

A Rare Cause of Mechanical Airway Obstruction: Aberrant Left Pulmonary Artery Also Named Vascular Sling

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Abstract

Aberrant left pulmonary artery also known as pulmonary sling, is a rare congenital vascular malformation, that is defined by anomalous course of this artery, passing behind the trachea, leading to tracheal stenosis, manifesting by respiratory symptoms, among new born and infants, the overall prevalence is estimated to occur one in every 17,000 school aged children, It originates from a lack of development of the sixth aortic arch during embryogenesis, the physiologic development of the left pulmonary artery consist in connecting it to the main pulmonary artery, anterior to the trachea, clinically it manifests generally by respiratory symptoms such as: dyspnea, wheezing, stridor, the dyspnea may be severe and hence life threatening, the diagnosis is generally made by Ct scan, the treatment is surgical, by repositioning the artery in front of the trachea, with high mortality procedure.

Keywords: Aberrant Left Pulmonary Artery; Pulmonary Artery Sling; Anomalous Left Pulmonary Artery; Airway Compression Imaging

Introduction

Aberrant left pulmonary artery is a rare anatomical variant, belongs to a broad category of congenital vascular anomaly, characterized by anomalous course of this artery, originating from the right pulmonary artery, and passing above the main right bronchus, and between esophagus and trachea, then it achieves the left lung [1].

The prevalence of pulmonary artery sling is estimated to be 59 per million, with a male predominance [2].

It manifests in neonatal period, by coughing, wheezing stridor, and severe dyspnea, which can be life threatening [3], due to compression of the trachea [3], the physiopathology consists in lack of development of the 6th aortic arch, physiologically the left pulmonary artery is linked to main pulmonary, anterior to the trachea, in our case, the left pulmonary artery migrates from the right pulmonary artery, resulting in a longer course, passing between esophagus and trachea [4].

The diagnosis is made by Ct scan, and bronchoscopy may be required for evaluation of the tracheal stenosis extent [4], 30% of patients have concomitant cardiac malformations, like ventricular septal defect, tetralogy of Fallot, atrial septal defect, that can be treated during PAS repair, hence the interest of making pre-operative cardiac screening [5].

The treatment is based on surgical repair, consisting in re-implantation of the aberrant artery, pulmonary artery sling associated with stenosed trachea is characterized by poor post-operative prognosis [4].

Case Presentation

We report the case of an infant aged one year, with no significant past medical history, who presented with severe respiratory distress requiring admission to the intensive care unit. Initial bronchoscopy evaluation revealed tracheal stenosis without evidence of an endoluminal lesion.

Contrast-enhanced computed tomography (CT) demonstrated an aberrant left pulmonary artery originating from the right pulmonary artery. The vessel followed a characteristic course between the trachea and the esophagus, passing just above the carina, which was located at the T5 vertebral level. This anomalous course resulted in extrinsic compression of the airway, particularly involving the distal trachea and the right main bronchus.

Quantitative assessment showed tracheal narrowing at the level of the carina measuring 6 × 2 mm (transverse × anteroposterior diameters). The anteroposterior diameter of the right main bronchus was markedly reduced to 1 mm, compared to 2 mm for the left main bronchus.

Based on imaging findings, the diagnosis of a type I aberrant left pulmonary artery (pulmonary artery sling) was retained, given the normal position of the carina and the absence of associated tracheobronchial anomalies. In particular, no bridging bronchus or abnormal bronchial branching pattern was identified.

Furthermore, no associated cardiovascular malformations were detected, including atrial septal defect, ventricular septal defect, or tetralogy of Fallot. The CT scan also revealed partial atelectasis of the left lower lobe.

Discussion

Pulmonary artery sling is a rare congenital vascular anomaly arising from abnormal development of the aortic arch system, in which the left pulmonary artery courses between the trachea and the esophagus, resulting in airway compression and potential tracheal stenosis [6].

It is a relatively uncommon congenital malformation, with an estimated prevalence of 59 per million and a reported male predominance [7]. However, this contrasts with our observation, as the present case involves a female patient.

The pathophysiology of pulmonary artery sling is well established and is best explained by the “space-available” embryologic theory. According to this concept, developing foregut and mesodermal structures—including the lung bud, bronchial buds, and the sixth aortic arch—compete for limited space. Premature branching of the right bronchus may create additional space adjacent to the trachea [8], allowing the left pulmonary artery to migrate abnormally and connect with the right ventral sixth aortic arch, thereby forming the vascular sling [9]. This mechanism also accounts for the frequent association of pulmonary artery sling with other congenital anomalies, such as patent ductus arteriosus [9]. In our case, however, no associated congenital malformations, including patent ductus arteriosus, were identified.

Pulmonary artery sling is symptomatic in approximately 90% of cases and is most often diagnosed during the first year of life. Clinical manifestations result from extrinsic airway compression and include respiratory distress, dyspnea, wheezing, stridor, and cyanosis. In addition, vascular compression may lead to pulmonary complications such as atelectasis involving one or both lungs [10]. These findings are consistent with our case, in which the patient presented with severe respiratory distress characterized by dyspnea, wheezing, and cyanosis, unresponsive to non-invasive ventilation and ultimately requiring orotracheal intubation.

Congenital tracheal stenosis is associated with pulmonary artery sling in approximately 65% of cases [11], a finding that aligns with the tracheal narrowing observed in our patient. Bronchoscopy is often performed as an initial diagnostic modality to exclude alternative causes of airway obstruction, such as foreign bodies or subglottic stenosis, and to assess for complete tracheal rings. Although

echocardiography has limited utility in directly diagnosing pulmonary artery sling due to its restricted field of view, it remains valuable for identifying associated cardiovascular anomalies and evaluating vascular rings [12].

Definitive diagnosis is generally established upon contrast-enhanced thoracic computed tomography (CT), which is considered the gold standard. CT imaging enables precise visualization of the anomalous origin and course of the left pulmonary artery, assessment of airway compression and tracheal stenosis, classification of the anomaly, and detection of associated pulmonary complications such as atelectasis [13]. In our case, bronchoscopy revealed tracheal stenosis without endoluminal obstruction, prompting CT evaluation, which confirmed the diagnosis of pulmonary artery sling and demonstrated associated left lower lobe atelectasis. Based on the normal position of the carina and preserved tracheobronchial anatomy, a type I pulmonary artery sling was diagnosed.

Airway anomalies commonly associated with pulmonary artery sling include tracheal and right main bronchial compression, complete tracheal rings, and tracheomalacia [15]. In our patient, only focal tracheal stenosis at the level of the carina was identified. Cardiovascular malformations are reported in up to 85% of cases and include ventricular septal defect, atrial septal defect, and patent ductus arteriosus [13], with tetralogy of Fallot being among the most frequently described lesions [16]. However, no associated cardiovascular anomalies were detected in our case.

The classification of pulmonary artery sling is based on tracheobronchial anatomy and the level of the carina. Two main types are recognized. Type I corresponds to a normal tracheobronchial branching pattern, with the carina located at the T4-T5 level, and may include subtypes depending on the presence or absence of minor airway anomalies. Type II, which is more frequently reported in the literature [14], is characterized by abnormal airway anatomy, a low-lying carina (T6-T7), long-segment tracheobronchial stenosis, and associated anomalies such as bridging bronchus and a T-shaped carina [12].

The main differential diagnosis is unilateral pulmonary artery agenesis, in which one pulmonary artery is absent and the contralateral artery represents the sole branch of the main pulmonary trunk [12].

Management is primarily surgical and consists of reimplantation of the left pulmonary artery anterior to the trachea with anastomosis to the main pulmonary artery [17]. However, in cases of significant or irreversible tracheal stenosis, airway intervention may be required. Tracheoplasty is indicated in patients with severe narrowing, with a tracheal diameter of less than 3 mm considered a critical threshold associated with poorer outcomes. Conversely, a larger tracheal diameter is generally associated with a more favorable prognosis [18].

Despite advances in surgical techniques and improved outcomes following pulmonary artery reimplantation, infants requiring concomitant tracheoplasty for long-segment tracheal stenosis or those with complex congenital cardiac anomalies continue to pose significant therapeutic challenges [19].



Figure 1: Axial Contrast enhanced thorax Ct scan displaying the aberrant left pulmonary artery, originating from the right pulmonary artery (Blue arrow), and coursing between the esophagus and the trachea, the carina is stenosed by this aberrant trajectory (Green arrow).

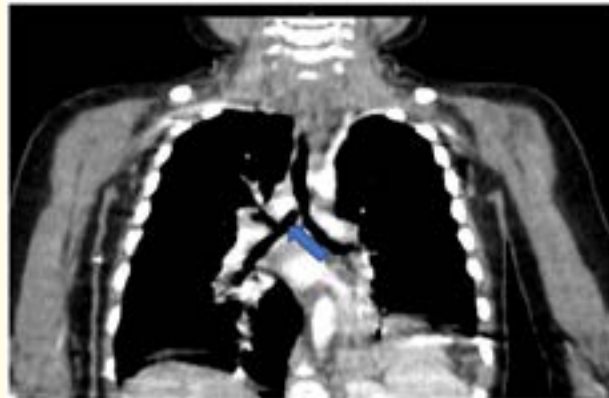


Figure 2: Coronal enhanced thorax Ct scan displaying aberrant left pulmonary artery compressing the carina and right bronchus (Blue arrow).

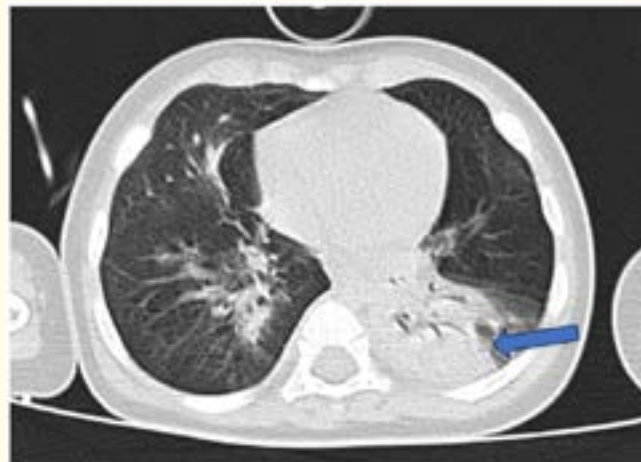


Figure 3: Axial thorax Ct scan, parenchyma window; displaying a passive atelectasis of posterior segment of the lower left lobe (Blue arrow).

Conclusion

Aberrant left pulmonary artery (pulmonary artery sling) is a rare congenital vascular anomaly that typically presents in early infancy with respiratory distress due to airway compression. Its clinical severity depends mainly on the degree of associated tracheobronchial involvement.

Contrast-enhanced CT is the diagnostic modality of choice, allowing precise assessment of the vascular anomaly, airway compromise, and associated abnormalities. Classification based on airway anatomy is essential for prognosis and management.

Treatment is primarily surgical, with generally favorable outcomes; however, associated tracheal stenosis or congenital anomalies may complicate management. Early diagnosis and a multidisciplinary approach are key to improving patient outcomes.

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