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Case Report / Olgu Sunumu

Pulmonary Artery Sling: Two Different Clinical Entities

Pulmoner Arter Slingi: İki Farklı Klinik Durum

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ABSTRACT

Pulmonary artery sling is an anomalous origin of the left pulmonary artery from the right pulmonary artery. The most common clinical presentations are respiratory symptoms. On the other hand, some patients may be asymptomatic. This report demonstrated two different clinic entities including symptomatic and asymptomatic disease courses. *The Journal of Pediatric Research 2014;1(2):87-8*

Key Words: Pulmonary artery sling, symptomatic, asymptomatic, infant, echocardiography

ÖZET

Pulmoner arter slingi, sol pulmoner arterin anormal olarak sağ pulmoner arterden ayrılmasıdır. En sık klinik bulgular solunum sistemi semptomlarıdır. Diğer taraftan, bazı hastalar asemptomatik olabilir. Bu makalede semptomatik ve asemptomatik klinik gidişatı içeren iki farklı klinik durum sunulmuştur. *The Journal of Pediatric Research 2014;1(2):87-8*

Anahtar Kelimeler: Pulmoner arter slingi, semptomatik, asemptomatik, infant, ekokardiyografi

Introduction

Pulmonary artery sling is a rare congenital anomaly in which the left pulmonary artery originates from the right pulmonary artery. In this condition, left pulmonary artery encircles the distal trachea and right main stem bronchus as it courses between the trachea and esophagus to reach the hilum of the left lung (1-3). Usually, the clinical outcome of these patients depends on the associated tracheal lesions and complex cardiac anomalies atrial and ventricular septal defects, patent ductus arteriosus, left superior vena cava, and tetralogy of Fallot (4). Respiratory symptoms are the most common clinical presentations during childhood and these patients often require early surgical interventions. Symptomatic newborns and infants with these complex lesions have a high mortality rate without surgical interventions. On the other hand, the prognosis is excellent in asymptomatic patients and no surgical procedure is not required (5). In this report, symptomatic and asymptomatic two patients with pulmonary artery sling were presented.

Case Report

Four year-old (patient 1) and 12-month-old (patient 2) female patients were referred to our clinic due to cardiac murmur and recurrent wheezing attacks, respectively. Their family histories did not revealed any cardiac diseases. Patient 2 had recurrent wheezing and pneumonia attacks from the birth while patient 1 was asymptomatic. The physical examination findings of the patients were normal except for a 3/6 pansystolic murmur on the left sternal border of patient 1. On laboratory, the chest radiograms of two patients were normal. However, the bronchoscopy of patient 2 revealed an external compression to the tracheal lumen (Figure 1a, video 1). On parasternal short-axis view of

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transthoracic echocardiography left pulmonary artery was not visualized on the expected location (Figure 1b). In addition, a perimembranous ventricular septal defect was showed in patient 1. The cardiac magnetic resonance imaging of patient 1 (Figure 1c) and catheterization-derived angiograms of patients (Figure 1d, video 2) showed that the left pulmonary artery originated from the right pulmonary artery and coursed leftward. Patient 1 has been clinically following-up for 6 months without any symptoms while successful left pulmonary artery reconstruction was performed on patient 2. After the surgery she became asymptomatic and during 3 months of follow-up period she had no respiratory symptoms.

Discussion

Pulmonary artery sling is an anomalous origin of the left pulmonary artery from the right pulmonary artery. Then, it courses posteriorly over the right main bronchus near its origin from the trachea, traverses between the trachea and the esophagus and enters the left hilum. Although, it is most frequently diagnosed in symptomatic infants, the condition can be found in asymptomatic children. Pulmonary artery sling results in clinical symptoms in approximately 90% of cases, and most of these cases are diagnosed during infancy in the first year of life (1). This condition usually presents with extrinsic airway obstruction and respiratory symptoms such as dyspnea, wheezing, stridor, cyanosis and apnea. Compression by the sling can also produce obstructive emphysema and/or atelectasis affecting either lung. While symptomatic pulmonary artery sling is almost always fatal if untreated (2), rare asymptomatic cases have been detected in older children and adults (3). The prognosis of pulmonary artery sling is variable depending on presentation. Surgery is the standard of care for all symptomatic patients (2). Recently, Fiore et al. have demonstrated a 79% survival rate in patients undergoing correction via transection of the left pulmonary artery and reimplantation anterior to the trachea (5). Additionally, they suggested that tracheal repair is not always necessary in the presence of pulmonary artery sling; that agenesis of the right lung is not a contraindication to successful complete repair; and that simultaneous correction of intracardiac defects can be safely performed in selected patients (5). In contrast to symptomatic patients, the prognosis for asymptomatic patients is excellent, and surgical intervention is not indicated. Recent patients' reports demonstrated two different clinic entities including symptomatic and asymptomatic disease courses. A successful left pulmonary artery reconstruction was performed on patient 2 and during the 3 months of follow-up period she has been still asymptomatic.

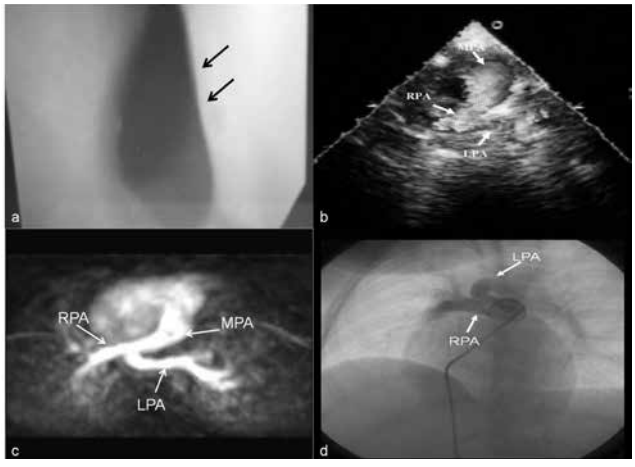

Figure 1. a,b,c,d. (a, video 1) Bronchoscopy of patient 2 showing external compression to the tracheal lumen, (b) parasternal short-axis view of transthoracic echocardiography showing that the left pulmonary artery was not visualized on the expected location in patient 2, (c) cardiac magnetic resonance imaging of patient 1 on axial gadolinium-enhanced image showed that the left pulmonary artery arising from the right pulmonary artery and coursing leftward, (d, video 2 of patient 1) pulmonary artery cineangiogram of patient 2 showed that the left pulmonary artery originated from the right pulmonary artery and coursed leftward. (MPA main pulmonary artery, RPA right pulmonary artery, LPA left pulmonary artery)

Finally, pulmonary artery sling may be presented as symptomatic or asymptomatic. Echocardiography is the primary and helpful technique for diagnosis. Symptoms are more important factors than the diagnosis, when deciding whether to perform tracheal surgery.

Consent

Written informed consent was obtained from the patients' parents for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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