CASE REPORT

A case of tracheal bronchus associated with right aortic arch and partial anomalous pulmonary venous connection

Sağ arkus aorta ve kısmi pulmoner venöz dönüş anomalisine eşlik eden trakeal bronş bulunan olgu

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Summary– Tracheal bronchus includes a variety of bronchial anomalies arising in the trachea or main bronchus and directed toward the upper-lobe territory. Reported incidence varies from 1–3% in the pediatric population. It is generally associated with other congenital malformations, including costovertebral anomalies, congenital airway and lung anomalies, vascular anomalies, and congenital heart defects. Presently described was the case of a 14-year-old female with tracheal right-upper-lobe bronchus, right aortic arch with mirror image, and abnormal left upper pulmonary venous return to innominate vein.

Özet– Trakeal bronş trakeadan veya ana bronştan kaynaklanan ve üst lob bölgesine doğru yönelen çeşitli bronş anomalilerini içerir. Pediyatrik popülasyonda insidansının %1-3 arasında değiştiği bildirilmiştir. Genellikle kostovertebral anomaliler, doğumsal havayolu ve akciğer anomalileri, vasküler anomaliler ve doğumsal kalp defektleriyle ilişkilidir. Bu yazıda, trakeal sağ üst lob bronşu, sağ aort kavsi ayna görüntüsü dallanması ve inominat vene sol üst pulmoner venin normal dışı girişi olan 14 yaşındaki bir kız hasta sunuldu.

A tracheal bronchus is an aberrant bronchus that arises most often from the right tracheal wall above the carina, and is the most common developmental airway anomaly of the trachea. Tracheal bronchus is present on the right side in 0.1–2% of the population, and on the left side in 0.3–1%. It usually originates 2–6 cm proximal to the carina.[1,2] Although a majority of patients are asymptomatic, tracheal bronchus may cause persistent or recurrent upper-lobe pneumonia, atelectasis, or air trapping and intubation complications.[1,3]

Tracheal bronchus associated with congenital heart defects has been previously reported.[4–7]

CASE REPORT

A 14-year-old asymptomatic girl was referred due to an incidentally found murmur. Physical examination revealed normal findings, with the exception of a 2/6 systolic murmur best heard at the left sternal border. Electrocardiography was normal. Transthoracic echocardiography revealed right aortic arch and vertical vein connecting to the right superior vena cava via the innominate vein. Cardiac catheterization and angiography demonstrated left upper pulmonary venous return to the innominate vein via the vertical vein, and right aortic arch with mirror image (Figure 1a, b). Pulmonary-to-systemic blood flow ratio (Qp:Qs) was 1.24, and pulmonary artery pressure was 28/10 (20) mmHg during cardiac catheterization and angiography. Computed tomography demonstrated these findings and an accessory tracheal bronchi to the right upper lobe, as well as mild compressing effect on the trachea (Figure 1c, d). She had no history of respiratory symptoms or recurrent pneumonia.

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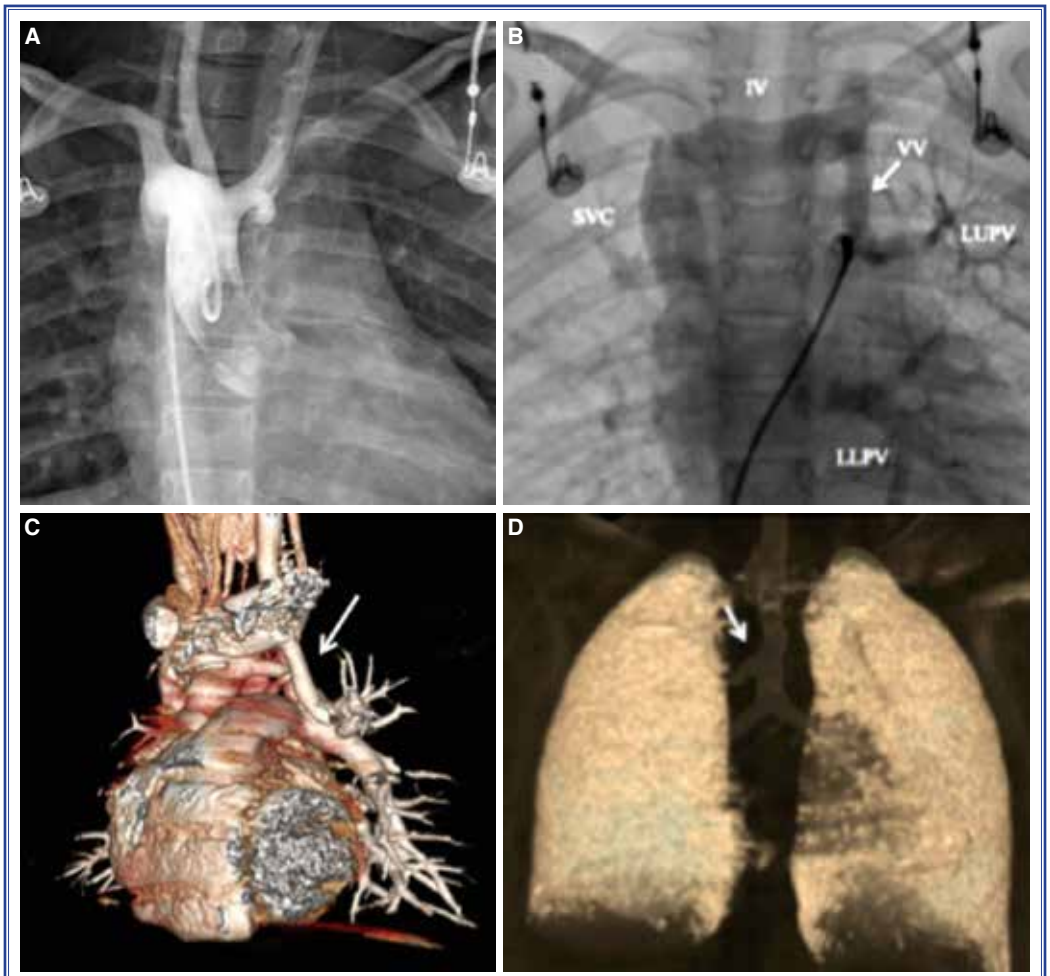

Figure 1. **(A)** Aortogram demonstrating right aortic arch with mirror image branching. **(B)** Pulmonary venous return phase of contrast material injection to the pulmonary artery, demonstrating normal left lower pulmonary venous return and abnormal left upper venous return to innominate vein via vertical vein. **(C)** A 3-dimensional reconstructed view of abnormal pulmonary venous return via vertical vein (arrow). **(D)** Tracheal bronchi to the right upper lung lobe (arrow). IV: Innominate vein; LLPV: Left lower pulmonary vein; LUPV: Left upper pulmonary vein; SVC: Superior vena cava; VV: Vertical vein.

DISCUSSION

Tracheal bronchus was described by Sandifort in 1785 as a right upper bronchus originating in the trachea.[1] The term tracheal bronchus includes a variety of bronchial anomalies arising in the trachea or main bronchus and directed toward the upper-lobe territory. This anomalous bronchus usually exits the right lateral wall of the trachea, and can supply the entire upper lobe or its apical segment.[1–3] It may be asymptomatic and incidentally located on chest computed tomography or bronchoscopic examination for other respiratory diseases.[3,8] Reported cases were associated with congenital costovertebral anomalies, congenital airway anomalies, tracheal stenosis, and vascular and congenital heart defects.[1–7] In addition, higher incidence of tracheal bronchus has been found in patients with Down's syndrome and VACTERL association (association of vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula and/or esophageal atresia, renal and radial anomalies, and limb defects).[4,9]

Reported congenital heart defects associated with tracheal bronchus include atrial septal defect, ventricular septal defect, pulmonary stenosis, atrioventricular canal defects, partial abnormal pulmonary venous return anomaly, and tetralogy of Fallot.[4–7]

Ioannis et al.[6] reported a young male with a combination of tracheal bronchus, right upper pulmonary

venous return to the superior vena cava, and right aortic arch, with respiratory symptoms attributed to asthma. Similar findings were observed in the present patient, with the exception of abnormal left upper pulmonary venous return. Combinations of bronchial anomalies and heart defects appear to reflect relatively specific developmental field defects, affected by both the spatial relationships of organs near the developing foregut and by temporal sequence. Patients with Turner's syndrome have a high incidence of aortic arch anomalies and abnormal partial pulmonary venous return, yet the present patient had no signs of Turner's syndrome. Computed tomography showed mild compression of the trachea due to right aortic arch, without respiratory symptoms. Due to the absence of symptoms and low Qp:Qs ratio, conservative follow-up without surgical intervention was planned.

Most patients with tracheal bronchus can be conservatively treated, but in patients with persistent symptoms, surgical excision of the involved segment is necessary. In patients with tracheal bronchus, endotracheal intubation may cause severe complications, including perioperative persistent hypoxemia, atelectasis of the involved lobe or segment, post-obstructive pneumonia, accidental intubation of the anomalous lobe resulting in pneumothorax, and inadequate ventilation of the remaining lung.[1,2] Thus, it should be preoperatively diagnosed in order to avoid such complications.

To our knowledge, this is the first reported case with such a combination. Due to the eventual need for surgery and endotracheal intubation in some patients with heart defects, diagnosis of bronchial anomalies is important, and should be considered in intubated patients, especially those with right-upper-lobe complications.

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