ABSTRACT

Pulmonary artery sling is a rare congenital vascular anomaly that results in the abnormal development of the left pulmonary artery. This case report describes a one-year-old male patient with a history of respiratory issues who was initially treated for suspected infantile asthma. However, further investigation revealed a diagnosis of pulmonary artery sling, a rare congenital vascular anomaly that can cause airway and esophageal compression. The patient underwent surgical treatment to correct the abnormality. This case emphasizes the importance of considering pulmonary artery sling as a potential diagnosis in patients with respiratory symptoms and highlights the need for prompt diagnosis and treatment to prevent potential complications.

Keywords: Congenital vascular anomaly, pulmonary artery sling, respiratory symptoms.

Pulmonary artery (PA) sling is a rare congenital vascular anomaly that involves an incomplete formation of the sixth pair of aortic arcs during embryogenesis. This results in the abnormal development of the left PA, which originates from the posterior wall of the right PA. The aberrant artery passes over the right main bronchus and in front of the trachea or carina and the esophagus before reaching the left lung hilus. This condition can compress the trachea or esophagus, leading to abnormal development of the tracheobronchial tree and airway obstruction.

Respiratory symptoms, such as dyspnea, stridor, and wheezing, typically manifest in the first years of life, and the degree of airway deformation is inversely related to the child’s age. Younger children are at higher risk of airway cartilage deformation. If left untreated, tracheal stenosis associated with PA sling can increase the risk of mortality. In this case report, we present a patient who was initially diagnosed with asthma but was later diagnosed with PA sling and underwent surgical treatment.

CASE REPORT

A one-year-old male patient who had a history of frequent hospitalizations due to reactive airway disease and recurring lower respiratory tract infections since the patient was 40 days old presented to our hospital's emergency unit with complaints of cough and fever. On physical examination, we detected prolonged expiration, wheezing, inspiratory difficulty, and stridor. A chest X-ray showed pericardiac infiltration, and the patient was diagnosed with a lower respiratory tract infection. A 15-day course of antibiotic therapy and inhaler drugs was initiated, but the patient continued to experience coughing and inspiratory stridor, and thorax computed tomography revealed bilateral air confinement in the posterobasal region. Suspecting infantile asthma, inhaler treatment was continued, and the patient was discharged on the 15th day.

However, the patient returned to the hospital after a week with a recurring cough, and bronchoscopy revealed a narrowing in the posterior lateral wall at the entrance of the right main bronchus due to compression. Further thorax computed tomography angiography showed that the left PA originated from...
the right PA and wrapped around the right arch of the carina, creating pressure on the airway and esophagus (Figure 1). The patient was diagnosed with PA sling and recommended surgical treatment.

During the procedure, the patient was closely monitored with an electrocardiogram, blood oxygen saturation, invasive blood pressure, Bispectral Index (Medtronic, Minneapolis, MN, USA), and near-infrared spectroscopy (INVOS; Medtronic, Minneapolis, MN, USA). The blood pressure was 106/68 mmHg, the heart rate was 135/min, and the peripheral oxygen saturation was 98%. Midazolam was intravenously administered, and anesthesia was induced with sevoflurane inhalation. Intravenous fentanyl and rocuronium were administered, and the patient was intubated. We also performed central venous catheterization.

After the median sternotomy, we partially removed the thymus and opened the pericardium. We observed that the aberrant left PA originated perpendicular to the hilus at one-third of the right PA. As it emerged, it passed between the posterior of the trachea and the anterior of the esophagus and oriented towards the left hilus, creating an incomplete vascular sling. It was noted that it partially compressed the trachea and the right main bronchus. We released the aberrant left PA by dissection. Afterward, under normothermic cardiopulmonary bypass (CPB) and aortic cross-clamping, the left PA was transected from the right PA where it originated and passed through the trachea and esophagus to the left side. The left PA was anastomosed with the continuous technique end-to-side to its new place in the main PA (Figure 2). The patient successfully came out of CPB after 60 min without any complications.

The patient was transferred to the intensive care unit, where he remained intubated and sedated. The following day, the patient was extubated, and the patient was discharged on the sixth day after the procedure.

**DISCUSSION**

Pulmonary artery sling is a rare form of a vascular ring that accounts for only 1% of congenital cardiovascular anomalies.[1] Pulmonary artery sling can cause respiratory symptoms in children and is often misdiagnosed as asthma due to respiratory system-related findings.[1,3] It has been reported that early surgical intervention reduces postoperative respiratory complications.[6] Therefore, early diagnosis and appropriate surgical intervention are crucial for the management of PA sling. Due to varying

![Figure 1. (a) Bronchoscopy image showing compression on the posterior lateral wall of the right main bronchus. (b) Computed tomography image showing pulmonary artery sling.](image-url)
degrees of respiratory distress in these children, airway management during surgery should be approached with caution. Vascular rings can act as anterior mediastinal masses during induction, leading to tracheal compression. It is recommended to use inhalation induction and to administer muscle relaxants only after ventilation is assured.[7]

Respiratory functions should be closely monitored since airway obstruction will continue after the operation.[3] Prolonged intubation, prolonged mechanical ventilation, and reintubations may be encountered in the postoperative period due to tracheomalacia. A study by Hong et al.[5] reported that postoperative early extubation, continuous positive airway pressure application, and early extubation might be useful to try again, even if reintubation was performed.

Early surgical intervention is recommended to prevent more severe complications that may arise as a result of long-term vascular compression of the airways.[6] In this type of vascular anomaly that requires surgical treatment, dissection at the point where the left PA originates and reimplantation to the main PA in front of the trachea are needed.[2] According to Backer et al.,[8] operations performed using median sternotomy and CPB allowed for the anastomosis of the left PA to the main PA in a safe and bloodless surgical area. The study found no complications associated with CPB, and reoperation was unnecessary.

In conclusion, when this extremely rare condition is diagnosed, early surgical intervention is necessary to prevent airway damage. Bronchoscopy should be performed to evaluate the airway condition before

Figure 2. (a) View of the left PA originating from the right PA. (b) Absence of the left PA. (c) Anastomosis of the left PA to its anatomical position by separating it from the right PA. (d) Final view of the anastomosis of the left PA.

PA: Pulmonary artery.
surgery. Inhalation induction is recommended, and early extubation should be considered in the postoperative period.

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