The Ectopic Tracheal Bronchus: Management of a Child by Excision and Segmental Pulmonary Resection

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In their classic study, Huizinga and Smelt described several variations of the congenital anomaly which they termed tracheal lobe. Essentially, an otherwise normal right upper lobe bronchus originates above the tracheal carina or bifurcation and provides the customary three segmental divisions. There may, however, be a complete duplication of the right upper lobe bronchus by an ectopic tracheal bronchus. Usually the anterior and posterior bronchopulmonary segments are served by the normal upper lobe bronchus, the ectopic tracheal bronchial evagination serving the apical segment only.1

If drainage of the appropriate bronchopulmonary segment is impaired, or bronchiectasis, cystic change or emphysema develop because of stenosis of the origin of the ectopic bronchus, then the patient may develop recurrent episodes of pneumonia or dyspnea.

The purpose of this publication is to present a child in whom recurrent episodes of right upper lobe pneumonia led to investigation and diagnosis of an ectopic tracheal bronchus serving the right apical segment, with cure of the condition by thoracotomy and resection of the ectopic bronchus and its associated apical pulmonary segment.

Case Report

K. T. is a three-year-old white girl who has been known to have a harsh systolic murmur in the second right interspace consistent with the diagnosis of congenital aortic stenosis. In the past year, she has had recurrent episodes of pneumonia involving the right upper lobe and on this admission, x-ray examination of the chest confirmed the presence of atelectatic pneumonia in the right upper lobe (Fig. 1) accounting for her pyrexial episode associated with cough which was productive of sputum and a white cell count of 10,400 per mm³ with 57 per cent segmented forms. In view of the recurrent episodes of pneumonia, it was felt that bronchoscopy and bronchography were indicated to exclude the possibility of an underlying congenital abnormality, particularly in view of the fact that an esophagram had demonstrated an area of mild constriction at the thoracic inlet suggestive of a vascular ring.

Under general anesthesia, bronchoscopy was carried out and the vocal cords were seen to be normal, the trachea appeared normal with no indentation or distortion, nor was there any apparent deformity attributable to extrinsic pressure, just above the bifurcation a small cryptic orifice was noted, but no other abnormality was seen.

Bronchography demonstrated an ectopic tracheal bronchus arising just above the carina. The tracheal bronchus had its origin 6 mm. above the bifurcation with an apparent diameter of 3 mm at its origin with minimal dilation in its mid portion. After coursing 1.8 cm, it divided into two branches, one running superiorly and one anterolaterally (Fig. 2.)

It was noted that the segment of the ectopic bronchus did not empty as well as the remaining bronchopulmonary segments and it was further noted that there was fusion of the basilar branches of both lower lobe bronchi.

In view of this congenital bronchial abnormality predisposing to recurrent episodes of atelectatic pneumonia it was decided to resect the bronchus with its appropriate parenchymal pulmonary tissue and one week later through a right fourth interspace posterolateral thoracotomy, the ectopic bronchus was resected together with the apical segment of the right upper lobe.

During the operative procedure, careful dissection was performed seeking an aberrant right subclavian artery to account for the indentation of the esophagus, but no such lesion was evident.

The child made an uneventful recovery from this procedure and was discharged from the hospital eight days later.

Description of the Ectopic Bronchus and Segment of Lung

The specimen consisted of a portion of lung and attached bronchus weighing 6 grams. The lung tissue measured 6.0 x 2.0 x 0.8 cm and...
was reddish-tan, soft and crepitant. One surface was covered with a thin translucent pleura with scattered petechiae. The bronchus measured 1.5 cm in length with the lumen measuring 0.4 x 0.2 mm, with a wall 1 mm in thickness. An adjacent lymph node was reddish-gray in color measuring 1.5 x 1 x 0.5 cm, its cut surface being dark red.

**Microscopy**

The section of the lung showed atelectasis over most of its extent, numerous scattered lymphoid follicles being present. The bronchi lining was composed of ciliated columnar epithelium. The submucosal glands and cartilage were unremarkable. The lymph nodes demonstrated prominent germinal centers and reticuloendothelial hyperplasia.

**Discussion**

An anomalous ectopic tracheal bronchus is an uncommon condition usually representing a "displaced" apical segmental bronchus and less commonly a true supernumerary bronchus. In his original investigation into the anatomy of this condition in 1889, Chiari described the occasional tracheal origin of the right upper lobe or eparterial bronchus, and he also noted the occasional presence of an accessory bronchus arising from the trachea proximal to the normal eparterial bronchus once per 1,200 necropsies. Herxheimer subsequently described a small functional "third lung" stemming from a tracheal bronchus 2.5 cm distal to the cricoid and, in fact, reflected a true accessory lung communicating with a supernumerary bronchus.

**Embryology**

The rudiments of the primitive respiratory system first appear in the fourth week of fetal life as a median laryngotracheal groove in the ventral wall of the pharynx. This groove deepens and its lips fuse to form a longitudinal septum, converting the groove into an entodermal lined tube from which the respiratory tract develops. Faulty development of this septum is responsible for varying degrees of tracheal stenosis, as well as tracheoesophageal fistula, the latter usually being associated with esophageal atresia.

Proximally the cephalic part of the tube forms the larynx which opens by a slit-like aperture into the pharynx. The succeeding part of the tube forms the trachea. Two lateral outgrowths develop caudally to provide the right and left lung buds, the surrounding mesoderm developing into the pulmonary parenchyma as well as providing the bronchial connective tissue. With fetal development the lungs migrate caudally so that by the time of birth the tracheal bifurcation is situated at the level of the fourth thoracic vertebra. The right and left lung buds normally divide into lobules, three appearing on the right side and two on the left. The subsequent delineation of septa leads to the formation of the segmental bronchi.

**Figure 1:** X-ray film of the chest demonstrating area of upper lobe atelectasis.

**Figure 2:** Bronchogram defines the ectopic tracheal bronchus with two divisions within the apical segment of the right upper lobe.
**Congenital Abnormalities of the Tracheobronchial Tree**

Tracheal or bronchopulmonary developmental anomalies include varying degrees of agenesis, absence or deformity of cartilaginous rings or intrinsic constriction by webs or stenosis. External pressure may, of course, be associated with vascular reduplications. Evaginations of the trachea or bronchi will result in the formation of diverticula, cysts, fistulae or an aerocoele (tracheocele or bronchocele). The evagination from the trachea may give rise to a tracheal lobe and from the bronchi may give rise to supernumerary bronchi with supernumerary lobes and fissures. Rarely, anomalous bronchial and pulmonary tissue may develop and such tissue may be anatomically related to some part of the respiratory system or may on occasion be related to tissue other than those of the respiratory system accounting for its occasional situation in the infradiaphragmatic locale.

Although the most frequent congenital tracheal abnormality is a tracheoesophageal fistula with esophageal atresia, usually a blind proximal pouch with a distal tracheoesophageal fistula, an occasional variant is a tracheoesophageal fistula without esophageal atresia.

Congenital stenosis of the trachea is a rare lesion and in their presentation of a case Cantrell and Guild described a seven-year-old child with stenosis of the distal trachea with anomalous origin of the right upper lobe bronchus proximal to the stenotic area, and referred to their inability to trace more than 24 acceptably documented cases. In describing three morphologic variants of stenosis, namely generalized hypoplasia, funnel-like stenosis and segmental stenosis they noted the not infrequent association of tracheal stenosis with abnormal origin of major bronchi. Evans has reported a tracheal origin of the right upper lobe bronchus in association with segmental stenosis while Holinger and co-authors described a case in which the entire right bronchial tree arose from the left main bronchus in association with segmental tracheal stenosis.

Such a tracheal evagination may occur without tracheal stenosis, and may on occasion compress the esophagus causing symptoms identical with those due to a vascular ring abnormality. An anomalous supernumerary bronchus is most frequently seen in relation to the right upper lobe and usually arises as did this case from the posterolateral margin of the trachea 1½ to 2 cm above the tracheal bifurcation.

The supernumerary bronchial orifice may be visualized bronchoscopically proximal to the carina, while bronchography will demonstrate the tracheobronchial architecture, as well as identifying the segmental area of pulmonary tissue subserved by the ectopic bronchus.

Surgical treatment includes ablation of the ectopic tracheal bronchus and resection of the corresponding area of pulmonary parenchyma, thereby preventing further pneumatic episodes and progression of bronchiectatic and other complications.

**References**


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