

Aberrant Right Subclavian Artery with Kommerell's Diverticulum and Patent Ductus Arteriosus: Unusual Combination of Congenital Heart Anomalies and Clinical Manifestations in A Neonate

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ABSTRACT

Background: A one-month-old neonate presented to the hospital with dyspnea and bloody stool, which happened suddenly and progressed over two days.

Case presentation: Computed tomography and three-dimensional reconstruction of the trachea and heart was done, demonstrating significant trachea stenosis, aberrant right subclavian artery arising from Kommerell's diverticulum, and patent ductus arteriosus. Reconstruction of the aberrant subclavian artery, resection of the diverticulum, and ligation of ductus arteriosus was performed.

Conclusion: The case reported a rare combination of congenital anomalies and rare clinical manifestations at the same time. We thought the anatomical anomalies caused necrotizing enterocolitis (NEC), which lead to bloody stool.

INTRODUCTION

The brachiocephalic trunk (BCT), left common carotid artery (LCCA), and left subclavian artery (LSA) from the proximal arch to distal arch is the typical branching pattern of the left-side aortic arch (LSAA), and the right common carotid artery (RCCA) and right subclavian artery (RSA) are the branch vessels of the BCT. Aberrant right subclavian artery (ARSA) is a variant branch arising from the aortic arch between the LSA ostium and distal arch, and it is one of the most common congenital aortic arch anatomical anomalies, which has a prevalence of 8% overall variant patterns [Natsis 2021]. About 60% of ARSA patients are complicated with Kommerell's diverticulum (KD) [Zhou 2017], which is a diverticulum at the proximal descending aorta of the aortic arch and gives rise to the aberrant subclavian artery [Tanaka 2015]. The expanded KD may press the adjacent trachea and

esophagus, causing symptoms of dyspnea and dysphagia. This case report describes a rare combination of congenital anomalies consisting of ARSA with KD and patent ductus arteriosus (PDA) in a neonate.

CASE PRESENTATION

A one-month-old male neonate presented to the emergency department with bloody stool and inspiratory dyspnea. These two main symptoms suddenly happened and progressed over two days. Other clinical manifestations included retraction signs of three fossae, feeding difficulties, and decreased activity. Medical history suggested the baby was a premature infant, and there were no noteworthy abnormal conditions with his mother's pregnancy. Physical examination of the nervous system did not find abnormal signs. Computed tomography (CT) and three-dimensional reconstruction of the

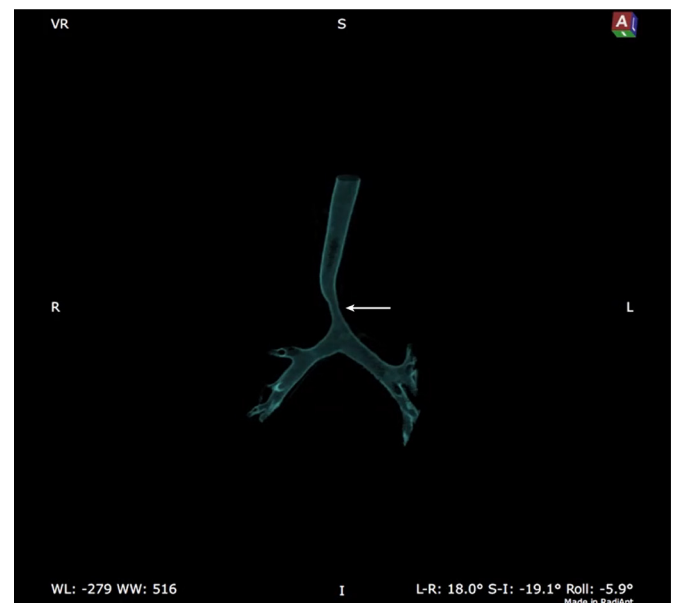


Figure 1. Three-dimensional reconstruction of the trachea demonstrated significant trachea stenosis (white arrow).

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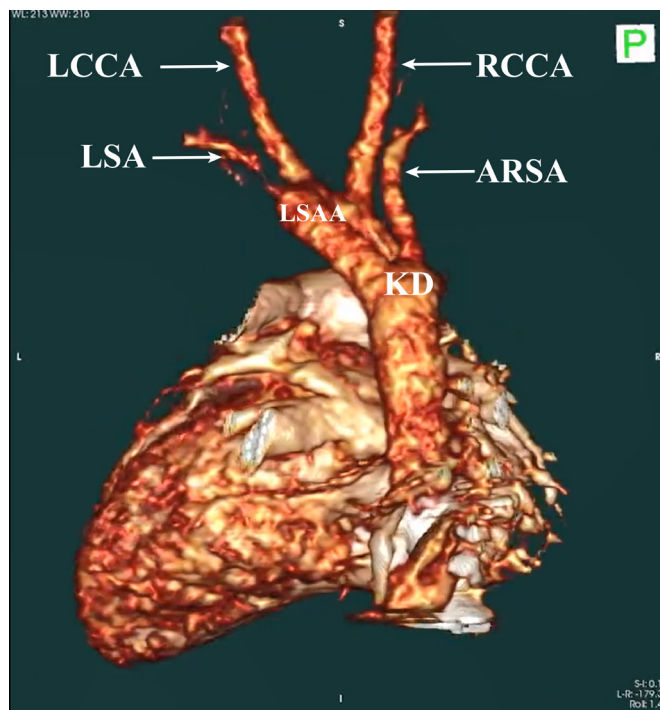


Figure 2. Three-dimensional reconstruction of the heart revealed four branches of the aortic arch (RCCA-LCCA-LSA-ARSA in sequence) and Kommerell's diverticulum. (LCCA, left common carotid artery; RCCA, right common carotid artery; LSA, left subclavian artery; ARSA, aberrant right subclavian artery; LSAA, left-side aortic arch; KD, Kommerell's diverticulum)

trachea and heart were performed to identify causes. Thin-section CT and three-dimensional reconstruction of the trachea showed significant stenosis of the trachea, especially at the position a little bit above the tracheal bifurcation. (Figure 1) Three-dimensional reconstruction of the heart revealed a variant pattern of LSAA, which consisted of four branches, including RCCA, LCCA, LSA, and ARSA from the proximal aortic arch to the distal arch. And, the origination of ARSA was dilated and formed KD. (Figure 2) Sight from another direction revealed a PDA of 3mm, connecting the KD and right pulmonary artery (RPA). (Figure 3) Thus, the left side segment of LSAA, retroesophageal KD, ductus arteriosus, and pulmonary artery composed a vascular ring around the trachea and esophagus.

Given the clinical features and radiological findings, the diagnosis of ARSA with KD and combined with PDA was confirmed. The compression to the trachea from the dilated KD and vascular ring caused symptoms, including dyspnea, feeding difficulties, and decreased activity. The bloody stool was considered secondary to the anatomical anomalies, which possibly was relevant to anoxia caused by trachea stenosis and the "diastolic steal phenomenon" caused by PDA. The infant underwent one-stage surgery, including reconstruction of ARSA, resection of the KD, and ligation of PDA. A median sternotomy was performed, and cardiopulmonary bypass was initiated through aortic and bi-caval cannulation. The patient

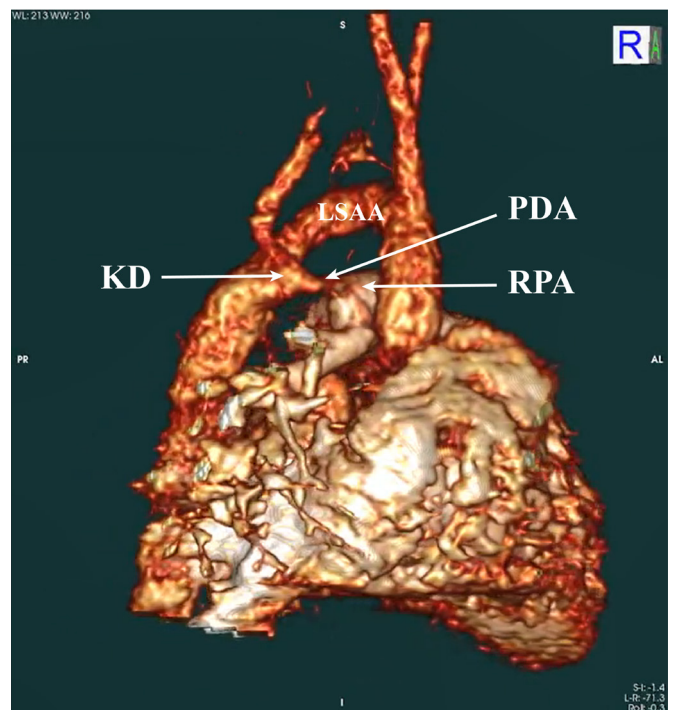


Figure 3. Three-dimensional reconstruction of the heart revealed a patent ductus arteriosus connecting the Kommerell's diverticulum and the right pulmonary artery. (PDA, patent ductus arteriosus; LSAA, left-side aortic arch; KD, Kommerell's diverticulum; RPA, right pulmonary artery)

was anticoagulated with 1 mg/kg heparin administered intravenously. Antegrade cardioplegia was administered to induce cardiac arrest. The avascular clamp was placed across the base of the KD and a portion of the descending aorta, and the portion of ARSA distal to the KD was also occluded, using a vascular clamp. The KD was resected, and the ARSA was then anastomosed to the opening of the descending aorta where the KD used to be, that means orthotopic transplantation. Finally, the ductus arteriosus was ligated at its pulmonary artery end and aorta end, and then was resected.

Postoperative vital signs were stable and the patient was discharged home without symptoms of dyspnea. At follow-up one month later, complete blood count demonstrated normal cell count and hemoglobin of 134g/L, and cardiac markers, including troponin, troponin T and CK-MB, returned to normal. TTE revealed no residual leaks at the PDA level, the dilation of the base of ARSA had disappeared. No dyspnea or bloody stool or other complications was found.

DISCUSSION

Typical branching pattern (BCT-LCCA-LSA sequence) of the aortic arch has a prevalence of 78% reported by a review of cadaveric studies, while variant patterns have 22% prevalence [Natsis 2021]. ARSA is a rare congenital anomaly with a prevalence ranging from 0.7 to 2.0% in the general population [Dumfarth 2015; Tanaka 2015]. ARSA usually

originates from a dilated base named KD, which is defined as a widening of the base of the subclavian artery that is more than 1.5 times the size of the distal subclavian artery [Backer 2012]. Considering the high risk of lethal complications like diverticulum rupture and aortic dissection [Azeem 2021], such patients usually are recommended to undergo the surgical procedure once KD was found [Mossad 2002; Kim 2014]. The most common surgical procedure was resection of the KD and reconstruction or translocation of ARSA. Many surgical incisions and operations have been reported. After considering age and the combination of PDA in this patient, a one-stage operation was decided. A median sternotomy and orthotopic transplantation of ARSA were mainly performed.

ARSA with KD is more likely to have pressure symptoms caused by direct compression from the KD or restriction from the vascular ring, such as dysphagia, dyspnea, coughing, chest pain, and right upper extremity ischemia [Stone 2011; Tanaka 2015]. Due to the absence of elastic tissue in neonates, the rigidity of the trachea is usually insufficient to resist the pressure, and dyspnea is more common in pediatric patients. This corresponded with the respiratory symptom in this case. Another noteworthy condition was the bloody stool, which suddenly appeared and disappeared after surgery. Drugs for the digestive system were not used. In this patient, besides ARSA with KD, a 3-mm PDA also should be taken into consideration, thus, the following reasons might conspire to cause this symptom.

First, duct-dependent systemic circulation, for example, hemodynamically significant PDA, may lead to impaired mesenteric blood flow through a phenomenon named “diastolic steal” [McElhinney 2000]. Ischemia of the mesenteric artery causes necrosis of intestinal mucosa, usually leading to bloody stool. Accordingly, some studies have reported correlation between congenital heart disease and necrotizing enterocolitis (NEC) in neonates [McElhinney 2000; Motta 2015]. One multicenter case-control study has considered hemodynamically significant PDA as an independent risk factor for NEC [El Manouni El Hassani 2021]. Secondly, decreased oxygenation secondary to the trachea stenosis would also increase the risk of NEC. Finally, a randomized controlled trial has confirmed that infants with congenital heart defects appear to have decreased diversity of gut flora [Ellis 2013]. Though the correlation between gut flora diversity and NEC has not been fully studied, it could be speculated that stability of intestinal environment would reduce, due to the change of gut flora, and bloody stool may happen. Without an examination of the digestive system, diagnosis of NEC could not be confirmed, however, through analysis of clinical manifestation and surgical findings, a great possibility of NEC caused by congenital heart defects should be taken into consideration.

CONCLUSION

In summary, this case report described a rare combination of congenital anomalies, including ARSA with KD and PDA, while the main clinical manifestations were dyspnea and

bloody stool. Stenosis of the trachea and NEC secondary to anatomical anomalies might explain these symptoms. A surgical procedure successfully was performed, and the patient safely was discharged home.

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