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Letter to the Editor

Aberrant left pulmonary artery – An uncommon cause of persistent stridor and respiratory distress in a neonate

Sir,

Aberrant left pulmonary artery (ALPA) is a rare vascular anomaly first described by Glaevecke and Doehle in 1897.¹ It arises from the right pulmonary artery, passes over the right main stem bronchus, loops to the left and courses between the trachea and esophagus to the left hilus. Respiratory symptoms like dyspnea, stridor or cyanosis result from compression of the trachea and right main stem bronchus by the anomalous vessel.² ALPA is frequently associated with tracheal anomalies, congenital heart disease, and lung abnormalities.³ Surgery is the only therapeutic modality for symptomatic patients, but results are often poor because of associated intracardiac defects and intrinsic tracheobronchial anomalies.⁴ Though the development of noninvasive imaging modalities such as computed tomographic (CT) scan, ultrasonography, and magnetic resonance imaging has led to increased reporting of this disease entity, diagnosis requires a high index of clinical suspicion in combination with careful clinical examination and use of appropriate imaging modalities. We report a neonate with ALPA who presented with persistent stridor and respiratory distress.

A 25-day-old male neonate, weighing 2150 g, presented to the Pediatric Emergency with rapid breathing and difficulty in feeding since birth, noisy breathing since one week of life and prominence of the chest since 2 weeks of age. He was delivered normally to a third gravida mother at hospital after an apparently uneventful antenatal period. There was no history of consanguinity and first two siblings (aged 3 years and 2 years, respectively) were normal and healthy. Birth weight of the infant was 2500 g. He cried immediately after birth but developed rapid and difficult breathing at the age of 6 h which never subsided but increased progressively over time. Since one week of age he developed noisy breathing which used to increase during crying and in supine posture and used to decrease in prone position and when held against the shoulder. The parents noticed prominence of the anterior chest wall since two weeks of age. The infant also had associated feeding difficulty since birth and was never been able to suckle properly. He was fed with diluted cow's milk with cup and spoon. He was shown to multiple practitioners and several

diagnoses were entertained like pneumonia, laryngomalacia, congenital heart disease etc.

On examination, the infant was anxious looking, irritable, having stridor in both phases of respiration and tachypnea (respiratory rate 72/minute) with marked suprasternal, intercostal and subcostal retractions. There was no cyanosis. Color of the face was dusker compared to the body but otherwise the infant had a normal facies without any dysmorphic feature of Pierre Robin sequence (Fig. 1). There was anteroposterior bulging of the chest with shifting of trachea to the left. On auscultation, air entry was poor over both the lung fields, more so on right side. Examination of the cardiovascular system was normal. Pulse oximetry showed an oxygen saturation of 92–94% in all the four limbs in room air. Pre- and post-ductal arterial blood gas analyses were normal. Chest X-ray showed hyperinflated right lung with mediastinal shift and partial collapse of the left lung (Fig. 2). A provisional diagnosis of tension pneumothorax was made and an

Fig. 1 – Clinical photograph of the infant.

Fig. 2 – Chest X-ray showing hyperinflated right lung with mediastinal shift and partial collapse of the left lung.

intercostal tube drainage was placed. But there was no clinical or radiological improvement after 24 h.

Keeping a clinical possibility of congenital lobar emphysema (CLE) or any other mass lesion producing supralaryngeal airway obstruction, a contrast enhancing computerized tomographic (CECT) scan of thorax was advised. CECT showed ALPA originating from the posterior aspect of right pulmonary artery, coursing around the right main stem bronchus and then from right to left, posterior to carina and anterior to esophagus to reach the hilum of left lung (Fig. 3). It compressed the lower trachea and the right main stem bronchus with luminal narrowing and air trapping producing obstructive emphysema of right lung. Esophagus was compressed posteriorly. Significant mediastinal shift and partial collapse of the left lung was also noted. Arch of aorta and its major branches were of normal caliber and normal flow of contrast. Echocardiography of the heart showed no structural abnormality. Karyotyping was normal. Cardio-thoracic surgical

opinion was taken and the infant was posted for corrective surgery. But the parents refused for any operative intervention and left the hospital against medical advice.

Incidence of ALPA is 3–6% of all anomalies of the aortic arch system.⁵ Embryologically, part of the pulmonary artery is derived from the ventral sixth branchial arch and the rest is derived from the postbranchial vessel. Usually, the left postbranchial vessels connect to the left sixth branchial arch to form LPA and the right postbranchial vessels receive vascular supply from the right sixth branchial arch to make the right pulmonary artery (RPA).¹ The lungs develop from a pair of lung buds originating from the foregut endoderm. With the progressive lengthening of the primitive trachea, the lung buds are carried progressively more caudally until they reach their definitive thoracic position. The lung buds then branch sequentially and terminal bronchiole and alveoli are formed.⁶ Development of lungs interacts with the development of the pulmonary vessels as the same embryonic foregut mesoderm gives rise to the developing primitive pulmonary arteries and the primitive respiratory system.⁷

The etiology of ALPA is probably multifactorial. Report of ALPA in identical twins, and in patients with trisomies 18 and 21 hints towards genetic-related factors.^{8–10} Pu, et al hypothesized that when the left postbranchial vessels cannot connect to the left sixth branchial arch, a vascular supply from the most nearby major artery can be captured.⁴ An ALPA results when a connection is made to the right sixth branchial arch through the embryonic peritracheal mesenchyme between the trachea and the esophagus caudal to the developing tracheo-bronchial tree.

Association of cardiovascular malformations is reported with ALPA. Common cardiovascular lesions associated with ALPA are persistent left superior vena cava and PDA.¹¹ In our patient, there was no similar family history. Normal karyotyping ruled out any associated chromosomal abnormality. There was no associated cardiovascular malformation which probably made the survival possible without any major intervention.

In conclusion, ALPA is a rare anomaly that is increasingly being recognized today because of easy availability of noninvasive imaging modalities. This possibility should be kept in mind as a differential diagnosis of persistent stridor or respiratory distress in neonates. Moreover, recognition of this

Fig. 3 – CECT thorax showing aberrant left pulmonary artery originating from the posterior aspect of right pulmonary artery.

anomaly during imaging investigations should alert the clinician to the possibility of other associated malformations and their possible impact on further clinical management.

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