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Case report / Olgu Sunumu

Concomitant occurrence of tracheal bronchus, pulmonary sequestration and azygos lobe

Trakeal bronkus, pulmoner sekestrasyon ve azigos lobunun eş zamanlı birlikteliği

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A 53-year-old man was referred with the complaints of persistent cough ongoing for approximately two years with intervals, weakness and dyspnea on effort. Posteroanterior lung radiography showed normal lung parenchyma, except azygos fissure and lobe view. Fiber optic bronchoscopy demonstrated tracheal bronchus on the right lateral wall of trachea, 2.5-3 cm above the main carina. Thoracic computed tomography revealed a view in soft-tissue density located in the left lung posterior basal segment, sized 4x2.5 cm, which had an aberrant arterial supply originating from thoracic aorta and was consistent with intralobar pulmonary sequestration. We present this case with a literature review, as we did not find any other case having all these three anomalies together in the literature search.

Key words: Azygos fissure; cough; pulmonary sequestration.

Elli üç yaşında erkek olgu, yaklaşık iki yıldır aralıklarla devam eden inatçı öksürük, halsizlik ve efor dispnesi yakınmaları ile sevk edildi. Arka-ön akciğer radyografisinde azigos fissürü ve lob görüntüsü hariç, akciğer parankimi normal izlendi. Fiber optik bronkoskopide trakeanın sağ lateral duvarında, ana karinanın 2.5-3 cm yukarısında trakeal bronkus görüldü. Toraks bilgisayarlı tomografisinde sol akciğer posterior bazal segmentinde torasik aorttan köken alan anormal arteriyel temini olan ve intralober pulmoner sekestrasyon ile uyumlu, 4x2.5 cm çapında yumuşak doku yoğunluğunda bir görüntü izlendi. Bu olgu, literatür araştırmasında bu üç anomalinin birlikte eşlik ettiği başka bir olguya rastlanmadığı için, literatür incelemesi ile birlikte sunuldu.

Anahtar sözcükler: Azigos fissürü; öksürük; pulmoner sekestrasyon.

Tracheal bronchus is a congenital anomaly located 2-6 cm above the carina, with a diameter of between 0.5 to 1.0 cm and a length ranging from 0.6 to 2.0 cm. The average incidence rate is 0.1-2% in the general population. Generally, tracheal bronchus is found on the right side of the trachea and is more common in men.[1] In addition, it is usually asymptomatic and is incidentally identified in patients who are referred with various pulmonary complaints.

Pulmonary sequestrations are non-functional pulmonary tissue masses that are not connected to the tracheobronchial tree and are supplied by an abnormal systemic artery. Like tracheal bronchus, they are generally asymptomatic and incidentally diagnosed.[2]

Azygos lobes are accessory lobes that occur at a rate of less than 1% in the lungs. They are mostly localized in the right lung and are not symptomatic unless an accompanying pathology exists.[3]

Embryologic process: Pulmonary sequestrations result from a developmental disorder within the first four months of intrauterine life. They are formed by segments which separate from the lung bud and develop together with the lung or the esophagus and then advance up to the distal part of the foregut. In newborns, they are observed via autopsy and are accompanied by another anomaly (e.g., diaphragmatic hernia) in 30% of the cases.[4]

Tracheal bronchus is frequently accompanied by other congenital defects such as Down syndrome,

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tracheal stenosis, or bilateral lumbar ribs and is caused by developmental pauses or interactions in the embryonic phase between the 26th day and the fifth week.[4]

Its embryological development is not clear. In the embryonic period, during its migration to join with the superior vena cava, the arch of the azygos vein entraps the apical or posterior portion of the upper lobe and produces a false image as if these segments were a separate lobe. Tracheal bronchus is rarely accompanied by additional pathologies such as pneumothorax, bronchial carcinoma, and vascular anomalies.[5]

However, to our knowledge, the triad of an azygos lobe, tracheal bronchus, and pulmonary sequestration, that we diagnosed in our patient has not been previously reported in the literature. Hence, due to the clinical significance of this case, we present the data related to this rare combination of anomalies.

CASE REPORT

A 53-year-old man presented to the pulmonary diseases outpatient clinic complaining of a persistent cough that had been ongoing for approximately two years (with intervals), weakness, and dyspnea on effort. There was nothing remarkable in either the patient's medical history or that of his family. On physical examination, fine crackles were heard from place to place in the left hemithorax infrascapular region. The pulmonary parenchyma appeared normal, except for an azygos fissure on the lobe as viewed on a posteroanterior lung radiograph, and the respiratory function tests were within normal ranges. A fiber-optic bronchoscopy (FOB) was performed on the patient under local anesthesia and revealed that the larynx and trachea were normal. However, it also showed a tracheal bronchus in the inferior one-third part of the trachea on the right lateral wall 2.5-3 cm above the main carina. The rest of the endobronchial system was normal (Figure 1a). Thoracic computed tomography (CT) verified the presence of the tracheal bronchus. It was approximately 1.8 cm in length and 1 cm in diameter and extended towards the right upper lobe parenchyma (Figure 1b). In addition, an soft-tissue mass was seen in the posterior basal segment of the left which corresponded with the intralobar pulmonary sequestration on thoracic CT angiography. It was 4x2.5 cm in size and was supplied by an aberrant artery originating from the thoracic aorta. (Figure 2a). There were also ectasic alterations in patches in the sequestration area (Figure 2b).

Furthermore, thorax CT detected an azygos lobe and fissure in the right lung apical segment (Figure 3). The patient was given conservative treatment, and resection of the pulmonary sequestration was recommended.

DISCUSSION

Although congenital pulmonary anomalies generally become symptomatic in infancy and early childhood, a part of them remains asymptomatic until adulthood, as was the case with our patient. They are then incidentally identified on radiographs performed for other reasons.

Tracheal bronchus, pulmonary sequestration, and azygos lobes are rare congenital anomalies of the lungs. Although they occur along with various anomalies involving the thoracic cavity, no cases with all three occurring in the same patient have been encountered in

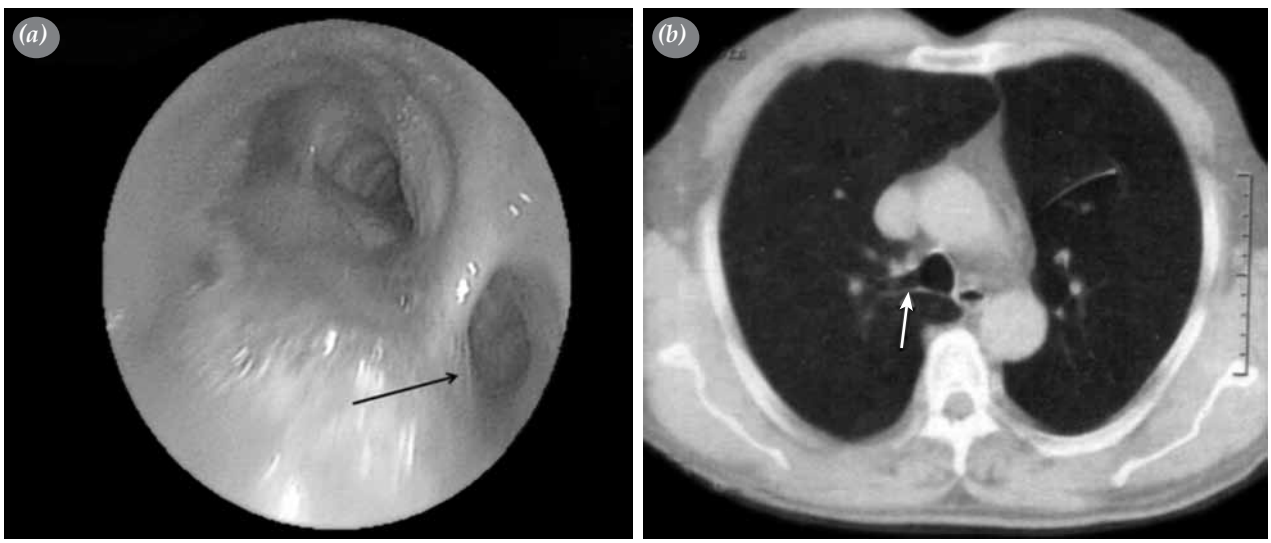

Figure 1. (a) Bronchoscopic views of the tracheal bronchus (arrow) on the right side of the trachea located 2.5 cm away from the main carina. **(b)** A view of the tracheal bronchus (arrow) in computed tomography.

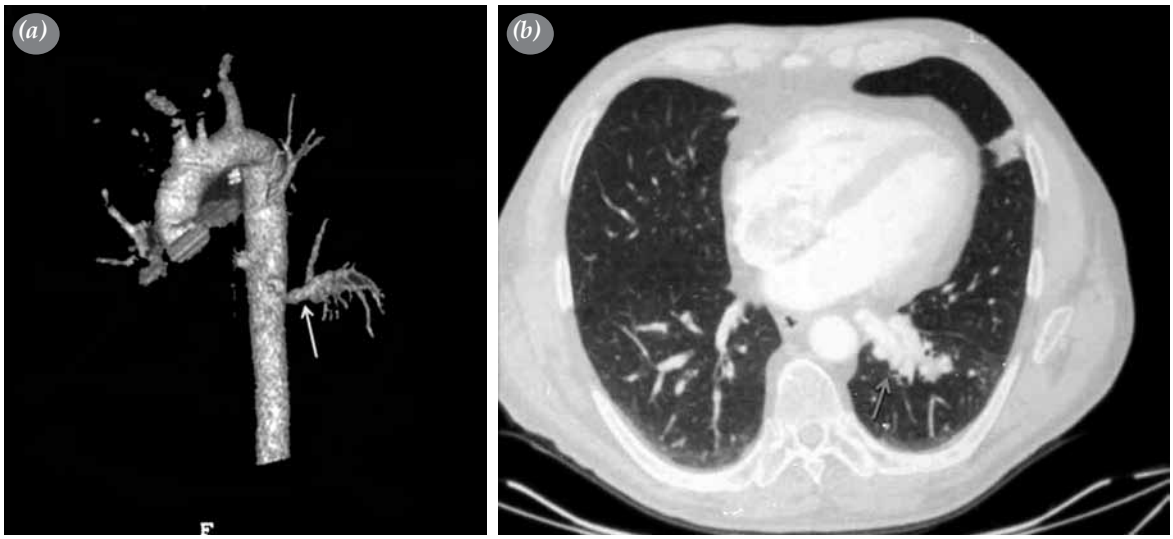

Figure 2. (a) Three-dimensional volume-rendering computed tomography angiography showing the aberrant arterial vascular structure (arrow) originating from the distal thoracic aorta and irrigating the sequestration tissue. **(b)** A view of the sequestration (arrow) in computed tomography angiography.

the previous literature. Our patient administered to the clinic with a persistent cough, and we identified a right tracheal bronchus, a pulmonary sequestration in the left lung inferior lobe, and an azygos lobe in the apical segment of the right lung.

Our patient had a tracheal bronchus originating from the right side of the trachea that was 1.8 cm in length and 1 cm in diameter. It is an anomaly generally located 2-6 cm above the carina and its diameter ranges between 0.5-1.0 cm and its length ranges between 0.6-2.0 cm.[1,6] This anomaly is generally asymptomatic; however, it may lead to recurring pneumonia, chronic bronchitis, and bronchiectasis in patients with bronchial narrowing or less local drainage.[7] Moreover, during intubation, the intubation tube may cause atelectasis by occluding the entrance of the tracheal bronchus or pneumothorax or even by entering the tracheal bronchus itself.[8]

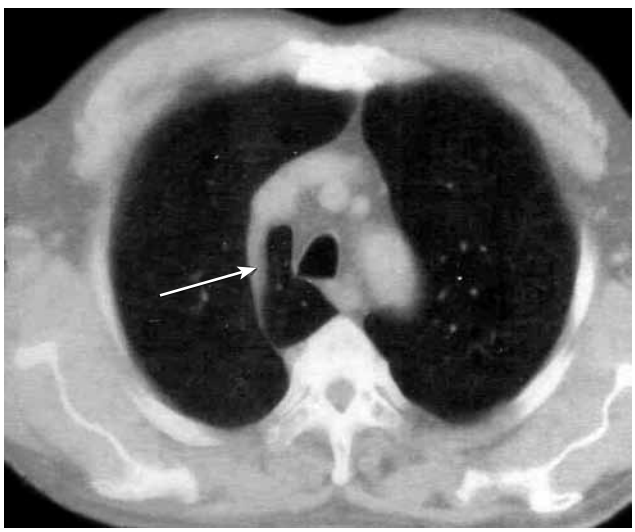

Figure 3. The view of the azygos lobe and fissure seen in thoracic computed tomography.

We also determined that there was sequestration in the left lung inferior lobe. It occurs in the population at a rate of 0.15-1.7% and is more common in men than women.[9] It is generally located in the inferior posterior and medial segments of the left half of the thoracic cavity and is supplied by aberrant arteries generated from the thoracic aorta in 74% of the cases and from the abdominal aorta in 18.7%.[9] Although pulmonary sequestrations are generally asymptomatic, they may lead to recurring infections, bleeding, and tumoral changes.[9,10] Hence, surgical resection is the therapy of choice for most patients. Our patient had a pulmonary sequestration in the left lung posterior basal segment that was supplied by an aberrant artery originating from the thoracic aorta.

Azygos lobes are created by the passage of the azygos vein anterior to the upper lobe of the lung. This process separates the apical or posterior segments in the intrauterine period.[5] Azygos lobes are generally located in the right lung at a rate of less than 1% and are not symptomatic unless an accompanying pathology exists.[3] We identified an azygos lobe in the right lung of our patient.

In conclusion, we identified the triad of tracheal bronchus, pulmonary sequestration, and azygos lobe in a single patient. Each of these anomalies would be rare

in and of themselves, but encountering them altogether was truly unique.

Declaration of conflicting interests

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