with Congenital Tracheal Diverticulum in the Presence of Esophageal Atresia and Tracheoesophageal Fistula

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ABSTRACT

A 32+4-week-preterm neonate was operated on day 1 for esophageal atresia with tracheoesophageal fistula with a tight primary anastomosis and closure of a tracheoesophageal fistula. Postoperatively, he needed ventilation for 6 days. Post-extubation, he needed continuous positive airway pressure support for increased work of breathing, increasing oxygen requirement, and respiratory acidosis when respiratory support was weaned. Further, during the post-operative period, he had right hemidiaphragmatic paresis and acute life-threatening events requiring manual high-pressure, non-invasive positive pressure ventilation resuscitation. These complications were considered, and a computed tomography assisted by three-dimensional reconstruction was performed. This revealed a congenital tracheal diverticulum and severe tracheomalacia which was confirmed with microlaryngoscopy and bronchoscopy. The presentation and the diagnostic dilemma surrounding this rare diagnosis are discussed in this case report.

Key words: Infant; Newborn; Tracheoesophageal Fistula; Esophageal atresia

INTRODUCTION

Common early complications following esophageal atresia (EA) repair and closure of tracheoesophageal fistula (TEF) include anastomotic leak, gastroesophageal reflux, anastomotic stricture, and missed proximal TEF [1,2]. One of the rare, late complications reported is tracheal diverticulum, usually diagnosed using bronchoscopy [3]. This occurs at the site of ligation of fistula and presents late at a median age of 12 months with repetitive respiratory symptoms not responding to conventional respiratory support. There are, however, no previous reports of congenital tracheal diverticulum associated with EA/TEF in the literature. Here, we describe a rare anomaly of congenital tracheal diverticulum associated with EA/TEF presenting early at 2 weeks of age in a pre-term infant. We also describe the feasibility of definitive diagnosis using the novel approach using computed tomography (CT) and three-dimensional (3D) reconstruction along with microlaryngoscopy and bronchoscopy (MLB) in the neonatal period. Congenital tracheal diverticulae are a poorly recognized finding in EA/TEF and can

be asymptomatic particularly if distal in location, but in the presence of severe tracheomalacia, they may present early with respiratory symptomatology as in our case [4].

CASE REPORT

The preterm (32+4 weeks) neonate was born vaginally with a late antenatally diagnosed EA. He underwent a tight, primary anastomosis of the EA on day 1 of life with closure of a TEF. Post-operatively, the neonate remained ventilated for 6 days. Following extubation, he needed continuous positive airway pressure due to rising oxygen requirements, respiratory acidosis, and increased work of breathing respiratory support which was weaned. A chest radiograph on day 5 showed a high right dome of the diaphragm suggestive of right hemidiaphragmatic paresis, a diagnosis that was confirmed on ultrasound.

On day 8, he developed acute life-threatening events (ALTEs) requiring high-pressure, manual non-invasive positive-pressure ventilation resuscitation. These epi-

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sodes persisted increasing in severity and were unresponsive to medical management of potential gastroesophageal reflux, and the electroencephalograph (at 6 weeks) was not suggestive of any seizure. MLB was undertaken and diagnosed severe, lower tracheomalacia (Figure 1). The neonate had a cardiorespiratory collapse at the start and during the procedure that necessitated resuscitation and premature termination of the procedure.

Subsequently, the baby underwent a CT chest with contrast at 4 weeks of age to exclude a vascular ring or any other external compressing structure. The CT found a posterior tracheal diverticulum with the origin just proximal to the carina, lower to the EA/TEF repair. The distal end of the diverticulum ended below the carina dividing into two blind ends like those anatomically expected from bronchiolar division; the anatomy expected of a congenital not acquired tracheal diverticulum (Figure 2) [5,6].

The neonate continued to suffer from ALTEs while intubated. 10 days following the CT, he had a second MLB, which confirmed the presence of the diverticulum with the entrance 1 cm proximal to the carina (Figure 3). As part of the MLB, he was intubated past the diverticulum; this stabilized his airway and the ALTEs resolved. Upper gastrointestinal contrast confirmed only mild stenosis of the esophageal repair, without a fistula.

Definitive treatment involved surgical resection of the diverticulum with aortopexy. Further imaging ruled out VACTERL sequence. Brain magnetic resonance imaging undertaken to investigate the extent of hypoxic damage from multiple episodes of resuscitation showed hemosiderin deposition over the left frontal convexity and hemorrhage in the cerebellum; there was no evidence of hypoxic brain injury in the diffusion-weighted imaging, T_1 and T_2 sequences. He did not require tracheostomy and now breathes without any respiratory support in room air and has done for 2 weeks post-aortopexy.

DISCUSSION

EA and TEF affect 1:300–4500 live births, most commonly in combination [1]. The majority are repaired in the 1st days of life by primary anastomosis if the esophageal gap will allow. It is preferable to extubate onto air and avoid reintubation or positive pressure ventilation to protect the anastomosis and avoid anastomotic leak. When a patient is born prematurely, the chance of extubation onto room air without non-invasive support is lower due to the lung immaturity, as in case of our case.

Tracheal diverticulum can also present with airway compromise and difficult ventilation [2]. Tracheal

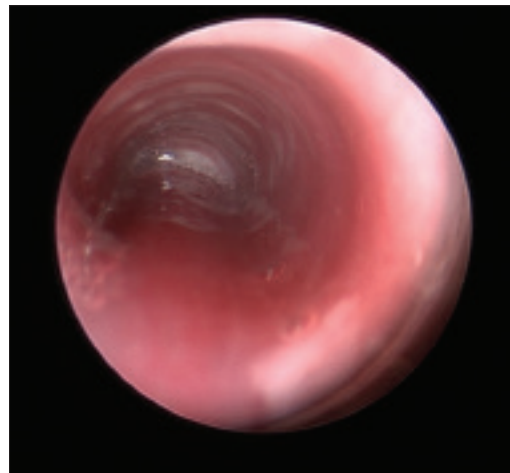


Figure 1: Microlaryngoscopy and bronchoscopy view of tracheomalacia

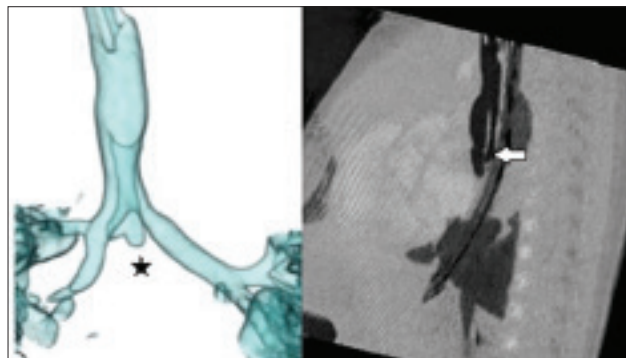


Figure 2: Three-dimensional computed tomography (CT) reconstruction of the tracheal diverticulum (starred) and two-dimensional CT in the sagittal plane (arrowed)

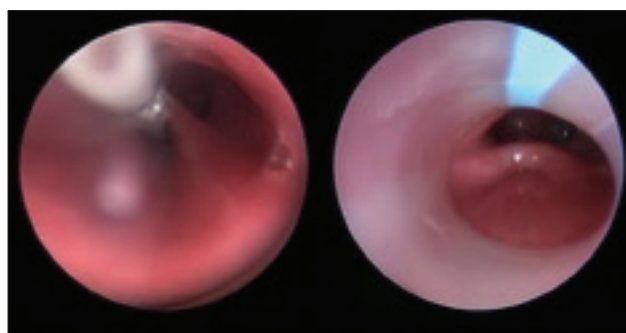


Figure 3: Microlaryngoscopy and bronchoscopy view of the diverticulum

diverticulum is best diagnosed with dynamic three-phase bronchoscopy; however, the airway instability of our patient hindered the attempts.

Although multidimensional CT with 3D reconstruction has been used in adults to diagnose tracheal diverticulum, this is the first report of use in a preterm neonate to diagnose tracheal diverticulum. As mentioned above, the bifurcation of the diverticulum as

seen in the 3D CT reconstruction suggests the potential development of bronchioles from the structure and thus the diverticulum was congenital and not acquired [5,6]. The right, posterolateral position of the diverticulum just above the carina also supports the congenital diagnosis, along with the fact that the EA closure was high and a TEF would not extend so low or indeed to beyond the carina, as the diverticulum does. Histology of the diverticulum was consistent with a tracheal pouch; however, the tissue had considerable diathermy artifact and so could not be considered diagnostic. We accept that this is a limitation along with the unfortunate absence of MLB confirmation but feel that the anatomical features specified above are significantly suggestive of a tracheal origin.

The understanding of the embryogenesis of this combination is poorly understood; however, the embryogenesis of EA/TEF alone also remains poorly understood. Currently, the most popular theories of EA/TEF formation consist of a failure of separation of the dorsal and ventral anterior foregut into the gastrointestinal and respiratory components, respectively, at around 3–5 weeks' gestation [7]. The drive for this failure appears to be dependent on the molecular expression of growth factors; however, the growth factors involved remains to be established. A congenital tracheal diverticulum is believed to occur from a defect of endodermal differentiation and tracheal cartilage formation during the 6th week of development [8]. It is plausible that altered growth factors resulting in EA/TEF formation could also impact the posterior wall formation of the trachea, resulting in diverticulum, but this is purely speculative.

Despite the progressive improvement in the surgical management of EA/TEF, the majority of patients experience long-term structural and functional esophageal and tracheal complications such as tracheomalacia [9]. Our patient had severe tracheomalacia, which was treated with aortopexy. Patients with severe tracheomalacia are more likely to experience ALTEs and require aortopexy or tracheostomy formation [10]. Tracheomalacia is evaluated most commonly with MLB or fluoroscopy. In a case series of patients with EA, 18.9% developed severe tracheomalacia at a median of 18 days postsurgery [11]. While 20% of these patients died, others went on to have aortopexy, fundoplication, or medical management. Patients managed with aortopexy had a shorter length of hospital stay and fewer readmissions for respiratory issues at 2-year follow-up [11]. This suggests that timely aortopexy for severe tracheomalacia might be the best treatment option.

Posterior tracheopexy is also emerging through the Esophageal and Airway Treatment team at Boston Children's Hospital as a primary or secondary treatment option for tracheomalacia associated with EA. Symptom resolution has been found to be significant

as it has ventilator dependence, compared with primary and secondary tracheopexy boasting comparable results [12]. Only 9.2% of 98 patients required reoperation such as aortopexy [13].

CONCLUSION

Congenital tracheal diverticulum in the presence of EA/TEF is a rare diagnosis, not previously addressed in the literature, and provides significant challenges in management in the presence of tracheomalacia. Bronchoscopy and CT are the gold standard of the investigation for tracheal diverticulum, and indication for surgical management is assessed on severity of symptoms. Current management of severe tracheomalacia consists of aortopexy or posterior tracheopexy, with surgical developments questioning the case for preventative primary tracheopexy at the time of EA/TEF repair.

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