

Breathing and Feeding Woes: A Neonate's Journey through Vascular Conundrum

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Abstract:

Vascular ring anomalies, such as a right-sided aortic arch with an aberrant left subclavian artery (ALSA) and Kommerell diverticulum, are rare congenital conditions that can cause significant oesophageal compression and respiratory distress in neonates. This case report describes the clinical presentation, diagnostic evaluation, and management of a baby boy with such a vascular ring anomaly. Born at 37 weeks of gestation via elective caesarean section due to non-progression of labour, with a birth weight of 2.78Kg, he had appearance, pulse, grimace, activity and respiration (APGAR) scores of 8 and 9 at 1 and 5 minutes, respectively. His antenatal course was unremarkable, with normal foetal echocardiogram and chromosomal microarray. The postnatal period was initially uneventful, but at 11 days of life, the baby presented with respiratory distress, feeding difficulties, and prolonged feeding times. He was admitted to another hospital and managed with intravenous (IV) fluids, orogastric feeds, oxygen, and antibiotics, but his symptoms persisted, prompting referral to our institution. On admission, the baby exhibited intercostal retractions, bilateral crepitations, and an oxygen saturation of 86%. Initial management included continuous positive airway pressure (CPAP) support, orogastric feeds, and IV antibiotics. Diagnostic tests revealed a moderate peri-membranous ventricular septal defect (VSD), a right aortic arch, and haziness in the right lower lung zone. Despite initial improvement, symptoms recurred and worsened on initiation of oral feeds, and further evaluation uncovered a 20% lumen compromise due to pulsatile vascular airway compression and significant oesophageal compression on flexible bronchoscopy and upper gastrointestinal (GI) fluoroscopy. A plain computed tomography (CT) chest scan confirmed a complete vascular ring with Kommerell diverticulum. The family opted for a delay in surgical intervention, and the baby was managed with nasogastric feeds. At 7 months, he underwent surgical division of the vascular ring, ALSA reimplantation, and VSD closure. Postoperative recovery included management of oesophageal stenosis with serial balloon dilatations. At 1.5 years of age, the child is thriving without further complications.

Key words: Vascular Ring, Right Aortic Arch, Aberrant Left Subclavian Artery, Kommerell Diverticulum, Oesophageal Compression.

Background

Vascular ring anomalies, such as a right-sided aortic arch with an aberrant left subclavian artery (ALSA) and Kommerell diverticulum, are rare congenital conditions that can cause significant oesophageal compression and respiratory distress in neonates. This case report describes the clinical presentation, diagnostic evaluation, and management of a baby boy with such a vascular ring anomaly.

Case Report

A baby boy, born via non-consanguineous marriage with a birth weight of 2.78Kg, presented at 11 days of life with respiratory distress. His antenatal course was uneventful, with a foetal echocardiogram showing a right aortic arch and a foetal chromosomal microarray ruling out any genetic issues. He was delivered by caesarean-section at 37 weeks due to non-progression of labour, with appearance, pulse, grimace, activity and respiration (APGAR) scores of 8 and 9 at 1 and 5 minutes, respectively. The immediate postnatal period was uneventful, and he was discharged on the third day.

At home, the baby experienced feeding difficulties, prolonged feeding times, and occasional non-bilious vomiting. He was predominantly breastfed. On the sixth day, fast breathing was noted, without fever or significant cough, and no family history of viral or flu-like illness was reported. He was admitted to another hospital, where he was managed with intravenous (IV) fluids, orogastric feeds, oxygen, IV antibiotics, and Lasix. However, his symptoms persisted, leading to a referral to our hospital for further management on the 11th day of life.

On admission, the baby had a temperature of 36.5°C, respiratory rate of 68/min with intercostal retractions, bilateral crepitations, saturation of peripheral oxygen (SpO₂) of 86% (Downe's score 4), heart rate of 138/min, a pan systolic grade 3/6 murmur, well palpable pulses, non invasive blood pressure (NIBP) of 68/40mmHg (mean, 50), and a random blood sugar (RBS) of 70mg/dL. He had good tone and activity, no dysmorphism, and no visible external malformations. Intermittent hoarseness was noted while crying. He was started on continuous positive airway pressure (CPAP) support (6 cm H₂O and fraction of inspired oxygen (FiO₂) 30%), orogastric feeds, and IV antibiotics pending blood culture results. Initial investigations showed a positive C-reactive protein (CRP) (25 mg/L) with sterile blood and cerebrospinal fluid (CSF) cultures. A chest radiograph showed haziness in the right lower zone, and 2D ECHO revealed a moderate peri-membranous ventricular septal defect (VSD), with a right aortic arch. An upper respiratory panel polymerase chain reaction (PCR) test was negative for common respiratory viruses. The baby was weaned off CPAP on the 15th day as his distress gradually settled. However, on the 16th day, after attempting oral feeds, he developed cough and respiratory distress again, necessitating re-initiation of high flow nasal cannula (HFNC) support and orogastric feeds. There was no tachycardia and no hepatomegaly. Repeat infection markers were negative. Chest radiograph showed increased bilateral infiltrates. Aspiration was considered due to worsening distress with oral feeds and no response to antibiotics. Further

evaluation for airway malformations and vascular anomalies was conducted. Flexible upper airway bronchoscopy revealed a 20% lumen compromise due to pulsatile vascular airway compression. Upper gastrointestinal (GI) fluoroscopy showed postero-lateral indentation on the contrast filled oesophagus, suggestive of significant oesophageal compression, with contrast aspiration into the right main bronchus (Figure 1).



Figure 1: Upper gastrointestinal (GI) fluoroscopy (oblique view) shows postero-lateral indentation on the contrast filled oesophagus (yellow arrow) and aspiration of contrast into right main bronchus (orange arrows).

A plain computed tomography (CT) chest scan identified a right-sided aortic arch with an aberrant left subclavian artery (ALSA) and Kommerell diverticulum, forming a complete vascular ring, causing significant oesophageal compression and aspiration pneumonia (Figure 2).



Figure 2: Computed tomography (CT) angiography axial view showing left common carotid artery arising from right-sided aortic arch coursing anterior to the trachea (yellow arrow); Aberrant left subclavian artery (ALSA) courses posterior to oesophagus (red arrowhead) on the left side-making a vascular ring.

Surgical correction was advised, but the family opted to wait until 6 months. In the interim, the baby was managed with nasogastric feeds at home. Repeat fluoroscopy at 6 months confirmed ongoing oesophageal compression and obstruction. At 7 months, the baby underwent vascular ring division with ALSA reimplantation and VSD closure. Post-surgery, he developed oesophageal stenosis, which was managed with serial balloon dilatation. Subsequently, there was no aspiration, and the baby is now thriving well at 1.5 years.

Discussion

Vascular rings are congenital anomalies of the aortic arch that result in compression of the tracheobronchial tree and or oesophagus, leading to respiratory and GI symptoms.^{1,3} They can be classified as complete (when both trachea and oesophagus are fully encircled by a vascular anomaly) or incomplete. Vascular rings with significant symptoms are usually complete type.¹ Double aortic arch, right aortic arch with aberrant vessel, innominate artery compression syndrome, aberrant left subclavian and pulmonary vascular sling are the major vascular rings.⁴⁻⁶ Kommerell diverticulum is an outpouching that arises from an aberrant subclavian artery, usually in the context of a right aortic arch with an aberrant left subclavian artery.^{7,8} This diverticulum can cause compression of the oesophagus and trachea, leading to symptoms like those caused by vascular rings. Vascular rings can either be isolated or be associated with other cardiovascular anomalies. Associated non-cardiac anomalies include tracheal stenosis (associated with a pulmonary sling), tracheomalacia, and laryngomalacia.⁹ Associated genetic abnormalities include CATCH 22 syndrome, Rubinstein-Taybi syndrome, and chromosomal aberrations.¹⁰ Vascular rings commonly present with respiratory symptoms which include inspiratory stridor, recurrent respiratory infections,

recurrent wheeze, cough, respiratory distress, and recurrent extubation failure.^{1,4,11} Symptoms indicative of oesophageal compression include dysphagia, choking while feeding, feeding difficulty, reflux, and vomiting.⁹ In infants experiencing repeated respiratory or oesophageal compressive symptoms, especially when arch anomalies are present, maintaining a high index of suspicion for diagnosing vascular rings is crucial.² Vascular ring causing oesophageal compression and aspiration in neonates is rare and is sparsely reported in literature. Diagnostic modalities include flexible bronchoscopy, fluoroscopy/ GI contrast study to see for posterior indentation of oesophagus, chest radiograph, CT angiograph/magnetic resonance imaging (MRI) to assess the arch and branching pattern and echocardiography with doppler and colour mapping.^{5,11} Surgical correction is the mainstay of treatment for symptomatic vascular rings, though not all vascular rings require surgery.¹ Asymptomatic babies need no surgical treatment even when the anomalies are found incidentally.² For infants with mild symptoms careful feeding with soft foods and aggressive treatment of pulmonary infections is required.⁵ Severe respiratory distress, recurrent pulmonary infections, oesophageal compression causing aspiration pneumonia, and apnoeic spells are few indications for surgical intervention.⁸ The timing of surgery depends on the severity of symptoms and is critical, as delaying intervention can lead to progressive airway and oesophageal damage, worsening symptoms, and an increased risk of respiratory infections.¹ Surgical approaches vary depending on the specific anatomy of the vascular ring or diverticulum but generally involve division of the vascular structures causing the compression.⁹ Postoperative complications include chylothorax, transient hypertension, oesophageal stenosis and vocal cord paralysis/paresis.

Conclusion

Vascular rings and Kommerell diverticulum, though rare, are significant causes of respiratory and feeding difficulties in neonates and infants. Early recognition and diagnosis are essential for effective management and to prevent serious complications. Advances in imaging technology have greatly improved the ability to diagnose these conditions accurately, allowing for timely and effective surgical interventions. Multidisciplinary care involving paediatric cardiologists, radiologists, and surgeons is key to optimising outcomes for affected infants.

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