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A Case of Incidental Bronchial Atresia

İnsidental Bir Bronşiyal Atrezi Olgusu

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Abstract

Bronchial atresia (BA) is a rare condition that appears radiologically as a perihilar mass lesion and increased peripheral aeration. A 29-year-old asymptomatic male patient was identified with a right perihilar mass during a routine medical examination linked to a job application. Before applying to our clinic, the patient had had undergone non-contrast thoracic CT and PET-CT. Physical examination was normal, while contrast-enhanced thorax tomography revealed a right central mass and signs of air trapped in its periphery. A pulmonary function test was normal. All bronchi were open on bronchoscopy, however a small blunt orifice was observed at the right upper lobe posterior segment entrance. We present this case due to it being a rare condition and the potential for confusion with cancer radiologically. BA with mass image and increased peripheral aeration on thorax CT can be diagnosed based on clinical and radiological findings, allowing unnecessary invasive interventions to be avoided.

Key words: *Mass, air trapping, bronchial atresia.*

Öz

Bronşiyal atrezi nadir görülen bir durumdur. Radyolojik olarak genellikle perihiler kitle lezyonu ve periferal havalanma artışı şeklinde karşımıza çıkar. Yirmi dokuz yaşında asemptomatik erkek hastanın iş başvurusu sırasında yapılan sağlık taramasında çekilen akciğer grafisinde sağ perihiler kitle saptanmış. Kliniğimize başvuru öncesinde hastaya kontrastsız toraks tomografisi (BT) ve PET-CT çekilmiş. Fizik muayenesi normal olan hastaya kontrastlı toraks BT çektik, sağ santral kitle ve periferinde hava hapsi saptadık. Solunum fonksiyon testi normaldi. Bronkoskopide tüm bronşlar açıktı, sağ üst lob posterior segment girişinde küçük kör sonlanan orifis görüldü. Bu olguyu, radyolojik olarak kanser le karışabilmesi ve nadir görülmesi nedeniyle sunmak istedik. Kitle imajı ve periferik hava hapsi ile karakterize olan bronşiyal atreziye klinik ve radyolojik olarak tanı koyulabilir ve gereksiz invazif girişimlerden kaçınılabilir.

Anahtar Sözcükler: *Kitle, hava hapsi, bronşiyal atrezi.*

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Bronchial atresia (BA) is a rare condition caused by focal interruptions of the bronchus that may be lobar, segmental or subsegmental, and usually related to mucus impaction (1). Radiologically, it presents with a central mass and peripheral air trapping (1). The central mass-like image may lead to malignancy being investigated, with potential for exposure to unnecessary invasive procedures. BA is more common in men than women, with an estimated prevalence of 1.2 cases in 100,000. Around two-thirds of patients are asymptomatic (2). We share here a case of bronchial atresia that was detected incidentally from a chest X-ray and investigated with a suspicion of malignancy.

CASE

The posteroanterior chest X-ray taken at another health facility during the routine health screening of a 29-year-old male patient with no smoking history, occupational exposure, comorbidity or symptoms revealed a right perihilar, well-defined, homogeneous opacity approximately 3 cm in size. A subsequent non-contrast thorax computed tomography (CT) scan taken at another health center revealed a 3.5 cm mass and led to him being directed to a 3rd-level health institution. The patient applied to another 3rd-level health institution where a PET-CT was made due to suspicions of cancer, and a SUV-max of 0 was recorded. The patient who had undergone a non-contrast thorax CT scan and a PET-CT then applied to out center for further examination and treatment.

The patient had no cough, sputum, weight loss, night sweats or loss of appetite, but described mild dyspnea occurring with heavy exertion. The patient had a history of frequent bronchitis as a child, but his family history was unremarkable. Upon physical examination, his general condition was good, oxygen saturation was 98% in room air, heart rate was 78/min, body temperature was 36.8 °C, respiratory rate was 13/min and respiratory sounds were normal.

Thorax CT scan and PET-CT images taken in previous health centers were re-examined, and it was decided to perform a thorax CT scan with contrast to rule out any vascular pathology. Following the injection of a contrast material into the left brachial vein to allow the examination of both the arterial and venous phases, a 64-channel multidetector CT was used to obtain arterial phase images (0.6 mm cross-sectional axial plane, coronal and sagittal MIP images, and 3D volume images).

In the central part of the right upper lobe, a 33x25 mm well-defined hypodense lesion was observed that did not show contrast in the obtained pulmonary arterial or late venous phase images. Wide air trapping covering primarily the posterior segment and lateral of the right lung upper lobe, beginning in the area of the lesion, and a mosaic attenuation pattern in the right lung upper lobe apical segment, were noted, (Figure 1 and 2). The 3D volume images are presented in Figure 3. Based on these findings, we made a preliminary diagnosis of bronchial atresia.

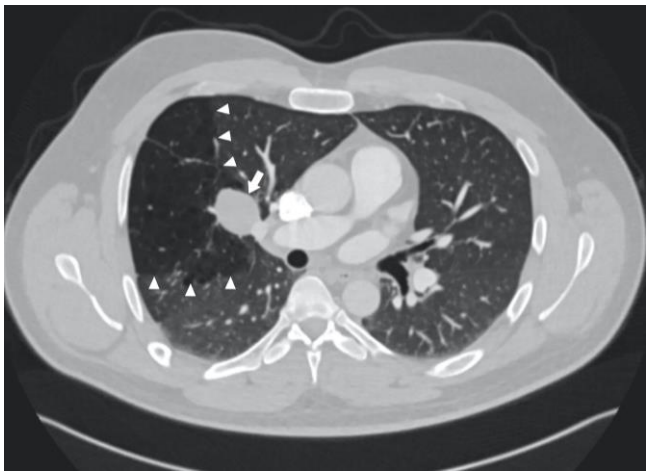


Figure 1: Axial CT image in parenchymal window showing the dilated atretic bronchus plugged with mucus (Arrow) and a radiolucent area due to air trapping and oligemia in the adjacent lung parenchyma (arrowheads)

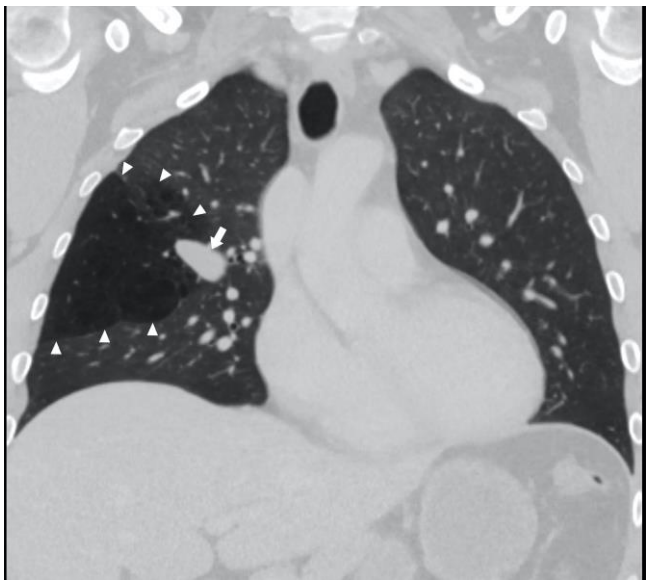


Figure 2: Coronal CT image in a parenchymal window showing the dilated atretic bronchus plugged with mucus (arrow) and a radiolucent area due to air trapping and oligemia in the adjacent lung parenchyma (arrowheads)

A bronchoscopy was performed to support our preliminary diagnosis and to rule out other possible pathologies. The bronchoscopy revealed the mucous membranes to be regular and the right upper lobe segments to be open, however, a narrowed and blind orifice was observed at the posterior segment entrance (Figure 4). The other lobes and segments were noted to have a clear and normal anatomical structure, and a pulmonary function test (PFT) was normal (FEV1/FVC: 83%, FEV1: 3450 ml 112%, FVC: 4100 ml 115%, reversibility test is negative). The patient was diagnosed with BA, and was followed up accordingly.

DISCUSSION

Bronchial atresia (BA) is an uncommon congenital anomaly characterized by focal obliteration of the proximal lumen of a lobar, segmental or subsegmental bronchus, and is related to peripheral mucus accumulation and the related hyperinflation of the obstructed lung segment (1). The most commonly affected areas are the apicoposterior segment of the left upper lobe, followed by the right upper lobe and middle lobe segments (3). Congenital BA was first described in 1953 by Ramsay and Byron (4).

The precise etiology of bronchial atresia is unknown. In a healthy person, during embryonic airway development, lobar bronchi, subsegmental bronchi and distal bronchioles emerge in the 5th, 6th and 16th weeks, respectively. In BA, it is assumed that it occurs as a focal bronchial obstruction before delivery (1). One theory suggests that BA occurs after the 16th week of the embryonic period due to intrauterine ischemia (5,6), while another theory claims that BA develops earlier than the 4th–6th week of intrauterin development, which is a period in which many congenital pulmonary anomalies occur (5-8). Raynor et al. (9), on the other hand, report secondary BA cases caused by the mucosal flap, bronchial mucosal hypertrophy, bronchial wandering related to herniation, or external compression of the bronchi through abnormal vascularization.

BA is usually diagnosed incidentally as patients are generally asymptomatic (1). Some patients may develop such asthma-like symptoms as dyspnea, wheezing and coughing. The most common clinical situation is recurrent lower respiratory tract infection (4,10). BA may be associated with pectus excavatus (11). It has been detected together with spontaneous pneumothorax in a few cases (12). BA, however, is usually an isolated anomaly. Pulmonary function tests (PFTs) are generally normal, but obstructive-type pulmonary dysfunction has been described (13). Our patient's PTF results were normal.

The optimum and most sensitive imaging technique for diagnosis is computed tomography. Mucocele, segmental hyperinflation and hypovascularity are typical features of bronchial atresia, and a combination of these findings on thorax CT is diagnostic (14,15). Radiographic findings include mass-like images and increased peripheral aeration due to collateral air trapping. Hyperlucency is seen in 90% of cases, perihilar masses in 8%, and both in 70% (12,15). Mass like images are caused by mucocele. In our patient's CT, diffuse air trapping in the lateral and posterior segments of the right upper lobe and a well-defined hypodense appearance of 33x25 mm in the central region of the right upper lobe were observed.

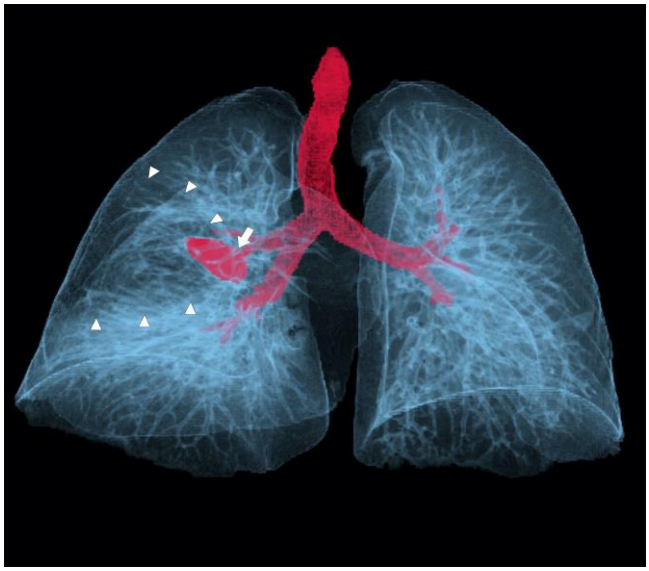


Figure 3: Coronal Colorized Volume Rendered image showing the atretic bronchus (arrow) and air trapped lung parenchyma (arrowheads)

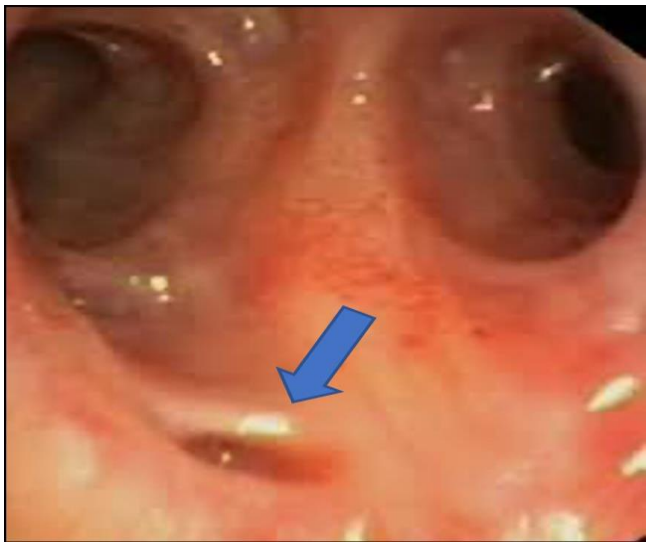


Figure 4: Narrowed blind orifice (blue arrow) at the right upper lobe posterior segment entrance

In patients with BA, generally the main bronchi are open upon bronchoscopy, and no intraluminal occlusive lesion is apparent (14). A bronchoscopic examination in our patient revealed no endobronchial lesion, while a small segment orifice was noted in the upper lobe of the right lung that was not suitable for the normal anatomical structure.

In a differential diagnosis, malignancies, benign neoplasms, allergic bronchopulmonary aspergillosis, arteriovenous malformations, abnormal pulmonary venous return, intralobar pulmonary sequestration, bronchogenic cyst and foreign body aspiration should be considered (13).

The treatment of asymptomatic BA is usually conservative and follow-up should include a chest X-ray. If the patient has severe and recurrent infections, surgical treatment may be considered (16).

There are some limitations related to this case. First of all, thorax CT with contrast should have been performed when a mass-like image was detected on the patient's chest X-ray, as there may have been no need for a PET-CT or a new CT-scan. It should not be forgotten, however, that recognizing BA, which is a rare disease, radiologically requires experience. We were lucky in this regard as the patient had applied to us after being examined by two other health institutions, and so we were able to make a diagnosis without the need for a more invasive procedure than bronchoscopy.

Our patient was diagnosed with BA based on the rudimentary segment orifices observed in bronchoscopy and typical radiological appearances observed in thorax CT. Radiological masses on images should not be assumed to be cancer. BA should be kept in mind in differential diagnosis. Considering BA in cases with mass-like radiological images, but accompanied by increased peripheral aeration and bullae, will save the patient from unnecessary invasive procedures.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - A.B., U.B.; Planning and Design - A.B., U.B.; Supervision - A.B., U.B.; Funding - A.B., U.B.; Materials - A.B., U.B.; Data Collection and/or Processing - A.B., U.B.; Analysis and/or Interpretation - A.B., U.B.; Literature Review - A.B., U.B.; Writing - A.B.; Critical Review - A.B., U.B.

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