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Left pulmonary artery sling: why is virtual CT bronchoscopy important? Case report

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Abstract

Background Many causes of vascular rings may present as stridor among children. Co-occurrence of tracheal anomaly in left pulmonary artery sling (LPAS) is very common and needs special attention.

Case presentation We report a case of an infant who presented with expiratory stridor and wheezing. A preliminary chest x-ray revealed no significant findings. CT and virtual bronchoscopy confirmed the diagnosis of LPAS with associated distal tracheal stenosis. He underwent surgical correction with good results in the follow-up period.

Conclusions Virtual bronchoscopy may help to decide the type of operation and to avoid a potentially invasive bronchoscopy in LPAS.

Keywords Left pulmonary artery sling, Tracheobronchial compression, Virtual CT bronchoscopy

Background

A pulmonary artery sling is an uncommon vascular anomaly that can produce severe airway obstruction. The exact prevalence of LPAS is difficult to conclude as the number of asymptomatic cases is unknown [1]. There are hospital-based registries reporting close to 0.14% prevalence for LPAS [2]. Compared with other vascular rings, the tracheobronchial stenosis in LPAS is predominantly anatomical due to congenital tracheal stenosis rather than dynamic or associated with trachea-bronchomalacia [3]. It is a life-threatening condition with an overall mortality of 20–70% within the first month of life, despite surgical correction due to the high occurrence of tracheobronchial and cardiac anomalies [1, 4]. The commonest symptoms are stridor and wheezing [1]. Virtual CT Bronchoscopy is a technique to identify complete tracheal rings with precision and ease.

Case presentation

A four-month-old male infant, first born to healthy non-consanguineous parents after a full-term pregnancy and uneventful delivery, presented at our hospital with sudden onset labored and noisy breathing. He was afebrile, acyanotic, and had no regurgitation during feeding. Physical findings at admission included stridor with prolonged expiratory wheeze.

A chest radiograph revealed a normal cardiothoracic ratio with normal pulmonary vascular markings (Fig. 1a). Echocardiogram (Fig. 1b–d) showed a 7 mm ostium secundum atrial septal defect (OSASD) and anomalous branching of pulmonary arteries which were of adequate size.

Contrast-enhanced ECG-guided cardiac CT was performed in a 256-slice CT scanner (Brilliance iCT, Philips Healthcare, USA) under sedation. A left pulmonary artery sling (LPAS) was present where the left pulmonary artery (LPA) had an aberrant origin from

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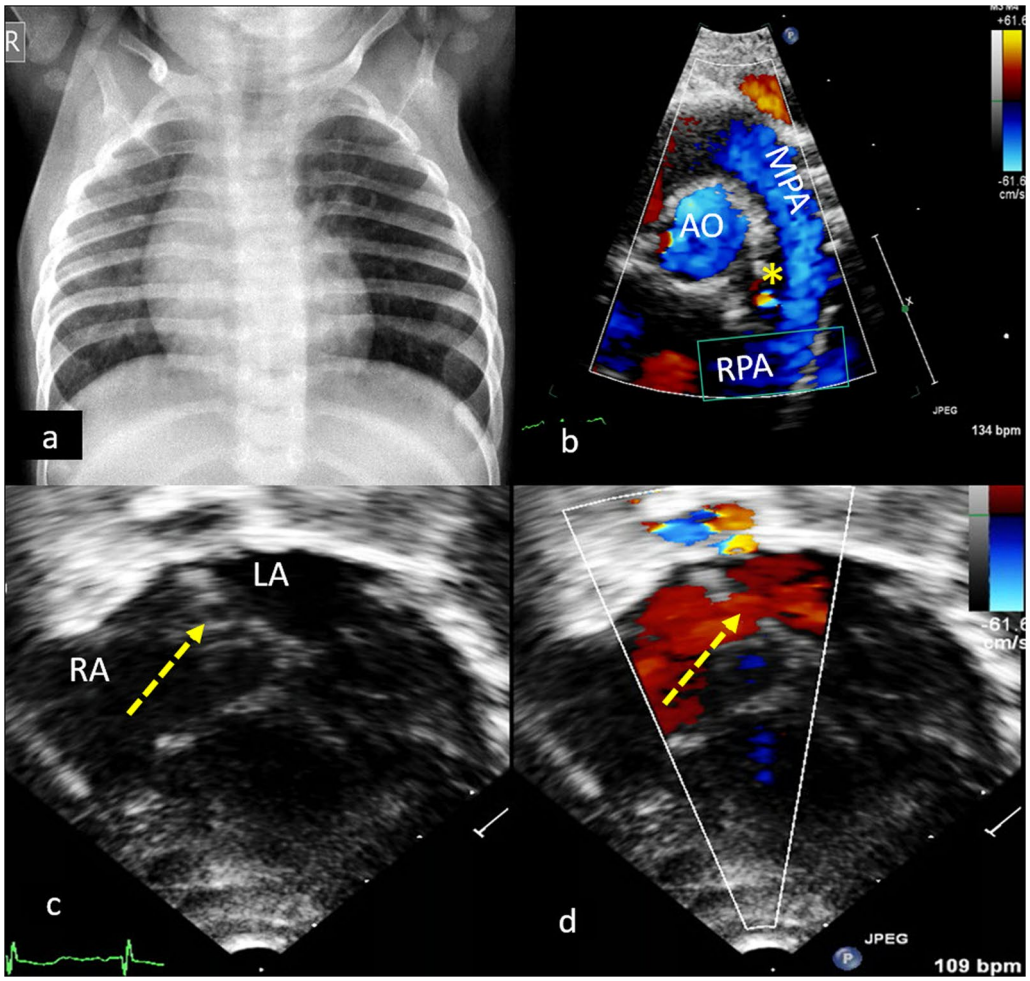

Fig. 1 **a** Chest radiograph revealed prominent right cardiac border possibly due to thymic shadow, with absent aortic shadow at the left cardiac border and normal pulmonary vascularity. **b** Echocardiogram in modified parasternal view demonstrates aberrant origin and anomalous course of LPA (yellow asterisk). **c** and **d** Echocardiogram in color mode in substernal view demonstrating OSASD (yellow dotted arrow) with left to right shunt

the right pulmonary artery (RPA) causing wrapping and compression of the trachea above the carina along with an unusual widening of the carina (Fig. 2a–c). Virtual bronchoscopy revealed short segment tracheal stenosis of 5.5 mm length, involving 4 tracheal rings (Fig. 2c–d and Additional file 1: video 1). The minimum diameter of the stenosed tracheal segment was 2 mm. The tracheal stenosis was treated by tracheal resection and anastomosis. LPA was reimplanted to the left side of the main pulmonary artery (MPA) via a thoracotomy along with direct closure of the ASD.

The hospital course was marked by a gradual improvement in respiratory symptoms. At the time of discharge, the child was asymptomatic. He was asymptomatic at a follow-up 2 years later also (Fig. 3).

Discussion

The embryology of LPAS has not been completely delineated with literature reviews postulating various theories [5, 6]. Abnormal fusion of the left lung bud plexus to the right sixth branchial arch instead of left during the time of vascular supply shift happening at 32 days is one of the mechanisms resulting in the formation of LPAS [6]. There is a tendency for tracheal stenosis in LPAS cases to begin at a higher tracheal level and extend to the carina; not just at the level where the LPA encircles the trachea due to a "squeezing and milking" hypothesis as heart descends along with carina [7]. Prevalence of congenital tracheal stenosis in LPAS is as high as 50–65% in many series and often described as the "ring- sling complex" [8]. Well's classification is

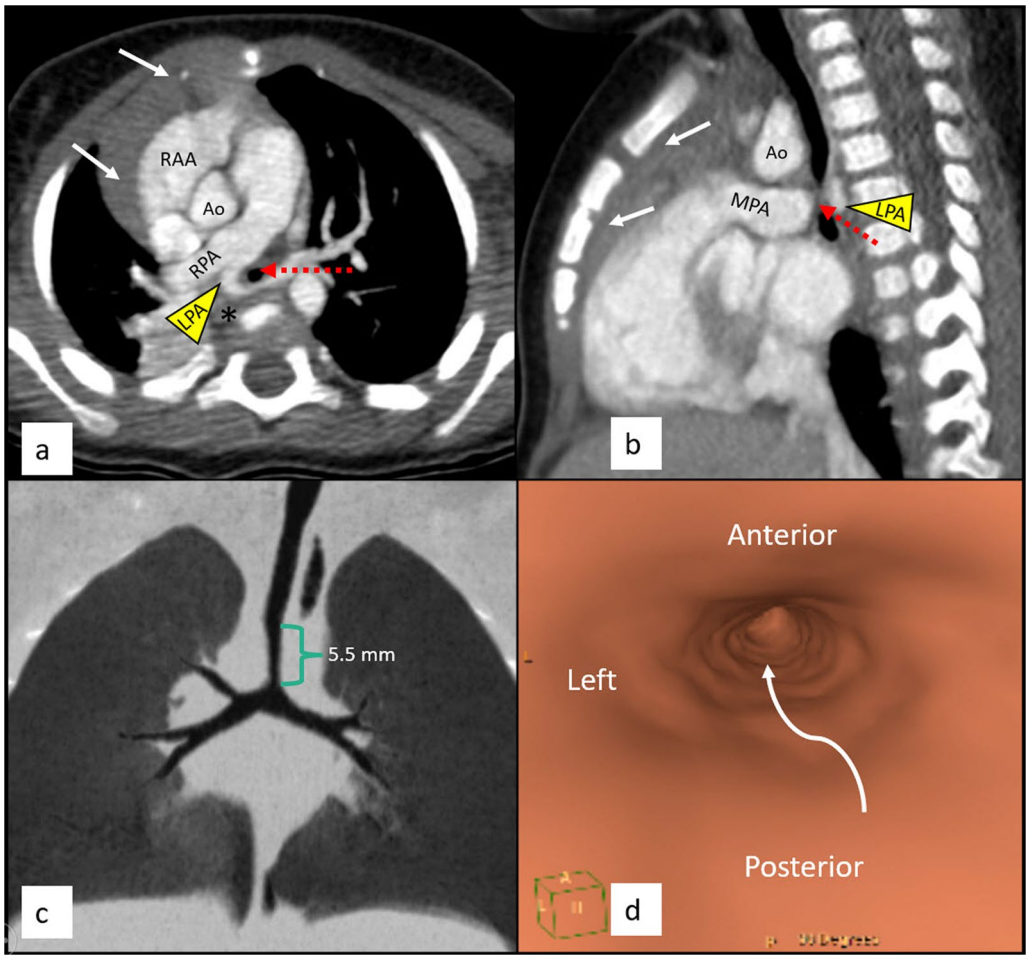

Fig. 2 MIP of CT Angiogram in **a** Axial and **b** Sagittal, demonstrates the posterior tracheal impression and narrowing (red dotted arrow) by the left pulmonary artery (yellow arrowhead in a). Position of esophagus is denoted by *. Enlarged thymus is noted anterior and lateral to the right atrial appendage, in the retrosternal location (white arrows in **a** and **b**) which is responsible for the prominent right heart border in Fig. 1a. **c** Coronal reconstruction demonstrates distal tracheal stenosis (Green Bracket). **d** Virtual bronchoscopy image reveals concave posterior margin and mucosal indentations of complete tracheal rings as ribbings (curved white arrow)

helpful for the management of LPAS where there are two main categories and two subtypes based on level of carina and presence of aberrant right bronchus [9]. This case was Type 1 A variety of LPAS. The complete cartilaginous ring of the trachea is one of the causes of persistent respiratory symptoms after surgical repair, usually detected by bronchoscopy [3].

Virtual CT bronchoscopy is a less invasive technique, usually performed in the non-contrast CT phase using a virtual bronchoscopy module that allows the operator to plan the path from the level of the laryngopharynx to the bronchi. The 3D virtual reconstruction of the airway mucosal surface as the cursor moves through the trachea captures images of any anomalies that may be present along the course, including complete tracheal rings [10]. Virtual bronchoscopy is a minimally invasive procedure that is fast, efficient, and involves less discomfort and procedural risks compared to traditional bronchoscopy. However in presence of mucosal edema, this techniques fails and a rigid bronchoscope is preferred [11].

A reduction tracheoplasty is performed in longer tracheal stenosis and resection anastomosis is performed in short-segment tracheal stenosis [12]. Our case had four complete tracheal rings and hence the latter treatment option was performed. Due to the high incidence of pulmonary arterial hypertension (PAH) in these patients, and left to right shunt should also be corrected for better outcomes as performed in this case [5].

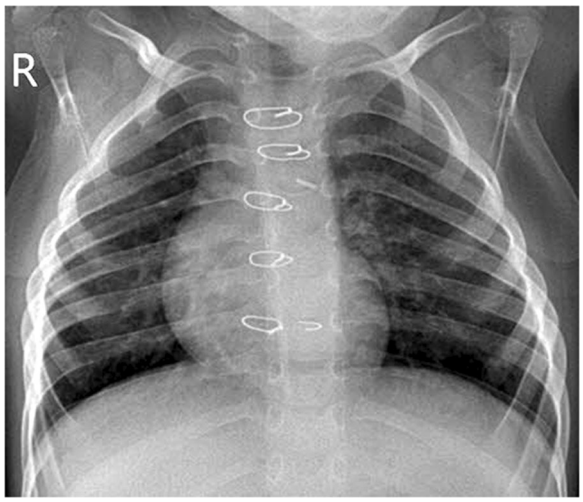

Fig. 3 Post-operative chest radiograph reveals normal bilateral lung fields without any pulmonary or tracheal complications

Conclusions

Successful treatment of LPAS requires early recognition of vascular rings as well as complete tracheal rings by appropriate imaging techniques. Virtual bronchoscopy may help to define degree and extent of tracheal stenosis and hence decide type of operation in LPAS and avoid invasive bronchoscopy.

Abbreviations

3D	3 Dimensional
Ao	Aorta
CT	Computed tomography
LA	Left atrium
LPA	Left pulmonary artery
LPAS	Left pulmonary artery sling
MIP	Maximum intensity projection
MPA	Main pulmonary artery
OSASD	Ostium secundum atrial septal defect
RA	Right atrium
RAA	Right atrial appendage
RPA	Right pulmonary artery
PAH	Pulmonary artery hypertension

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s43055-023-01115-9.

Additional file 1: Video 1. The video of virtual bronchoscopy reveals complete cartilaginous rings in the trachea with distal tracheal stenosis along its course from laryngopharynx to tracheal bifurcation.

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Author AA, KDK contributed in concept, drafting article, critical revision of article and approval of article, whereas JV, AG, BSD contributed in drafting article, critical revision of article and approval of article.

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Availability of data and materials
Not applicable.

Declarations

Ethics approval and consent to participate
Institutional Ethical Approval was obtained.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Competing interests
The authors declare no competing interests.

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