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Case Report

Unexpected Vascular Anomaly; Right Subclavian-Pulmonary Artery Connection Unraveled during Routine PDA Closer; Case Report

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Abstract
Aberrant origin of the right subclavian artery (ARSA) is an extremely rare congenital vascular anomaly. A systemic connection to the pulmonary artery occurs due to abnormal embryogenesis of the aortic arch. The clinical presentation of the patient depends on the degree of shunt & associated cardiac problems, making most of the patients asymptomatic, or they complain of respiratory distress, differential cyanosis, and feeding difficulties. Echocardiography might suspect the problem, but computed tomography angiography and cardiac catheterization are essential for definitive diagnosis & for delineating the exact anatomy and hemodynamic significance. Early recognition is important to prevent complications such as pulmonary overcirculation, congestive heart failure, or vascular steal phenomena. Surgical or transcatheter correction is usually required to re-establish systemic arterial supply and disconnect the anomalous pulmonary connection. Reporting such rare cases adds to the understanding of vascular embryology and helps optimize diagnostic and therapeutic strategies.
Keywords. *Aberrant Right Subclavian Artery; Patent Ductus Arteriosus; Echocardiography; Angiography.*

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Introduction

The aberrant right subclavian artery (ARSA) is an extremely uncommon pathology with an estimated incidence of 0.5-2% [1]. It has a male-to-female ratio of 2:1 and tends to be accidentally discovered during imaging. Sometimes it may be associated with complications such as debridement of the blood supply from the upper limb or respiratory distress, in ARSA, in which the RSA arises directly from the AA instead of originating from the brachiocephalic artery. The most common combined malformation in ARSA is intracardiac malformation [2]. We are discussing a case of isolation of the RSA, which was initially diagnosed as bilateral patent ductus arteriosus (PDA).

Case Presentation

A 7-month-old term infant was referred to Shaheed AL Mihrab Tertiary Center of Cardiology with a complex cardiac congenital heart defect. The general examination revealed the child had growth faltering, recurrent chest infection, he was complaining of dyspnea, poor feeding, and a cough that was worsening due to right-sided heart failure. The Echocardiography revealed, see table 1. (Fig.1A), and (Fig.1B). The child was admitted to the hospital for cardiac catheterization. In the catheterization laboratory, angiography results are highlighted in Table 1. And (Fig.2A), and (Fig.2B).

Table 1. The results of the imaging test

Investigation done	Results of the test
Echocardiography study	• Systemic & pulmonary venous return: normal • Great vessel relationship: normally related • Aortic arch: left-sided with anomalous right subclavian artery (ARSA) • Main pulmonary artery: markedly dilated • ASD: large secundum type, 9 mm • PDA: small left-sided PDA, additional PDA like flow at the origin of the right pulmonary artery • Pulmonary hypertension: severe

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Cardiac Catheterization / Angiography	• Confirmed restrictive Type E PDA • Left aortic arch with three-branch pattern (R CCA, L CCA, L SCA) • ARSCA arising anomalously from the right pulmonary artery instead of aortic arch • Collateral flow from descending aorta → RSCA → right pulmonary artery • Pulmonary vasculature: markedly dilated, severe pulmonary hypertension • Pulmonary venous drainage: normal

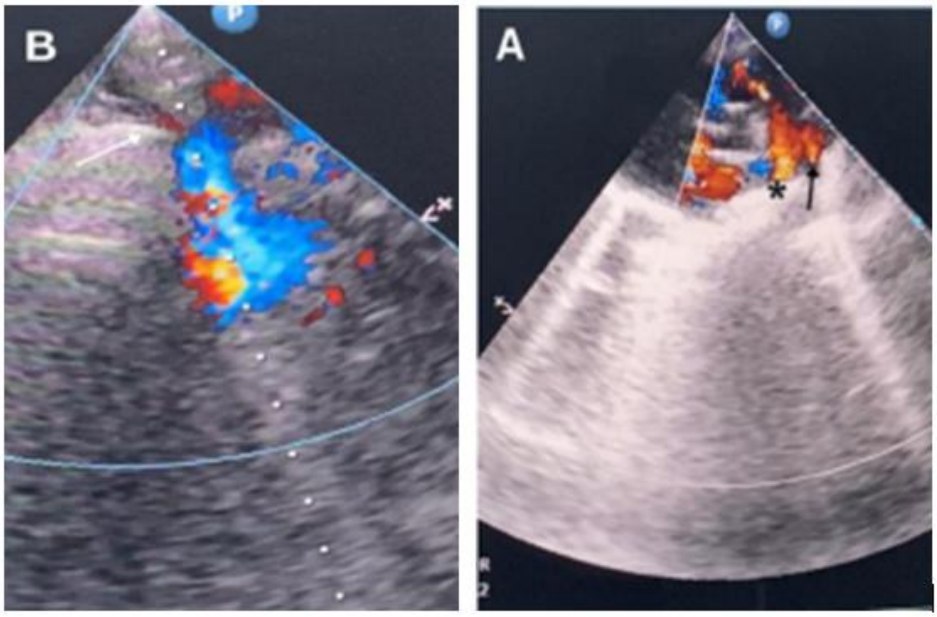

Figure 1. A two-dimensional color Doppler echocardiograph image showing A.PSAX view; there are bilateral arterial ducts (black arrow)highlighting a left-sided PDA shunt, (*= right-sided PDA shunt) .B a suprasternal long-axis view (white arrow) describing a flow of ARSA from descending thoracic aorta.

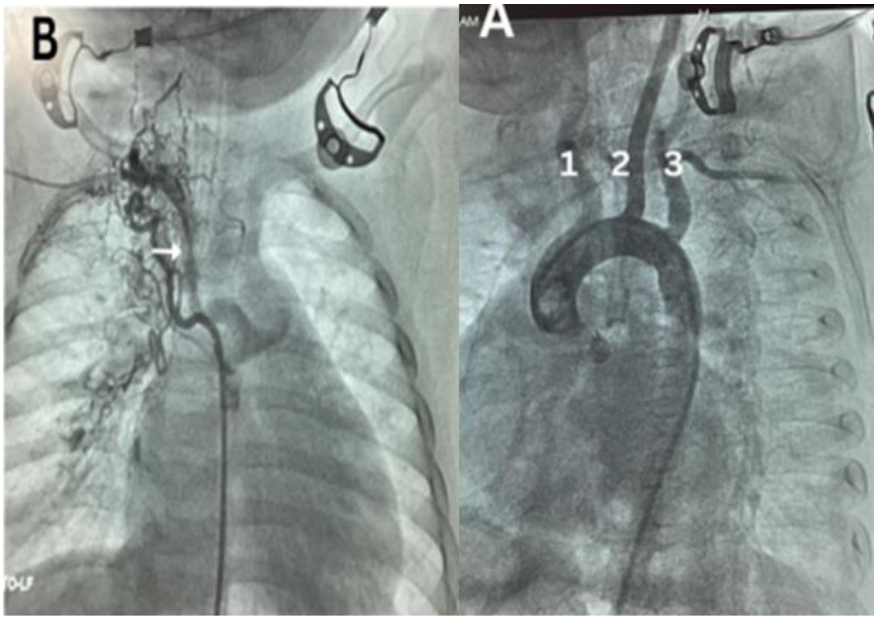

Figure 2. The Rt. Side photo is showing ascending aorta (AA) angiograph; there are 3 vessels in the AA, no.1 the left carotid and from it the LSA. No. 2 another left carotid branch no. Three right carotids with no RSA. The right-sided photo shows the RSA receiving blood supply from the descending aorta collateral to the upper right lung.

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Discussion

The embryological basis of ARSA lies in the faulty regression of the RT fourth AA, which lies between RCCA and the RSA. Since the RSA will fail to fuse with the ascending aorta, it will arise distally. Ideally, the left AA will persist and give rise to RCCA, LCCA, and LSA, with the RSA arising as an aberrant fourth branch from the distal arch or descending aorta [2,3]. ARSA is connected to the RPA by a right-sided arterial duct. During its path to the right arm, it passes the midline posterior to the esophagus in 85% of cases and is defined as ARSA. There is more than one classification for ARSA; one is Adachi William's classification of the subclavian artery and ARSA [4,5]. Type I (Type G) Four-branched patterns in the sequence of RCCA, left common carotid artery, left is called retroesophageal ARSA [4,5]. Additionally, it may pass between the esophagus and trachea or even go anterior to the trachea [5] or take a course behind the esophagus and trachea.

Table 2: Adachi William's classification of the subclavian artery & ASRSA

Type	Branching type	Key finding	Common course
Type I (Type G)	RCCA → LCCA → LSA → ARSA	Four distinct branches; ARSA arises last	Retroesophageal in 85% of cases
Type II (Type CG)	RCCA → LCCA → Left vertebral artery → LSA → ARSA	Additional left vertebral artery between LCCA and LSA	Retroesophageal
Type III (Type H)	Bicarotid trunk (RCCA + LCCA) → LSA → ARSA	Common carotid trunk for both carotids	Retroesophageal

Common carotid artery=CCA, right common carotid artery =RCCA, left common carotid artery=LCCA, Left subclavian artery=LSA; Aberrant right subclavian artery (ARSA)

According to the above classification, our case was type 1G; however, in the current case, the anomaly was further complicated by patent ductus arteriosus, where the flow to RSCA is maintained by flow from the aorta through left-sided PDA to pulmonary arteries and to RSCA. This dual circulation-maintained perfusion of the right arm but contributed to pulmonary overcirculation and severe pulmonary hypertension. From the hemodynamic point of view, the aberrant-systemic pulmonary shunt can create concerns if we attempt to close it, since it supplies the right arm. Our initial concern was that a PDA device led to obstruction of the aortic orifice of left-sided PDA, which maintains systemic pressures in the pulmonary arteries. Flow then decreased to RSCA and led to decreased perfusion of the right arm. This anatomical configuration illustrates the complex interplay of the embryological, vascular anatomy, and clinical decision-making in a rare congenital vascular anomaly. The ARSA was initially described by Hunauld [6,7] following an autopsy dissection. Later, in 1794, Bayford [8] described the first case in a 62-year-old female who passed away years suffering from chronic dysphagia. Apley presented the first angiography showing the disorder in 1949[9]. Our case is the first case reported with this specific physiology, which presented with bilateral PDA with a left AA, ARSA, and anomalous origin of ARSA from the right PDA arising from RPA. In asymptomatic cases, adopting observation only is enough if no indication exists for intervention. Surgical intervention is indicated for symptomatic aberrant RSA m presented with persistent dysphagia, upper limb ischemia, or the presence of an aneurysmal dilatation [6]. Any surgical intervention aims to relieve the symptoms caused by the aberrant artery and to re-establish right upper extremity blood flow. There are 3 main options for surgical repair: open repair, thoracic endovascular aortic repair (TEVAR), and hybrid repair.

The appropriate approach depends on the vascular anomaly, its size, patient symptoms, acuity, and body build. The first successful surgical treatment of ARSA was described by Gross in 1946[3], using a left thoracotomy and ligation of the aberrant vessel. This “classic” method has the risk of developing ischemia, so currently, revascularization is more recommended. Endovascular and hybrid techniques have been reported too, with thoracic endovascular aortic repair (TEVAR) joint with carotid-subclavian bypass providing effective exclusion of the aberrant origin while keeping arm perfusion intact [10].

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CONCLUSION

Recognition of variations of aortic arch branching is important because they may cause symptoms due to tracheoesophageal compression or complications during surgical or endovascular interventional procedures of the aorta and its branches. If Echocardiography shows flow of bilateral PDA at the origin of LPA&RPA, CT Angio or cath study is important to rule out ARSA.

Conflict of interest. Nil

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