

Carinal trifurcation associated with isolated partial anomalous pulmonary venous return

İzole parsiyel pulmoner venöz dönüş anomalisi ile birlikte karinal trifurkasyon

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ABSTRACT

Carinal trifurcation is described as an opening of the right upper lobe or accessory lobe close to the tracheal bronchus and carina. The etiology of these malformations has not been clearly understood since they commonly occur with cardiac anomalies resulting from their common embryonic development. Isolated partial anomalous pulmonary venous return with an intact atrial septum is a congenital anomaly where one or more of the pulmonary veins are connected to the venous circulation. To the best of our knowledge, the coexistence of a carinal trifurcation, tracheomalacia, and an isolated partial anomalous pulmonary venous return with an intact atrial septum has not yet been reported in the literature. In this article, we report the diagnostic approach and management of a seven-year-old girl with a delayed diagnosis and management of a coexisting carinal trifurcation, tracheomalacia, and an isolated partial anomalous pulmonary venous return with an intact atrial septum.

Keywords: Carinal trifurcation; isolated partial anomalous pulmonary venous return; tracheomalacia.

ÖZ

Karinal trifurkasyon, sağ üst lobun ya da aksesuar lobun trakeal bronkusuyla karınaya yakın olarak açılması olarak tanımlanır. Yaygın olarak, ortak embriyonik gelişimlerinden kaynaklanan kardiyak anomalilerle görüldüklerinden, bu malformasyonların etyolojisi tam olarak anlaşılamamıştır. İntakt atriyal septumlu izole parsiyel pulmoner venöz dönüş anomalisi bir veya daha çok pulmoner venin, venöz sirkülasyonla bağlantılı olduğu bir doğuştan anomalidir. Karinal trifurkasyon, trakeomalazi ve intakt atriyal septumlu izole parsiyel pulmoner venöz dönüş anomalisi birlikteliği bildiğimiz kadarıyla literatürde bildirilmemiştir. Bu yazıda tanı ve tedavisinde geç kalınan karinal trifurkasyon, trakeomalazi ve intakt atriyal septumlu izole parsiyel pulmoner venöz dönüş anomalisi birlikteliği olan yedi yaşında bir kız çocuğunun tanınal yaklaşımları ve tedavisi sunuldu.

Anahtar sözcükler: Karinal trifurkasyon; izole parsiyel pulmoner venöz dönüş anomalisi; trakeomalazi.

Tracheal bronchus is a rare congenital malformation with an incidence rate of only 0.1-2% on the right side and 0.3-1% on the left. Although displaced and supernumerary tracheal bronchi can be found, the displaced type is more common. Carinal trifurcation, a very rare subtype, is defined as an opening of the right upper lobe or accessory lobe via the tracheal bronchus close to the carina. Additionally, the coincidental

nature of tracheomalacia and carinal trifurcation has provoked much interest.^[1-3]

Isolated partial anomalous pulmonary venous return (PAPVR) with an intact atrial septum is an anomaly in which one or more of the pulmonary veins are connected to the venous circulation, but this only occurs in approximately 0.4-0.7% of the general population.^[4]



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To the best of our knowledge, the coexistence of carinal trifurcation, tracheomalacia, and an isolated PAPVR with an intact atrial septum has never been reported. Herein, we present a patient in which there was a delay in both the diagnosis and management of coexisting carinal trifurcation, tracheomalacia, and isolated PAPVR with an intact atrial septum.

CASE REPORT

A seven-year-old girl was admitted to the hospital with complaints of fatigue and shortness of breath on exertion. There was nothing in her medical records or family history that indicated her ultimate diagnosis, and she was in good general condition, conscious, and cooperative during her preliminary examination. Her body temperature was 36.7 °C, her pulse rate was 90 beats per minute, her finger oxygen saturation (OS) was 95%, her blood pressure was 115/70 mmHg, and her respiratory rate was 14 breaths per minute. A physical examination revealed a normal respiratory system and a grade II/VI midsystolic ejection murmur that was heard best along the left sternal border. The patient's chest X-ray and electrocardiogram were normal, but two dimensional (2D) echocardiography revealed mildly dilated right-sided structures of the heart, moderate tricuspid valve regurgitation, and an intact interatrial septum. Therefore, transesophageal echocardiography (TEE) was performed to better visualize the interatrial septum, and this determined that there were no other cardiac anomalies.

The patient was referred to the Department of Pediatric Pulmonology to investigate which

pulmonary diseases might be contributing to the mildly dilated right-sided structures of the heart and moderate tricuspid valve regurgitation. In her physical examination, the respiratory system was normal, and before salbutamol treatment the pulmonary function test results showed a forced vital capacity (FVC) of 83% and a forced expiratory volume in one second (FEV₁) of 76%. After salbutamol treatment FVC was measured as 86% and FEV₁ as 91%. As FEV₁ increases by more than 12%, it is indicative of reversible airway disease. In addition, reversibility was observed, but thoracic computed tomography (CT) was still performed to identify the differential diagnosis of parenchymal lung disease that stemmed from an absence of the typical clinical findings associated with asthma. A tracheal anomaly was suspected, and we expected to find a tracheal bronchus on the thoracic CT (Figures 1a, b). A fiberoptic bronchoscopy was then performed which found a third bronchial entrance (carinal trifurcation) at the right side of the carina that continued into the accessory lobe. The right and left main bronchi orifices had lost their contours and were shaped like a fish mouth (tracheomalacia) (Figures 2a, b). Since the airway anomaly did not account for the dilated right-sided structures of the heart and moderate tricuspid valve regurgitation, cardiac catheterization was performed to confirm the presence and severity of pulmonary hypertension (HT) and congenital heart disease. Blood gas samples were also taken from the accessed sites, and the OS levels from the superior vena cava (SVC), main pulmonary artery (MPA), and left ventricle (LV) were 84%, 88%,

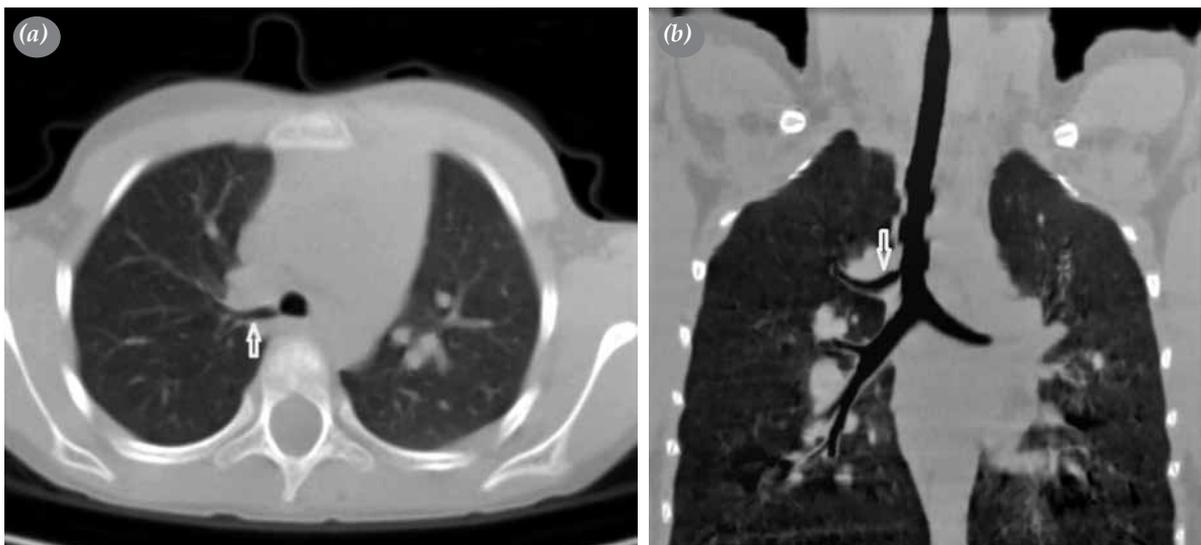


Figure 1. (a, b) View of the tracheal bronchus via thorax computed tomography.

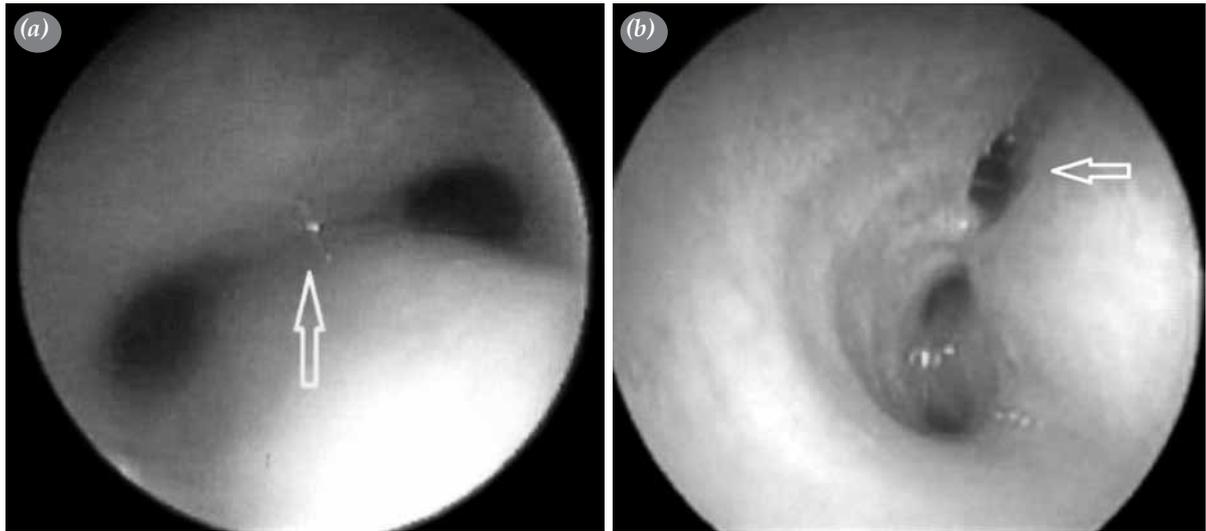


Figure 2. (a) Tracheobronchomalacia as seen via a fiberoptic bronchoscopy. (b) The view from the fiberoptic bronchoscopy on the right shows the third bronchus entrance next to the carina (carinal trifurcation).

and 95%, respectively. Furthermore, there was a significant left to right shunt with a Qp/Qs ratio of 1.5:1. Imaging of a non-ionic contrast medium in the pulmonary arteries identified that the left pulmonary veins had drained into the left atrium while the right pulmonary veins had drained into the right atrium. It also revealed that there was an intact interatrial septum (Figure 3). This case was debated



Figure 3. Angiographic view of an isolated partial anomalous pulmonary venous return.

by the cardiovascular surgery council, which decide that an operation was the best treatment option. Additionally, medical drug therapy was initiated for the patient's congestive heart failure.

DISCUSSION

A tracheal bronchus is an extremely rare congenital malformation that typically occurs in less than 2% of the population.^[3] In fact, Akoğlu et al.^[5] reported a tracheal bronchus incidence rate of only 0.2% in Turkey. Initially defined by Sandifort in 1785, a tracheal bronchus originates from the trachea as an upper right bronchus.^[1] It is usually located in a tracheal segment 2 cm above the main carina but has been reported as high as 6 cm above this ridge.^[2,3,6] As previously stated, there are two types of tracheal bronchus: displaced and supernumerary. If the anatomic upper lobe bronchus or its single branch is missing, the tracheal bronchus is classified as being displaced, which may indicate that the entire upper lobe is displaced or that it is only displaced at its apical segment (high apical lobe). If the right upper lobe bronchus has normal trifurcation into the apical, posterior, and anterior segmental bronchi, the tracheal bronchus is categorized as being supernumerary, and these may end blindly.^[2] Ghaye et al.^[2] examined 35 cases and reported that 77% had ectopic bronchi and 23% had accessory bronchi (supernumerary). During the fiberoptic bronchoscopy procedure there was a third bronchial entrance (carinal trifurcation) at the right side of the carina which continued with the accessory lobe.

A tracheal bronchus is generally asymptomatic and is often incidentally detected. However, it can sometimes be accompanied by laryngomalacia, tracheomalacia, tracheal stenosis, infantile lobar emphysema (especially in the upper lobe), pulmonary sequestration, congenital heart disease, congenital diaphragm hernia, Down syndrome (DS), bronchiectasis, atelectasis, and cystic adenomatoid malformation.^[6,7] Together with vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities (VACTERL) anomalies, tracheobronchial stenosis and tracheobronchomalacia anomalies may also be present.^[6-8] Sarin^[8] reported that tracheal trifurcation and a supernumerary bronchus ended up in a blind manner in a newborn and indicated that the tracheobronchial anatomy must be evaluated via a bronchoscopy in patients with esophageal atresia. In our patient, the tracheomalacia accompanied the carinal trifurcation and accessory lobe.

Tracheobronchial anomalies frequently occur alongside cardiac anomalies because of their common embryonic development. Likewise, in our case, the PAPVR was associated with carinal trifurcation. In addition, the embryonic connections between the pulmonary and systemic veins may persist and result in pulmonary venous drainage abnormalities.^[9] In the literature, there have been five cases with both a tracheal bronchus and PAPVR,^[10] but to the best of our knowledge, our patient represents the first case in which carinal trifurcation, tracheomalacia, and PAPVR appear together.

Tracheal bronchus treatments vary according to the severity of the symptoms. While conservative treatment is sufficient in most cases, segment excisions may be required in the presence of persistent and recurrent upper lobe pneumonia, atelectasis, and air trapping.^[2,11,12] Furthermore, even patients who have no complaints related to an active respiratory passage should continue to be followed up. However, tracheal bronchus patients are usually asymptomatic.^[13,14] Moreover, in a study that involved 101 cases, Butler et al.^[13] found that 71% (n=101) of those who underwent a tracheoplasty also had cardiovascular anomalies, primarily pulmonary artery slings and ventricular septal defects (VSDs).

In conclusion, we believe that our patient represents the first case in which carinal trifurcation, tracheomalacia, and PAPVR have occurred concomitantly. There have been other reports that presented instances of carinal trifurcation and tracheal bronchus occurring together, and one case

report even had a case of carinal trifurcation with PAPVR, but no tracheomalacia was present.^[9,13] In this study, we chose to emphasize that airway anomalies may be encountered without any complaints of airway disease and that cardiac abnormalities should be investigated in the presence of pulmonary abnormalities due to their embryological connections.

Declaration of conflicting interests

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