

Bronchial Atresia Presented with a Cystic Lesion in a Child: Case Report

Kistik Lezyonla Görülmüş Olan Bronşiyal Atrezili Bir Çocuk

Şule ÇAĞLAYAN SÖZMEN, MD,^a
Duygu ÖLMEZ, MD,^a
Arzu BABAYİĞİT, MD,^a
Tuba TUNCEL, MD,^a
Oğuz ATEŞ, MD,^b
Nevin UZUNER, MD,^a
Özkan KARAMAN, MD,^a
Handan ÇAKMAKÇI, MD^c

^aDepartments of ^aPediatrics Health and Diseases, ^bPediatric Surgery, ^cDepartment of Radiology, Dokuz Eylül University Faculty of Medicine, İzmir

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Yazışma Adresi/Correspondence:
Duygu ÖLMEZ, MD
Dokuz Eylül University Faculty of Medicine, Department of Pediatrics Health and Diseases, Allergy Disciplines, İzmir, TÜRKİYE/TURKEY
duyguolmez@hotmail.com

ABSTRACT Congenital bronchial atresia is an infrequent pulmonary anomaly usually characterized with a blindly terminating bronchus, mucoid impaction, and hyperinflation of the peripheral pulmonary parenchyma. It is usually diagnosed incidentally in asymptomatic young adults males but is rarely diagnosed in children. Pectus excavatum is the most frequent deformity of the thorax. However, bronchial atresia together with pectus excavatum is a rare condition. Costisternal retraction to overcome the obstruction in bronchial atresia can lead to hyperinflated lung parts which can be a cause of pectus excavatum. Herein we presented a six years old girl with congenital bronchial atresia presented with a cystic lesion on thorax computerized tomography and pectus excavatum, which are rarely defined together in the literature. Nuss procedure was performed for correction of pectus excavatum. Right middle and superior lobectomy was successfully performed for bronchial atresia.

Key Words: Bronchial diseases, abnormalities, pectus excavatum, child

ÖZET Konjenital bronşiyal atrezi genel olarak; kör sonlanan bir bronş, mukus tıkaçı ve periferik akciğer parankiminin aşırı havalanması ile karakterize olan, sık görülmeyen bir akciğer anomalisidir. Genellikle asemptomatik erişkin erkeklerde rastlantısal olarak ortaya çıkar, çocuklarda ise nadiren görülür. Pektus ekskavatum toraksın en sık görülen deformitesidir. Ancak bronşiyal atrezi ve pektus ekskavatum birlikteliği nadir bir durumdur. Bronşiyal atrezideki obstrüksiyonu yenmek için gelişen kostokondral retraksiyon akciğer bölümlerinde aşırı havalanmaya, bu da pektus ekskavatumu neden olabilir. Bu çalışmada literatürde birlikteliği nadiren tanımlanmış olan toraks bilgisayarlı tomografide kistik lezyon olarak görülen konjenital bronşiyal atrezili ve pektus ekskavatumlu altı yaşında bir kız çocuğunu sunduk. Pektus ekskavatumun düzeltilmesi için Nuss prosedürü uygulandı. Bronşiyal atrezi için başarılı bir şekilde sağ orta ve üst lobektomi yapıldı.

Anahtar Kelimeler: Bronş hastalıkları, anomaliler, pektus ekskavatum, çocuk

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Congenital bronchial atresia usually presents incidentally in asymptomatic young adult males, but is rarely diagnosed in children.¹ It is a short and juxtahilar bronchus interruption that leads to an obstructive emphysema often associated with mucoid impaction.² Herein, we report a symptomatic child who has both bronchial atresia presented with a cystic lesion on thorax computed tomography (CT) and pectus excavatum. This association was rarely reported in the literature.³

CASE REPORT

Six years old girl admitted to our hospital because of recurrent respiratory tract infections and thorax deformity. Her recurrent respiratory symptoms began at nine months of age. She was given antibiotics several times for lower respiratory tract infections. When she was three years old, right middle lobe emphysema and 2 x 3 x 3cm cystic lesion containing fluid-air was detected on CT of thorax. All laboratory investigations including serology of cyst hydatid were in normal limits. She was followed up with the diagnosis of bronchogenic cyst and lobar emphysema in different hospitals. Her most common complaints were cough and sputum production. It was learned that sternal depression was present shortly after birth and became more prominent in time. She was referred to Pediatric Surgery Department of our hospital for lobectomy and correction of the thorax deformity. Physical examination revealed rib cage rigidity, protracted shoulders, inferior rib flares and sternal depression. Chest X-ray revealed a hyperinflated right lung. Thorax CT was consistent with right superior and middle lobe atresia (Figure 1). Nuss procedure was applied for correction of pectus excavatum. In addition, right middle and superior lobectomy was performed. Lobectomy specimen contained intraparenchymal cyst lined with bronchial epithelium and a mucoid material in it. Histopathologic examination revealed bronchial atresia. There was no serious post-operative complications.



FIGURE 1: Thorax CT of the patient

DISCUSSION

Congenital bronchial atresia is a rare disorder that develops due to a blind terminating segmental or lobar bronchus, in which mucus accumulates to form a bronchocele, and this causes hypoventilation of the lung.⁴ Clinical features of congenital bronchial atresia in childhood and infancy are totally different from adults. Most common symptoms seen in childhood are fever and cough as a consequence of recurrent pneumonia.¹ Our patient's most common complaints were cough and sputum production. She received antibiotics several times because of recurrent pneumonia. Respiratory distress can be seen in infancy. In our patient respiratory symptoms started when she was nine months old. The disease is predominantly seen in females (59%) and right upper lobe is frequently affected, as in our case. Chest X-ray findings show a broad spectrum from a large cyst to an infiltrative pneumonia. It is mostly defined in subsegmental level.¹

Pectus excavatum is the most common congenital chest wall abnormality, affecting 1 in 400 live births.⁵ Deformity can cause cardiorespiratory dysfunction by compressing the chest wall. It also affects psychological state with unpleasant appearance and leads to psychosomatic disturbances.⁶⁻⁹

Bronchial atresia together with pectus excavatum is a rare condition. Airway obstruction can be the major pathophysiological mechanism underlying this condition. Costosternal retraction, to overcome this obstruction, can lead to hyperinflated lung parts which can be the cause of pectus excavatum.³

Our case did not have a diagnosis until she was six years old although she had persistent respiratory symptoms. We postulated that cystic lesion on thorax CT was not suspected as bronchial atresia and was investigated for infectious diseases. Cystic lesion on thorax CT with a relevant thorax deformity should be considered as bronchial atresia.

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