Case Report

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TANAFFOS

Pulmonary Artery Sling Associated with Stridor from Early Infancy: A Case Report

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Background: Pulmonary artery sling is a rare condition in which the left pulmonary artery anomalously originates from a normally positioned right pulmonary artery. The left pulmonary artery arises anterior to the right main bronchus, courses between the trachea and esophagus then enters the left hilum. Respiratory symptoms such as wheezing, stridor, cough, and dysphasia are common in this anomaly.

Case presentation: We describe a 16-month-old male infant presenting recurrent cough, stridor, and wheezing from early infancy. He underwent computed tomography angiography, bronchoscopy, and transthoracic echocardiography, confirming the left pulmonary artery sling diagnosis. Surgical correction of pulmonary artery sling was successfully performed as a new anastomosis between the main pulmonary artery and the left pulmonary artery, as well as tracheoplasty. The infant was discharged without any complications. Follow-up after two years revealed no respiratory symptoms and feeding difficulty.

Conclusion: In the presence of chronic cough, stridor, recurrent wheezing, and other prolonged respiratory symptoms, investigation for possible detection of pulmonary artery sling is recommended.

Key words: Pulmonary Artery Sling; Stridor; Wheezing; Surgical Repair

INTRODUCTION

Pulmonary artery sling (PAS) is an uncommon congenital vascular anomaly in which the left pulmonary artery is separated from the right pulmonary artery (1). It travels between the esophagus and the trachea before entering the left lung, so it creates a compressive effect on these organs (2). The PAS presents tracheal symptoms such as stridor, wheezing, recurrent pneumonia, and respiratory distress in the 1st year of life (3). However, the pulmonary sling cannot be diagnosed until adulthood or remain as asthma (4).

Croup is another cause of upper airway obstruction with stridor and recurrent cough. Other differential diagnoses of respiratory distress included epiglottitis, tracheomalacia, bacterial tracheitis, foreign body aspiration, peritonsillar abscess, and allergic reaction (5). Early diagnosis and surgical treatment are necessary to prevent mortality in patients with pulmonary artery slings (3). Variable modalities are available for diagnosis of PAS, such as chest x-ray (CXR), computed tomography (CT) angiography, cardiac magnetic resonance (CMR), and echocardiography (6).

CT-angiography is a highly sensitive technique for PAS diagnosis, evaluation, and choosing the best surgical method (3). Other congenital heart defects may be associated with PAS, such as patent ductus arteriosus, which may play a role in enclosing the trachea (3). Herein, we report a case of pulmonary artery sling that was presented with chronic wheezing and stridor.

CASE SUMMARIES

A sixteen-month-old male infant was admitted with a history of recurrent stridor, cough, and wheezing from early infancy in Modarres Hospital, Tehran. His past medical history determined that he had been hospitalized three times during the last year due to respiratory distress. On physical examination, his weight was 10 Kg (<5th percentile) with subcostal retraction, nasal flaring, and no heart murmur. Sepsis workup was done (except lumbar puncture because of unstable condition). Complete blood count (CBC) with differential was normal. White blood cell count (WBC) of 10500 cells/µl, neutrophils of 60% and negative C-reactive protein (CRP) were reported that all of them were unremarkable.

Chest radiography showed right-sided infiltration (Figure 1). Transthoracic echocardiography demonstrated that the left pulmonary artery (LPA) was originating from a normally positioned right pulmonary artery (RPA) (Figure 2). Moreover, CT angiography revealed an anomalous beginning of the LPA as separated from the posterior aspect of RPA and coursed between the trachea and esophagus, causing the trachea compression (Figure 3). A bronchoscopy was performed. Multiple complete rings from the thoracic inlet to one cm above the carina (approximately eight cartilage) led to the significant tracheal narrowing at three last rings (>50% obstruction, Grade 2) and severe bronchomalacia of the right main bronchus (RMB) and left upper lobe bronchus (LUL) were reported in bronchoscopy (Figure 4).

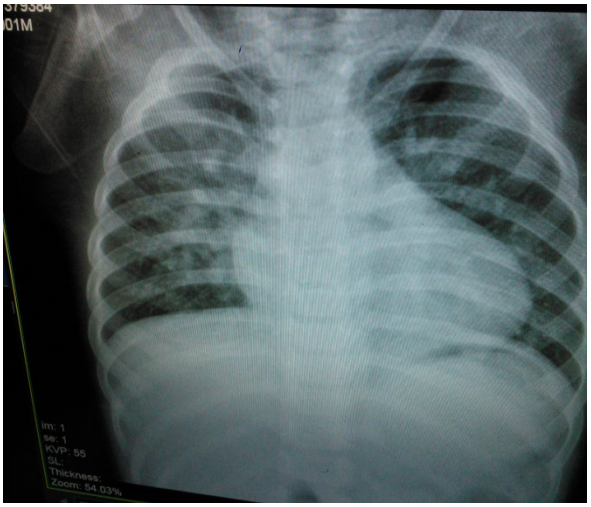

Figure1. CXR revealing right-sided infiltration.

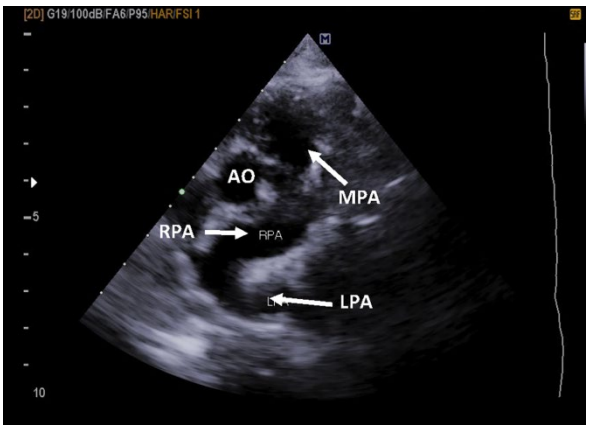

Figure 2. Echocardiography showing the abnormal origin of LPA from RPA.
MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; AO, aorta.

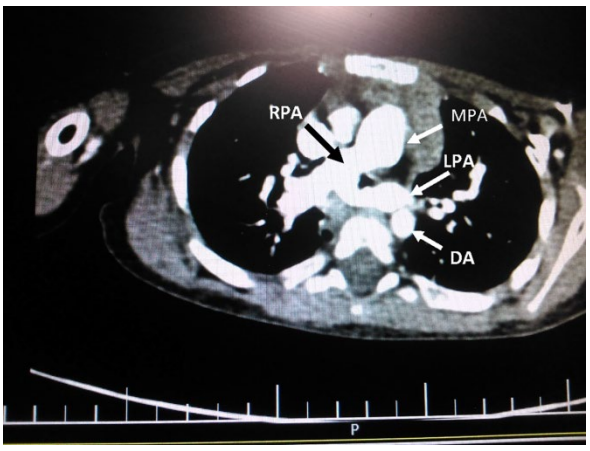

Figure 3. CT angiography showing abnormal arising of LPA from RPA.
MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; DA, descending aorta.

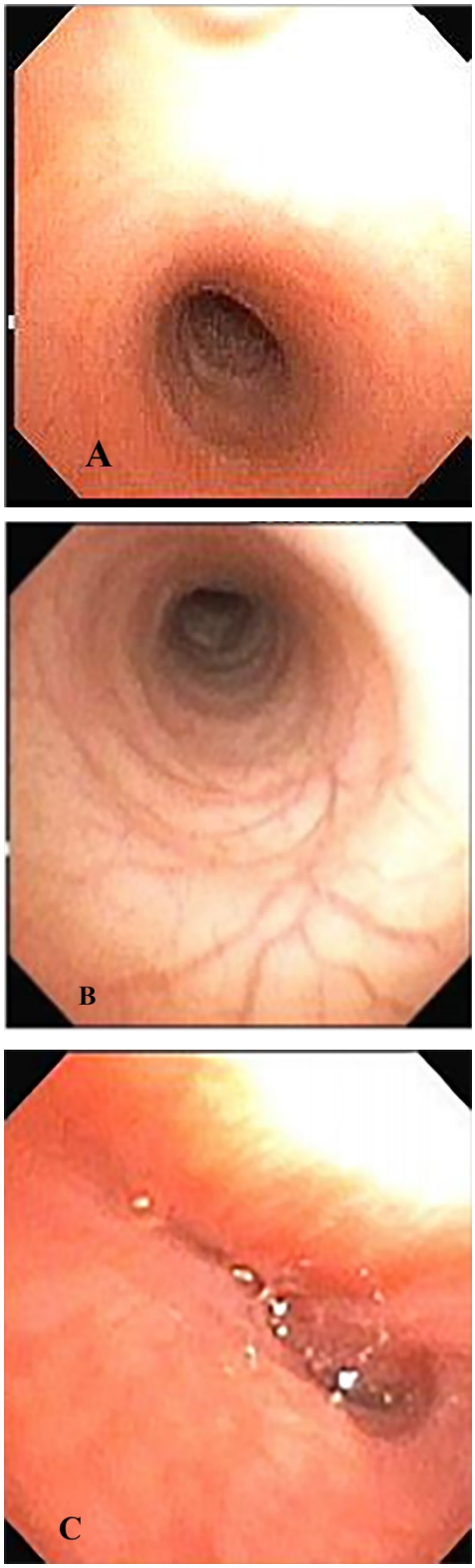

Figure 4. Bronchoscopy showing complete tracheal rings with significant narrowing (A, B). Severe bronchomalacia of RMB and LUL (C).

RMB, right main bronchus; LUL, left upper lobe bronchus.

The patient underwent surgical correction of PAS. A median sternotomy was performed. After heparin administration, canulation of aorta and atria followed by cardiopulmonary bypass was done. On a beating heart, the LPA was detached and repositioned to the main pulmonary artery in the proper anatomical location (Figure 5). Given severe tracheomalacia, tracheoplasty was performed. After surgery, all respiratory symptoms were eliminated and the patient was discharged ten days after surgery in a good condition. Regular follow-up was preserved and during two-year follow-up, improvement of symptoms with no complications was reported.

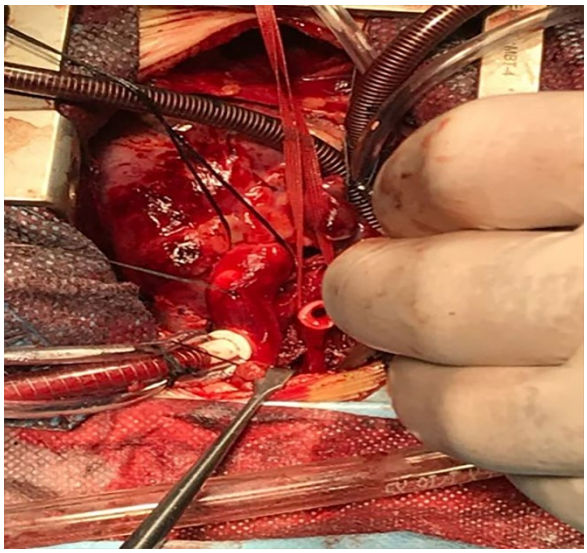

Figure 5. Surgical repair of pulmonary artery sling

DISCUSSION

Pulmonary artery sling is a rare condition of respiratory distress with non-specific symptoms such as cough, stridor, wheezing, and dysphasia (7). The prevalence of PAS has not been recognized in the United States and globally (8). In some studies, PAS might be asymptomatic until late childhood (9). Undiagnosed PAS result in morbidity and mortality. Therefore, timely diagnosis is very important. In our case, the patient has respiratory symptoms in early infancy. After three times hospitalization and missed diagnosis of asthma, echocardiography was performed with suspicion of pulmonary artery sling, and PAS was established after CT

angiography. Pulmonary artery sling can be diagnosed by fetal echocardiography using a three-vessel view (10). Although echocardiography can be a non-invasive technique for screening PAS, it is not competent to confirm this diagnosis because of low sensitivity (11). The CT angiography is a helpful modality not only for PAS diagnosis but also for pre-operation evaluation and the position of trachea compression (8).

Bronchoscopy is another choice modality for the diagnosis of pulmonary artery sling. The term "stovepipe trachea" is used in PAS in which the tracheal cartilage is circular, which should normally be U-shaped (7). In the present study, tracheal cartilage was similar in bronchoscopy. Also, tracheal obstruction due to narrowing caused by complete rings led to respiratory symptoms such as stridor and wheezing. The optimal treatment for pulmonary artery sling is surgical repair. Sternotomy via midline incision followed by cardiopulmonary bypass, detachment of LPA, and anastomosis between LPA, and main pulmonary artery are preferred (like the procedure performed in our patient) (7). Tracheoplasty should be considered in symptomatic tracheal stenosis, similar to the current case. The clinical outcome after surgery depends on coexisting other cardiac malformations and tracheal anomalies. In our patient, a two-year postoperative follow-up demonstrated that all respiratory symptoms were removed and the best condition was achieved for the patient.

In conclusion, a pulmonary artery sling can cause death in early infancy. Thus, in the presence of chronic cough, stridor, recurrent wheezing, and other prolonged respiratory symptoms, investigation for possible detection of pulmonary artery sling is recommended. Early surgical repair can reduce morbidity and mortality.

Ethical Approval

Informed consent was provided for the purpose of publication of images and other clinical information in this case report. Also, in this paper, no identifiable personal details are included.

Conflict of Interest

The authors have no conflicts of interest to declare.

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