

Aortic Arch Anomaly, Respiratory Distress and Difficult Feeding in a Premature Infant: A Case Report

Archana Bottu¹ | Siddharth Mahajan² | Shabih Manzar^{3*}

***Correspondence:** Shabih Manzar

Address: ¹Intern, Department of Pediatrics, Louisiana State University Health Sciences Center at Shreveport 1501 Kings Hwy, Shreveport, LA; ²Pediatric Cardiologist, Department of Pediatrics, Louisiana State University Health Sciences Center at Shreveport 1501 Kings Hwy, Shreveport, LA; ³Neonatologist, Department of Pediatrics, Louisiana State University Health Sciences Center at Shreveport 1501 Kings Hwy, Shreveport, LA

e-mail ✉: shabih.manzar@lsuhs.edu

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ABSTRACT

We present a case of a preterm infant with persistent respiratory distress and poor feeding. A modified barium swallow study showed posterior indentation on the esophagus. A cardiac computed tomography angiography showed a right aortic arch with an aberrant left subclavian artery from Kommerell's diverticulum. The infant underwent surgery to release the compression. The infant tolerated the procedure well.

Keywords: Preterm Infant, Right Aortic Arch, Cardiac Computed Tomography Angiography

Introduction

In neonates, aortic anomalies could lead to clinical symptoms, including respiratory distress, cyanosis, and dysphagia due to the compression of the trachea and esophagus (Reynolds *et al.*, 2015; Backer, 2020). With the availability of ultrasound technology, these conditions could be diagnosed prenatally (Bornaun *et al.*, 2021; Babaoğlu *et al.*, 2022). We present a case of a preterm infant diagnosed with an aortic arch anomaly that was causing persistent respiratory distress and feeding intolerance.

Case Presentation

A preterm infant was born at 30 1/7 weeks of gestational age via cesarean section to a 28-year-old gravida G6, para 3 mother with pregnancy complicated by di-di twin gestation and fetal growth restriction (FGR), required positive pressure ventilation (PPV) and continuous positive airway pressure (CPAP) in the delivery room. She was admitted to the neonatal ICU due to prematurity and respiratory

distress, briefly requiring intubation on the day of delivery. Her birth weight was 870 grams.

She required prolonged ventilation support in the form of CPAP for nearly a month before starting a high-flow nasal cannula (HFNC). She was further weaned to NC after 10 days but had difficulty weaning to room air after. In the interim, she was feeding via nasogastric tube and had eating difficulty. The neonate had good growth but could not fully tolerate oral feedings. Two weeks after birth, she had several apnea, bradycardia, and desaturation episodes and often choked while feeding. The chest x-ray was not suggestive of advanced pulmonary pathology. A modified barium swallow (MBS) study was obtained (Fig. 1), which showed posterior indentation.



Figure 1: Modified barium swallow study showing indentation in the middle part of the esophagus.

The differential diagnosis included a vascular ring, aortic arch anomaly, cardiac tumor, posterior mediastinal mass, acquired esophageal atresia, congenital esophageal stenosis, and pulmonary artery sling (Reynolds *et al.*, 2015; Backer, 2020; K Rahmath and Durward, 2023). Based on the findings of the MBS, a cardiac computed tomography angiography (CTA) was obtained that showed the right aortic arch with descending thoracic aorta on the right and an aberrant left subclavian artery with a short diverticulum of Kommerell (Fig. 2-3).

The CTA confirmed the suspicion raised by an earlier echocardiogram that showed a probable primum atrial septal defect versus a low secundum atrial septal defect with concern for a right aortic arch with an aberrant left subclavian artery from Kommerell's diverticulum vs. double aortic arch. The infant was transferred to Cardiac PICU for further management. The surgeons decided not to excise the Kommerell diverticulum because the diverticulum was not very prominent on direct inspection, and the

infant's weight was only 2.5 kg. The infant underwent a left posterolateral muscle-sparing thoracotomy. The surgeons ligated the ligamentum arteriosum and freed the tissues around the esophagus (Fig. 4). There was no remaining extrinsic compression on the esophagus. The infant tolerated the procedure well.

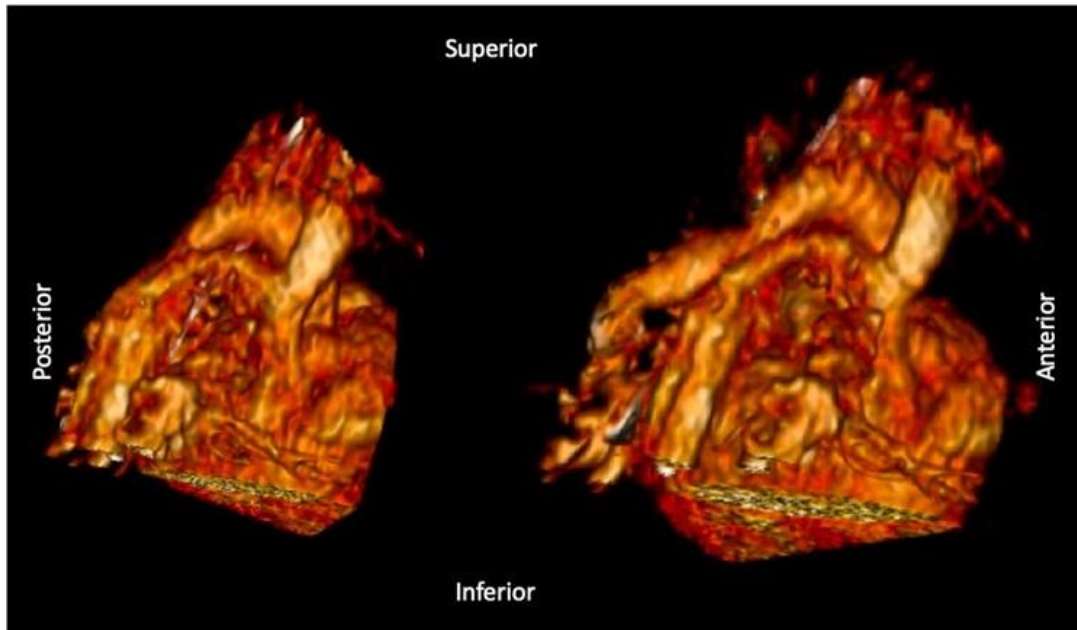


Figure 2: The reconstructed image obtained by the cardiac computed tomography angiography (CTA) showing the right-sided aorta.

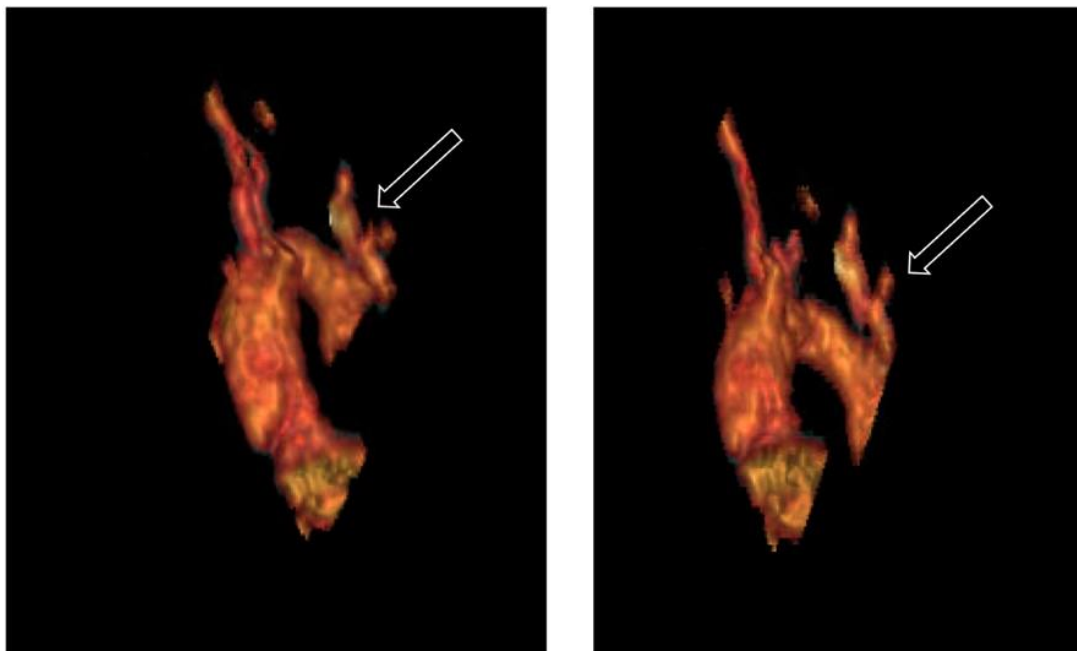


Figure 3: The reconstructed image obtained by the cardiac computed tomography angiography. (CTA) showing the branches of the right-sided aorta. An open white arrow points to the aberrant left subclavian artery arising from the diverticulum of Kommerell.

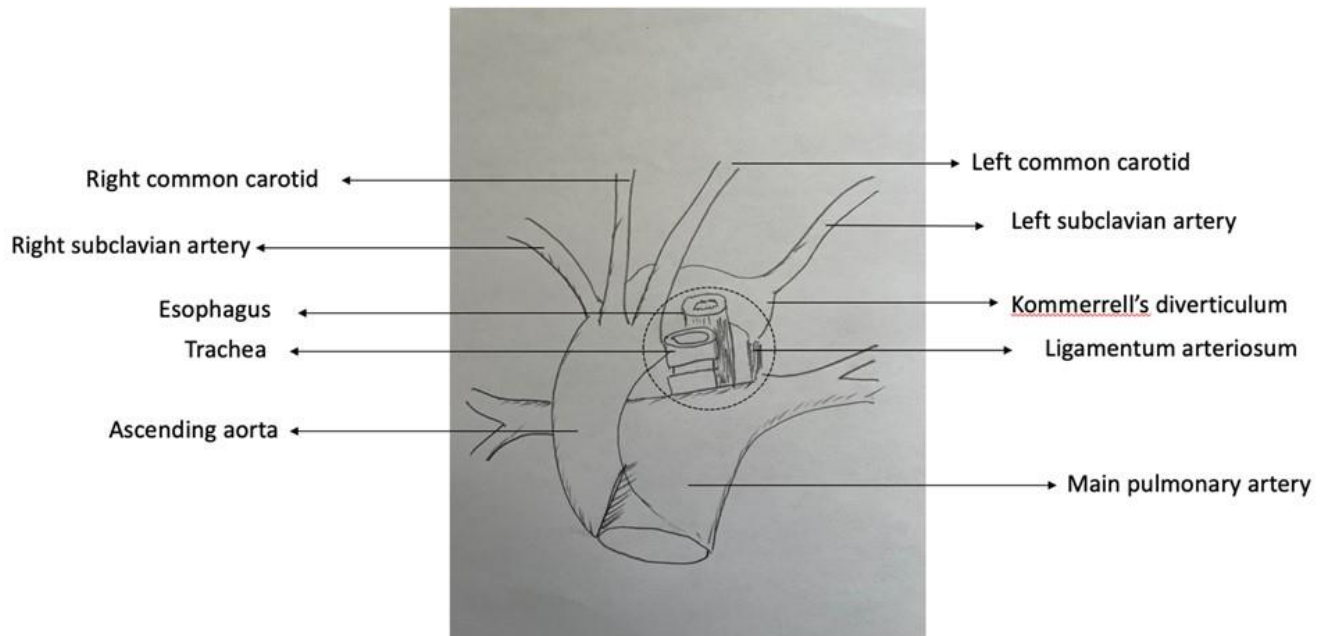


Figure 4: Animation showing the altered anatomy. The dotted circle represents the compression area.

Discussion

The indentation seen in the MBS was suggestive of an external compression on the esophagus. The acquired esophageal atresia and congenital esophageal stenosis were ruled out as these are intrinsic anomalies. A cardiac tumor or mediastinal mass would have presented with other signs like cardiac dysfunction or a mass effect. A complete vascular ring was ruled out after the CTA. It was not the case of a double aortic arch encircling and forming a complete ring around the esophagus. The obstructive symptoms manifested by the infant were due to the compression caused by the ligament arteriosus (Fig. 4).

The compression-related symptoms have been reported by (Renoyld, *et al.* 2015) in association with the aberrant right retroesophageal subclavian artery. The other reason for compression could be the Kommerell diverticulum (Backer, 2020). We did not find any other anomalies in neck vessels. Cyanosis has been reported with Kommerell's diverticulum with right-sided aortic arch and other vessel anomalies (Kanwal *et al.*, 2021). The treatment would be the resection of the diverticulum; however, symptoms might persist after surgery (Luciano *et al.*, 2015; Callahan *et al.*, 2020).

Conclusion

An infant who cannot be weaned from respiratory support and has difficulty feeding for no apparent reason should be evaluated for aortic arch anomalies.

Acronyms and Abbreviations:

PMA-postmenstrual age

MBS-modified barium swallow (MBS) study

CTA-Cardiac computed tomography angiography

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