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Case Report

A case of pulmonary artery sling, unilateral lung hypoplasia, and congenital heart disease

Joyce Ashong[1], Sandra Kwarteng Owusu[2], Adwoa Pokua Boakye Yiadom[2], Naana Wireko Brobby[2]

[1]Department of Child Health, Cape Coast Teaching Hospital, Cape Coast, Central Region, [2]Department of Child Health, Kwame Nkrumah University of Science and Technology, Komfo Anokye Teaching Hosptial, Kumasi, Ashanti Region, Ghana.

***Corresponding author:**
Sandra Kwarteng Owusu, Department of Child Health, Kwame Nkrumah University of Science and Technology, Komfo Anokye Teaching Hosptial, Kumasi, Ashanti Region, Ghana.

abenaboamah18@gmail.com

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ABSTRACT

Vascular rings are rare congenital anomalies that primarily result from an embryological derangement of the paired aortic arches or branching pulmonary arteries. They make up <1% of all cardiac defects. Double aortic arch and right-sided aortic arch with the left ligamentum arteriosum are the most common types, making up 85–95% of cases, with pulmonary artery slings making up about 10% of cases. The prevalence is estimated as 59 per million children with very few cases reported worldwide. The clinical presentation is variable and this includes severe acute life-threatening episodes, recurrent apnea, severe respiratory distress, feeding difficulties persistent cough, wheezing, or stridor. In some cases, affected individuals may be completely asymptomatic. A high index of suspicion is needed to suspect and diagnose vascular rings as they are uncommon but surgically treatable. We present a case of a 5-day-old term neonate referred to our facility with respiratory distress associated with stridor which started 12 h post-delivery.

Keywords: Congenital anomalies, Congenital heart disease, Lung hypoplasia, Pulmonary artery sling

INTRODUCTION

Respiratory distress in newborns is one of the most common reasons for admission into the neonatal intensive care unit.[1] The causes although diverse and multisystemic can be broadly grouped into two – pulmonary and non-pulmonary. Common pulmonary causes include transient tachypnea of the newborn, respiratory distress syndrome, neonatal pneumonia, meconium aspiration syndrome, and persistent pulmonary hypertension of the newborn (PPHN). Cardiac disease, airway obstruction – from conditions such as subglottic stenosis, laryngeal webs, vascular rings, chest wall abnormalities, and neuromuscular disease are non-pulmonary causes of respiratory distress seen in neonates.[1,2]

Vascular rings are rare congenital anomalies that primarily result from an embryological derangement of the paired aortic arches or branching pulmonary arteries.[3] They were first described in 1737 by Hommel; however, the term vascular ring was first used in 1945 by Gross in the *New England Journal of Medicine*. They were subsequently classified by Klinkhamer and Stewart into complete – which includes; a double aortic arch (Type 1), right aortic arch with persistent left ligamentum arteriosus (Type 2), and a Type 3 which involves a Kommerell diverticulum, in which a ring is formed by a remnant of an incompletely regressed left or right arch, from which the left or right subclavian originates and incomplete – which include innominate artery compression syndrome and pulmonary artery sling (PAS).[4] Double aortic

arch, right aortic arch, and the left ligamentum arteriosum make up 85–95% of cases, with approximately 1% of aortic arch anomalies due to a vascular rings.[3,4]

PASs are less common, forming about 10% of cases. No gender, racial, or geographical predilection has been reported.[3] The estimated prevalence of PASs worldwide is reported as 59 per million children with very few cases reported.[4]

This case report looks at a term neonate presenting with severe respiratory distress and stridor 12 hours post-delivery and highlights the importance of considering vascular ring as an important differential in babies with such presentation. It also highlights the challenge in the diagnosis as well as management of this condition, with emphasis on the limited imaging modalities in the low- and middle-income countries, as well as the absence of well-resourced specialized units needed to manage the condition.

CASE REPORT

A 5-day-old female infant, born at term to non-consanguineous parents, with a birth weight of 2.6 kg was referred to our facility on account of the difficulty in breathing and stridor, noted 12 h after delivery. She was delivered through elective cesarean section at 38 weeks gestation on account of maternal history of three previous cesarean sections, systemic hypertension, and gestational diabetes mellitus. An initial diagnosis of congenital pneumonia and upper airway obstruction was made, intravenous Cefuroxime and Gentamicin were started before the baby was referred to our center. On arrival, the baby had no fever, she was in severe respiratory distress – with a respiratory rate of 82 cycles/min, flaring of the ala nasae, severe subcostal and intercostal recessions, grunting, and inspiratory stridor. Oxygen saturation was between 92 and 95% on intranasal oxygen at 2 L/min. Air entry was adequate bilaterally, breath sounds were bronchial with transmitted sounds noted on auscultation. Heart rate was 140 beats/min and was regular with heart sounds 1 and 2 present. A systolic murmur was noted, loudest at the aortic area. Other systemic examination findings were normal. The working diagnosis made was upper airway obstruction with differentials of subglottic stenosis, vocal cord paralysis, and extrinsic airway compression from vascular abnormalities. In addition, PPHN with congenital heart disease was considered.

Non-invasive ventilation, continuous positive airway pressure with positive end-expiratory pressure of 8 cm H_2O and FiO_2 of 90% was initiated to support the increased work of breathing and intravenous Cefotaxime 50 mg/kg 6 hourly, Dexamethasone 1 mg 12 hourly for 72 h, and Sildenafil 2.6 mg 6 hourly were started after blood samples were sent for investigations. Feeding was also started through the orogastric tube with 20 ml of expressed breast milk 3 hourly. Results from an initial round of investigations done showed: Hemoglobin of 16.7 g/dL; total white blood cells of 9.97 × 10^9/L; neutrophil count – 3.59 × 10^9/L; platelets – 100 × 10^9/L; urea – 1.07 mmol/L; and creatinine – 37 umol/L with negative blood cultures. Initial chest radiograph done showed opacification of the left lung with silhouetting of the left cardiac border and obliteration of cardiophrenic and costophrenic angles [Figure 1]. Features were noted to be suggestive of a left lung consolidation with no stenosis/narrowing noted in the upper airway. We were unable to carry out bronchoscopy due to the unavailability of the rigid or flexible bronchoscope in pediatrics sizes. A Chest CT scan was done, however, showed hypoplasia of the left lung with narrowing of the left main bronchus [Figure 2]. Pulmonary vessels were not clearly visualized.

We could not do an echocardiogram for technical reasons. A computerized tomography pulmonary angiogram could only be done after 26 days on admission. Results showed features suggestive of coarctation of the aortic arch distal to the origin of the left subclavian artery. In addition, there was a patent ductus arteriosus (PDA), an atrial septal defect (ASD), and dilatation of the right atrium. A vascular ring made up of the left pulmonary artery originating from the

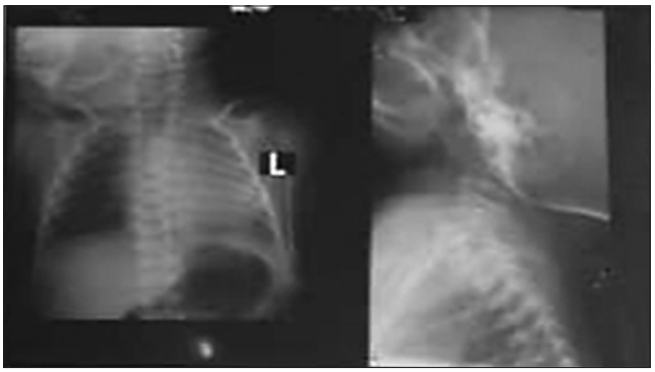

Figure 1: Chest X-ray + Lateral neck X-ray of the patient presented showing the homogeneous opacification of the left hemithorax with contralateral shift of the trachea.

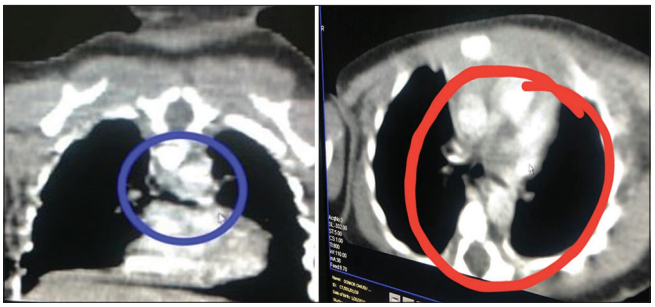

Figure 2: CT chest-coronal and an axial view showing narrowing of the left main bronchus. Blue circle: Chest CT in coronal view showing a narrowed left main bronchus at the level of the carina. Red circle: Chest CT in axial view showing a narrowed left main bronchus.

right pulmonary artery, which travels posterior to the trachea before entering the left pulmonary hilum, and left lung hypoplasia was also seen [Figure 3].

The baby was persistently in respiratory distress with associated stridor throughout the admission and stayed on non-invasive ventilation continuous positive airway pressure, as this was the best mode of ventilation that was immediately available. Throughout her period of stay, she also had feeding challenges, with multiple episodes of desaturation with feeds especially when the volume of feeds was increased. The baby died after 33 days on the admission of respiratory failure.

DISCUSSION

PAS, also known as an aberrant left pulmonary artery, was first described by Glaevecke and Doehle in 1897[5] and is reported to constitute 10% of all aortic arch anomalies.[3] The anomaly results from a failure of development of the left sixth aortic arch, coupled with the development of a collateral branch from the posterior aspect of the right pulmonary artery to supply the left lung.

Pu *et al*. hypothesized that when the left post-branchial vessels are unable to connect to the left sixth branchial arch, a collateral blood supply is derived from the nearest major artery. A PAS 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 tracheobronchial tree.[6] The etiology is likely to be multifactorial with reports suggesting genetic-related factors (close association of band 22q11 deletion).[3]

PAS is frequently associated with other tracheobronchial abnormalities with approximately 50–65% developing the ring-sling complex – defined as a combination of a PAS with a complete tracheal ring, producing upper airway obstruction.[3,5] Other tracheobronchial abnormalities include tracheomalacia, tracheal stenosis, and right tracheal bronchus which is described as an aberrant bronchus that arises from the right tracheal wall above the carina[3,5] and an underdeveloped right lung, all of which were absent in the case presented. Congenital heart defects are found in 50% of the cases, most commonly ASD, PDA, and aortic stenosis/aortic arch anomalies which were present in our case.[3,5,7] Others include ventricular septal defect, left superior vena cava, and a common ventricle. Clinical presentation is variable running along a spectrum of no symptoms to acute life-threatening episodes including severe respiratory distress, apnea, and stridor as was seen in our case. Other symptoms include recurrent respiratory tract infection, wheezing, dysphagia, recurrent vomiting, and failure to thrive.[3,5] It is worth noting that two-thirds of patients with PAS present with symptoms related to airway disease during the 1st month of life.[8] Useful diagnostic tools include CXR which is usually an initial test that may show unilateral hyperinflation or atelectasis of the right lung and an abnormal horizontal course of the left and right main bronchi, resulting in a T-shaped trachea. Findings are, however, non-specific[3] as seen in the case presented. Echocardiography is another useful diagnostic tool that can provide detailed images of the heart and central great vessels, including the initial segments of the pulmonary artery branches as well as intracardiac anomalies. It is preferred in children because of its non-invasive nature and is thus most often referred to as the primary diagnostic tool.[9] In the case presented, echocardiography was not done as it was not available during the period of admission. Other investigative modalities include magnetic resonance imaging, which although can evaluate both intra and extracardiac anomalies noninvasively, is not commonly used because of the need for deep sedation or general anesthesia.[3] Bronchoscopy, rigid, or flexible is the gold standard diagnostic tool especially when tracheal narrowing or obstruction is suspected, with some literature advocating that all cases of PAS should have bronchoscopy done.[5] Bronchoscopy was, however, not done in our case, as bronchoscopy services were not available at the time. CT angiogram is another diagnostic tool that gives a detailed description of the anatomy of the anomalous left pulmonary artery, the location, degree, and extension of trachea stenosis, and the spatial relationship among the PAS, trachea, and the esophagus. Echocardiography and CT angiogram have a complementary relationship and provide a comprehensive evaluation of the heart (both intra and extracardiac), which is necessary for the definitive diagnosis that is essential for precise operative planning. Definitive diagnosis, however, is often provided by CT angiogram[10] as was done in our case. Management is mainly surgical and survival of symptomatic infants is said to be unlikely without surgical intervention.[7] Early surgery decreases morbidity and allows for normal tracheal growth. Early diagnosis

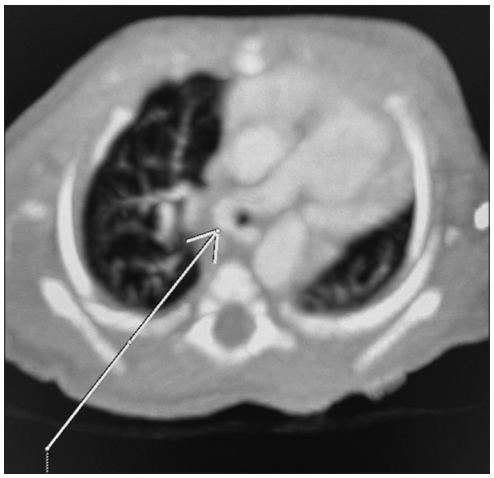

Figure 3: CT pulmonary angiogram showing pulmonary artery sling (PAS) (shown with arrow).

is, therefore, necessary to improve outcomes in affected patients. The left pulmonary division with reimplantation into the main pulmonary artery with/without tracheal repair is the preferred procedure with about 90% of patients having normal respiratory function with a resolution of their symptoms postoperatively.[3] Limited cardiovascular/thoracic surgery and pediatric intensive care centers in most low- and middle-income countries preclude the opportunity of these patients to be salvaged surgically especially in the complex cases such as our case who had other associated congenital cardiac anomalies challenging the surgical options.

CONCLUSION

Although uncommon vascular rings must be considered as one of differentials in newborns with unexplained respiratory distress.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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