Congenital Trifurcation of the Trachea

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Introduction

“Tracheal trifurcation” with an ectopic bronchus or foregut fistula is exceedingly rare. A “tracheal bronchus” is uniformly found within 2 cm of the carina supplying the right upper lobe. In children undergoing bronchoscopy for respiratory symptoms, an ectopic bronchus was found in 2% of the cases. These may be asymptomatic or associated with bronchiectasis, focal emphysema or cystic lung malformations. Implications at intubation, bronchoscopy, and surgery can be complex.

Case Presentation

A 2-year-old girl suffered right lower lobe pneumonia. On resolution, she remained symptomatic with a dry cough and reduced exercise tolerance. She had an antenatal history of a right echogenic lung mass. Computed tomographic imaging at 3 months demonstrated tracheal trifurcation at the carina with normal vascular anatomy (►Fig. 1). Flexible bronchoscopy confirmed bronchomalacia of the ectopic bronchus (►Fig. 2).

At thoracotomy, a single artery supplied the lower lobe draining via its own inferior pulmonary vein (►Fig. 3). The right lower lobe was excised and histology confirmed bronchiectasis. Recovery was uneventful and at 3-month follow-up exercise tolerance and energy levels had improved.

Discussion

To our knowledge, the only previously reported case of an ectopic bronchus supplying the right lower lobe (as opposed to the upper lobe) was associated with a sequestration. A rare “bridging bronchus” crossing the mediastinum has previously been reported, representing an origin of the right lower lobe bronchus from the left bronchial tree.

The relationship between esophageal atresia and tracheal bronchus has been demonstrated in the Adriamycin rat model, in which approximately one-fifth had a tracheal bronchus. This corresponds with case reports of VACTERL children with tracheobronchial malformations and highlights the need for comprehensive evaluation of the tracheobronchial tree in esophageal atresia before repair is undertaken. Although it is suspected that these malformations may carry an increased malignant risk this has not been quantified in the literature. There have been multiple cases of lung cancer (squamous cell carcinoma, adenocarcinoma, small cell carcinoma, and bronchial carcinoid) associated with a tracheal bronchus, but these have generally been in adults over 50 years of age with a history of smoking or other lung pathology.
Conflict of Interest
None.

References

