MEDICAL FORUM

LEFT PULMONARY ARTERY SLING: REPORT OF FIVE CASES ON MDCT FROM VIETNAMESE CHILDREN

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SUMMARY

Left pulmonary artery sling (LPAS) is a rare congenital anomaly in which the left pulmonary artery originates from the posterior aspect of the right pulmonary artery and courses between the trachea and esophagus to reach the left lung. This anomaly causes distal tracheal and/or right main-stem bronchus compression. Most LPAS cases are associated with early symptom onset, around 2 month-old, and have severe respiratory distress within the first year of life. There are two major types of LPAS based on the location of LPA and abnormal bronchial branching. The diagnosis can be made by using various imaging modalities. Herein, we present the imaging characteristics on multi detectors computed tomography of 5 LPAS cases with respiratory distress (2 months to 12 months).

Keywords: *pulmonary sling, computed tomography, respiratory distress.*

INTRODUCTION

Left pulmonary artery sling (LPAS) is a rare congenital anomaly (4% of congenital vascular anomalies) in which the LPA originates from the posterior aspect of the right pulmonary artery and courses between the trachea and esophagus to reach the left lung. Approximately 50% of LPAS cases are associated with intrinsic tracheal stenosis. There are two major types of LPAS based on the origination of LPA and the congenital anomalies of the tracheobronchial tree. Type I is defined by a normal tracheobronchial pattern, with the tracheal bifurcation at the fourth to fifth thoracic vertebral level. Type I slings are further divided into type IA and IB delineated by the presence (A) or absence (B) of a pre-eparterial tracheal bronchus. Type II slings are subdivided into types IIA and IIB, which are both characterized by an abnormal bridging bronchus and a tracheal bifurcation at thoracic vertebral level 6 [1]. Both types of slings, but especially type II, are associated with other cardiovascular, pulmonary, and other abnormalities [2].

We report 5 cases (2 boys) of LPAS, presented with dyspnea, aged 2-12 months, underwent Multiple Detector Computed Tomography at Radiology center, Bach Mai hospital.

CASE REPORT

Case 1: Type IA (Figure 1)

2-month-old girl, with a long history of wheezing, stridor. On MDCT, LPA originates from the right pulmonary artery (D4 level) and courses between the trachea and esophagus, compressing the distal of the trachea. The anatomy of the tracheobronchial tree is normal. She has also a perimembranous ventricular septal defect.

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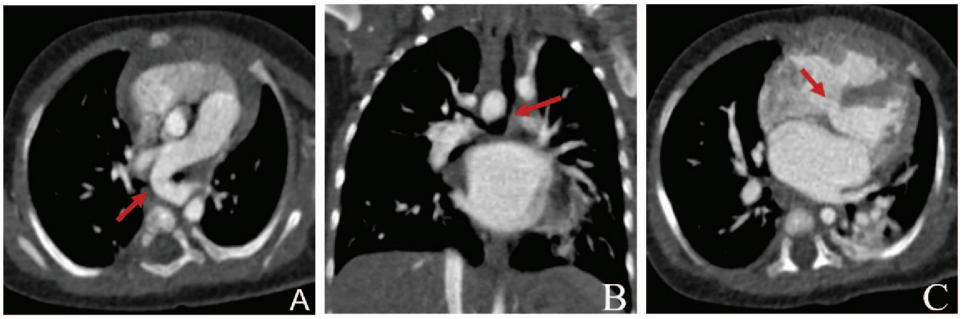

Figure 1: A, B: LPA originates from the right pulmonary artery (D4 level) and courses between the trachea and esophagus, compress the distal of the trachea. The anatomy of the tracheobronchial tree is normal. C: Note also perimembranous ventricular septal defect.

Case 2: Type IA (Figure 2)

12-month-old boy, presented with wheezing, dyspnea for 1 month. LPA originates from the right pulmonary artery (D5 level) and courses between the trachea and esophagus, compressing the distal of the trachea. The anatomy of the tracheobronchial tree is normal. The patient has also a right-sided aortic arch.

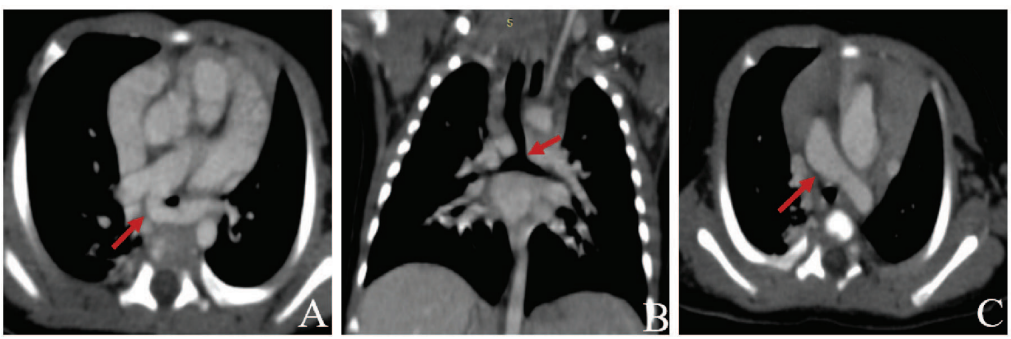

Figure 2: A, B: LPA originates from the right pulmonary artery (D5 level) and courses between the trachea and esophagus, compress the distal of the trachea. The anatomy of the tracheobronchial tree is normal. C: Note also right-sided aortic arch.

Case 3: Type IA (Figure 3)

6-month-old girl, presented with wheezing, dyspnea from newborn. LPA originates from the right pulmonary artery (D5 level) and courses between the trachea and esophagus, compressing the distal of the trachea. The anatomy of the tracheobronchial tree is normal.

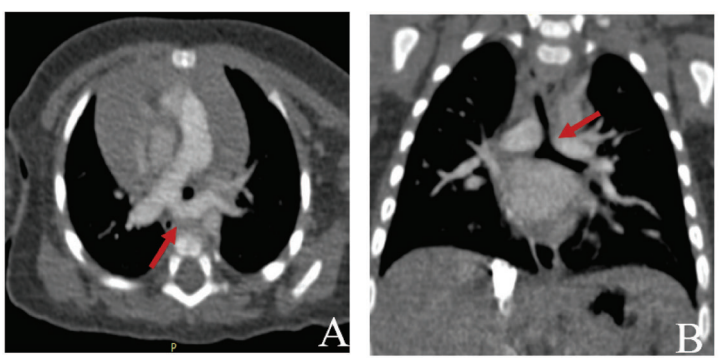

Figure 3: A, B: LPA originates from the right pulmonary artery (D5 level) and courses between the trachea and esophagus, compress the distal of the trachea. The anatomy of the tracheobronchial tree is normal.

Case 4: Type IIB (Figure 4)

9-month-old boy, with a long history of cough, wheezing. LPA originates from the right pulmonary artery (D6 level) and courses between the trachea and esophagus, compress the same level of trachea. MDCT shows a low T-shaped carina (D7 level). In the complete absence of the right bronchial tree, the right lung is being supplied by a bridging bronchus from the left main bronchus. The lungs infection was shown on MSCT.

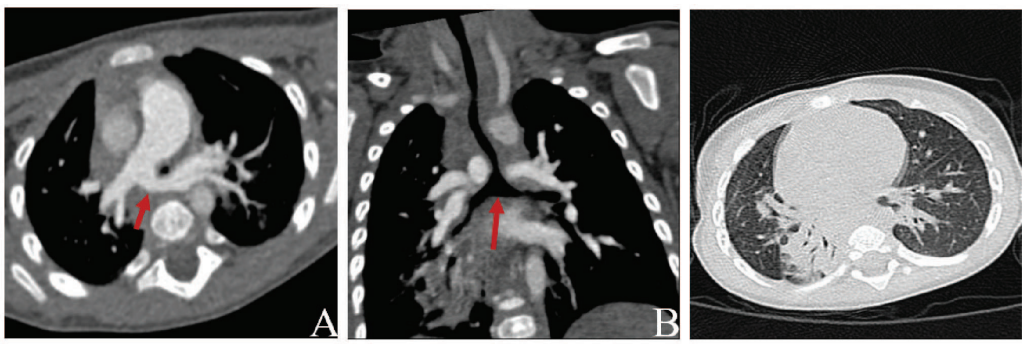

Figure 4: A, B: LPA originates from the right pulmonary artery (D6 level) and courses between the trachea and esophagus, compress the same level of trachea. Low T-shaped carina (D7 level). In the complete absence of the right bronchial tree, the right lung is being supplied by a bridging bronchus from the left main bronchus. C: Lungs infection.

Case 5: Type IA (Figure 5)

12-month-old girl, with a long history of cough, wheezing and recurrent bronchitis. LPA originates from the right pulmonary artery (D5 level) and courses between the trachea and esophagus, compress the same level of trachea. The anatomy of the tracheobronchial tree is normal. Note also hiatal hernia and perimembranous ventricular septal defect.

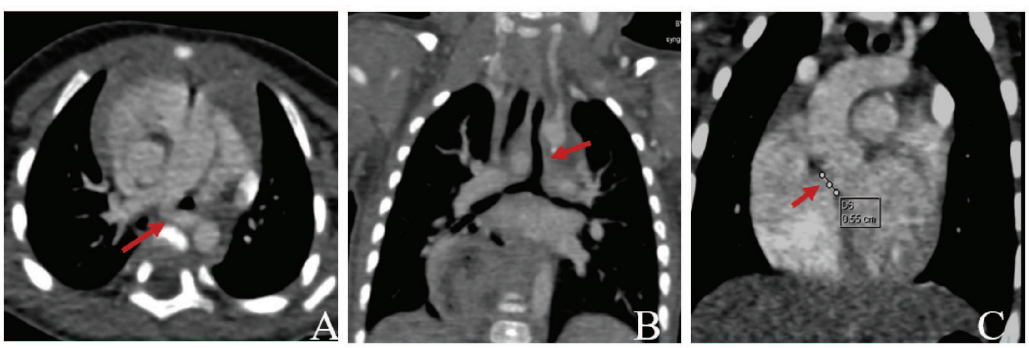

Figure 5: A, B: LPA originates from the right pulmonary artery (D5 level) and courses between the trachea and esophagus, compress the same level of trachea. The anatomy of the tracheobronchial tree is normal. Note also hiatal hernia. C: Perimembranous ventricular septal defect.

DISCUSSION

LPAS was first defined in 1958 by Contro et al and recognized as a rare cause of congenital broncho-obstruction (4% of congenital vascular anomalies) [2]. LPA originates from the posterior aspect of the right pulmonary artery and courses between the trachea and esophagus to reach the left lung. Approximately 50% of LPAS cases are associated with intrinsic tracheal stenosis [3].

The development of the branchial apparatus begins during the second week of gestation and is completed by the seventh week. The aortic arch then gives rise

to several major thoracic arteries. The formation of a pulmonary sling is the result of a failure of the proximal left 6th arch to properly involute. An anastomotic vessel, connecting the primitive pulmonary circulations, becomes the anomalous left pulmonary artery, arising from the right pulmonary artery. This vessel then travels above the main pulmonary bronchus to reach the left lung hilum by passing between the trachea and esophagus, often leading to a compression of these structures [3].

Approximately 50% of LPAS cases are associated with intrinsic tracheal stenosis. The most common presenting symptoms of pulmonary artery sling are wheezing, stridor, and vomiting/feeding difficulties. Cough and recurrent infections are less common presentations. All of these symptoms are very common in children; however, in most cases, they will have other, most commonly infectious, etiologies [4].

There are two major types of LPAS. Type I is defined by a normal tracheobronchial pattern, with the tracheal bifurcation at the fourth to fifth thoracic vertebral level. Type I slings are further divided into type IA and IB delineated by the presence (A) or absence (B) of a pre-eparterial tracheal bronchus. Type I malformations are less complex and are normally associated with tracheobronchomalacia. Symptomatic cases produce significant morbidity and mortality and are usually managed by left pulmonary artery reimplantation surgery. Type II slings are subdivided into types IIA and IIB, which are both characterized by an abnormal bridging bronchus and a tracheal bifurcation at thoracic vertebral level 6. In Type IIA, the bridging bronchus originates from the left main bronchus and supplies the right middle and upper lobe. Type IIB is defined by a complete absence of the right bronchial tree, with the right lung being supplied by a bridging bronchus from the left mainstem bronchus. The right lung is commonly hypoplastic. Type II is more common than type I and is frequently complicated by long segment tracheal stenosis. Management of this type needs to address not only the aberrant pulmonary artery but also the airway abnormality. Both types of slings, but especially type II, are associated with other cardiovascular, pulmonary, and other abnormalities [2].

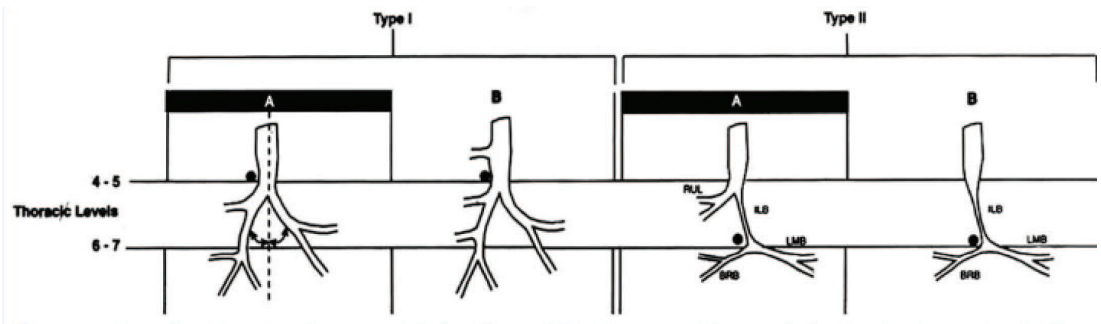

Figure 1. Anatomic types of PA sling (Wells, et al.) – solid circle denotes left pulmonary artery origin. PA: pulmonary artery, RUL: right upper lobe bronchus, ILB: intermediate left bronchus, BRB: bridging right bronchus, LMB: left main bronchus.

Turner et al showed that the median age for the diagnosis of the pulmonary sling is one month with the range being from birth to 3 months [3]. Primary means of diagnosis differ from an institution; however, the emphasis has shifted away from the barium swallow and angiography to CT scanning, MRI, and echocardiography. It has been demonstrated that chest X-ray films are the least sensitive for detecting vascular sling, but they are still the first-line examination modality, occasionally revealing compression of the trachea and/ or mainstem bronchus. The barium swallow and bronchoscopy have better sensitivity than chest X-ray, but not as high as echocardiography, angiography, and CT. Some institutions use echocardiography as the modality of choice, but as with all ultrasonographic studies, the sensitivity is operator-dependent. Echocardiography is a relatively low-cost, non-invasive, radiation-free modality, which may be used as an initial step in

pediatric patients undergoing a workup of vascular anomalies. MRI is a viable means of diagnosing pulmonary slings, with the ability to reveal the aberrant vasculature without relying on contract. However, it is associated with higher costs and requires the infants to be sedated [5].

Asymptomatic patients need no surgical treatment, even when anomalies are found incidentally. Medical management is recommended for infants with mild symptoms. Respiratory distress, history of recurrent pulmonary infections, apneic spells, FIT are indications for surgical. Gikonyo et al reported a 90% mortality rate in patients with a pulmonary sling that was managed only medically.

CONCLUSIONS

Left pulmonary artery sling (LPAS) is a rare congenital anomaly in which the LPA originates from the posterior aspect of the right pulmonary artery and courses between the trachea and esophagus to reach the left lung. There are two major types of LPAS based on the location of LPA and abnormal bronchial branching. Diagnostic imaging, particularly MSCT and MRI identify and assess the level of tracheal stenosis.

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