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(RESEARCH ARTICLE)

An unusual discovery: Aberrant subclavian artery - symptoms, embryological pathology, surgical approach, and outcome analysis

Muhammed Saheen KV [1,*], Yasvanth Kumar [1], Swaminathan V [2] and Neville AG Solomon [2]

[1] *Department of Cardiothoracic Surgery, Apollo Hospitals, Chennai, India.*
[2] *Faculty, Department of Pediatric and Adult Congenital cardiac Surgery, Apollo Children's Hospital, Chennai, India.*

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Abstract

Introduction: Aberrant subclavian artery (ASA) anomalies, though rare, present unique challenges in clinical practice due to their varied clinical manifestations and associated anatomical complexities. This study aims to comprehensively elucidate the embryological basis, clinical spectrum, diagnostic modalities, management strategies, and outcomes of ASA anomalies.

Methods: A retrospective analysis was conducted on pediatric patients diagnosed with ASA anomalies between 2019 and 2023. Clinical data, including presenting symptoms, diagnostic findings, management approaches, and post-operative outcomes, were collected and analyzed.

Results: Four pediatric patients with ASA anomalies were included in the study, with three cases of aberrant right subclavian artery and one case of aberrant left subclavian artery. Most patients were presented with dysphagia secondary to esophageal compression, while associated anomalies such as Kommer ell's diverticulum and cardiac anomalies were commonly observed. Diagnostic modalities including prenatal ultrasound, CT angiography, and barium swallow studies aided in accurate diagnosis and evaluation of associated anatomical variations. Management strategies ranged from conservative measures to surgical interventions, with the hybrid approach demonstrating promising outcomes. Post-operative recovery was generally uneventful, with isolated cases of complications resolving spontaneously.

Keywords: *Arteria Lusoria*; *Dysphagia Lusoria*; Aberrant Subclavian Artery; Pharyngeal Arch Arteries; Dysphagia

1. Introduction

In 1971 David Bayford has made his discovery of the unique and bizarre cause for the compression of the esophagus by the aberrant right subclavian artery of a fatal case of obstructed deglutination for which he coined the term *dysphagia lusoria* [2].

Aberrant subclavian artery is the most common congenital aortic arch anomaly with an incidence of 0.4–1.8 % in the general population [3].

2. Embryology

The pharyngeal arch arteries are in six paired embryological vessels and seven paired cervical intersegmental arteries, Kommerell's diverticulum.

* Corresponding author: Muhammed Saheen KV

Table 1 Arteries arising from pharyngeal arches.

Pharyngeal Arch	Arteries
First	A portion of maxillary artery
Second	Stapedial artery
Third	Common carotid arteries Portion of internal carotid arteries
Fourth	Right side: Proximal part of right subclavian artery and pulmonary arteries. Left side: Segment of aortic arch.
Sixth	Pulmonary trunk

The proximal portion of the right subclavian artery is formed from the right fourth pharyngeal artery and the distal portion is formed from part of the seventh intersegmental artery. The left subclavian artery arises from the seventh intersegmental artery [4].

The aberrant origin of the right subclavian artery occurs due to the involution of the right fourth pharyngeal vascular arch. This in turn results in the persistence of the seventh intersegmental artery that originates from the proximal descending thoracic aorta [5].

2.1. Associated anomalies

It has been reported that an aberrant right subclavian artery is linked to chromosomal abnormalities such as trisomy 21, 22q11.2 microdeletion syndrome, and Turner syndrome. Even in fetuses without chromosomal abnormalities, there is a possibility of other associated structural abnormalities, with cardiac anomalies being the most frequently reported [6].

It can be left or right aberrant subclavian artery and is commonly associated with any other anomalies such as Kommerell's diverticulum, PDA, ASD, VSD, LSVC, right sided aortic arch and DORV [7].

Kommerell's diverticulum refers to the aneurysmal dilatation of the descending aorta at the origin of an aberrant left subclavian artery, which is seen in both right- and left-sided aortic arches, usually reported incidence was 0.05–0.1% [8].

3. Discussion of cases

From 2019 to 2023, we operated on four aberrant subclavian artery pediatric patients in which three were, aberrant right subclavian artery and one was aberrant left subclavian artery. Most of the patients were presented with other anomalies, only one patient is without any other anomalies.

3.1. Symptoms

Mostly they are asymptomatic, around 10 % of patients will be symptomatic due to compression of esophagus. The clinical implications of aberrant right subclavian artery primarily revolve around the potential for symptoms and complications. There are variations in the course of the aberrant right subclavian artery as it travels towards the right upper limb. One common variation is the retroesophageal course, where the aberrant right subclavian artery passes behind the esophagus can compress the esophagus and leads to dysphagia. Another variation is the cervical course, where the aberrant right subclavian artery travels upwards in the neck instead of the normal course through the chest. This can cause compression of the nearby nerves and leads to shoulder pain and arm weakness [9].

Moreover, aberrant subclavian artery has been associated with an increased risk of aneurysm formation [10], dissection, and thrombosis [10].

In our case series, most of the patients presented with symptoms of dysphagia mainly to solids due to compression of esophagus.

Table 2 Representing patient's data, including symptoms, diagnosing modality and compresing structure.

S.No	Sex	Age	symptoms	Pathology	Diagnosing modality	Compressing structure
1	F	4y	Dysphagia	Right aberrant subclavian artery with tiny PDA	BA swallow and CT aortogram	Esophagus
2	M	17Y	Dysphagia	Aberrant left subclavian artery arising from kommerell diverticulum compressing the esophagus	BA swallow and CT aortogram	Esophagus
3	F	5y	Failure to thrive	Ostium Secondum atrial septal defect with PAPVC (left upper pulmonary vein draining into left innominate vein via vertical) + right aberrant subclavian artery	BA swallow and CT aortogram	Mild esophageal obstruction
4	M	11 Y	Dysphagia	Aberrant right subclavian artery	BA swallow and CT aortogram	Esophagus

3.2. Diagnostic modalities

Table 3 Representing surgery done, approach used, difficulties, complications, and outcome.

Patient	Surgery	Approach	Difficulties	Complications	Outcome
1	Detachment and reimplantation of aberrant right subclavian artery to right common carotid artery with PDA division	Right thoracotomy through 3rd intercostal space. And right upper median sternotomy	Difficulty in anastomosing the right subclavian to carotid via thoracotomy hence done hemi sternotomy	Chylothorax	Good
2	Detachment and reimplantation of aberrant left subclavian artery to left common carotid artery with repair of kommerells diverticulum and ligation of ligamentum arteriosus.	Left postero lateral thoracotomy in 4th intercostal space and sternotomy	Difficulty in anastomosing the left subclavian to left carotid via thoracotomy hence done hemi sternotomy	no	Good
3	ASD with partial anomalous pulmonary venous connection repair + right aberrant subclavian artery reimplantation.	Primary median sternotomy	no	no	Good
4	Detachment and reimplantation of aberrant right subclavian artery to right common carotid artery with tiny PDA division	Right thoracotomy through 3rd intercostal space. And right upper hemi sternotomy	Difficulty in anastomosing the right subclavian to carotid via thoracotomy hence done hemi sternotomy	no	Good

Prenatally we can diagnose with help of ultrasound in the 2nd, and 3rd trimesters, respectively[11].

Accurately diagnosing Aberrant Subclavian Artery is crucial for effective management. Diagnostic modalities such as CT scan can help identify a vascular ring. The diagnosis of a vascular ring is often incidental, discovered during a CT scan that was performed for another indication. An echocardiogram can be used to check for any additional heart anomalies,

while bronchoscopy can be performed to look for any additional tracheal pathology [12] . Awareness of associated anatomical variations, such as Kommerell's diverticulum or vascular rings, is essential for a comprehensive evaluation.

We investigated dysphagia with barium swallow study, in view of indentation of esophagus at the 3rd and 4th vertebral level. We had proceeded with computed tomography angiography (CTA) to confirm the diagnosis and determine the appropriate course of treatment.

3.3. Management

Treatment options for ASA depend on the presence of symptoms, associated complications, and the patient's overall health status.

Conservative management with lifestyle modifications and medication is often recommended for asymptomatic individuals.

A hybrid approach using carotid subclavian artery bypass with TEVAR (thoracic endovascular aortic repair) is the new technique. It is often performed in two stages, whereby a carotid subclavian bypass is performed via a supraclavicular approach, followed by a thoracic stent graft placement via a percutaneous femoral approach. There are some complications which can be expected such as endo leaks, stroke, spinal cord ischemia [13].

Surgical interventions such as subclavian artery revascularization or bypass procedures, are considered in cases of severe symptoms or significant complications. We mobilize the aberrant subclavian artery and bring Infront of trachea and esophagus. End to side anastomosis has to be performed to common carotid artery. We tried to repair ASA with minimal invasive thoracotomy incisions but almost all the time we faced Difficulty in anastomosing the subclavian to carotid via thoracotomy hence we were forced to do at least a hemi sternotomy for completion of procedure.

3.4. Outcome

Almost all patients in post-operative period were uneventful except one patient who underwent Detachment and reimplantation of the aberrant right subclavian artery to right common carotid artery with PDA division has developed chylothorax, which settled without any intervention in early post operative period. All of them made steady recovery, no signs of recurrent laryngeal nerve injury, phrenic nerve injury or bleeding.

4. Conclusion

This study contributes valuable insights into the embryological basis, clinical spectrum, diagnostic approach, and therapeutic strategies pertaining to aberrant subclavian artery anomalies. By elucidating these facets, the study aims to inform clinical practice, enhance diagnostic accuracy, and optimize patient outcomes in the management of this intriguing vascular anomaly. Further research and collaborative efforts are warranted to refine existing management algorithms and advance our understanding of aberrant subclavian artery anomalies in pediatric populations.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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