Case Report

Journal of Cardiovascular Medicine and Surgery
Volume 3 Number 1, January - June 2017
DOI: http://dx.doi.org/10.21088/jcms.2454.7123.3117.10

Isolated Right Subclavian Artery in a Patient with Tetralogy of Fallot with Single Pulmonary Artery

Neeraj Kamat*, Ashish Gaur, Sandeep Sinha**, Anvay Mulay*****

Authors Affiliation
*Attending Consultant **Senior Physician Assistant ***Director & Head, Cardiac Surgery & Transplant Division, Fortis Hospitals, Mulund-Goregaon Link Road, Mulund (West), Mumbai, Maharashtra, India-400078.

Reprints Requests
Neeraj A. Kamat, Attending Consultant, Cardiac Surgery & Transplant Division, Fortis Hospitals, Mulund-Goregaon Link Road, Mulund(West), Mumbai, Maharashtra, India-400078

Received on 05.02.2017,
Accepted on 23.02.2017

Abstract

Isolated RSCA from the right pulmonary artery is a very rare vascular arch anomaly. Complete loss of continuity of subclavian artery from the aortic arch was named ISOLATION by Stewart and colleagues [4] in 1964. They are commonly associated with DiGeorge syndrome [2] or intracardiac anomalies [3]. We report a case of 6 years old female child diagnosed with Tetralogy of Fallot with single pulmonary (right) artery with no left pulmonary artery with an anomalous Right Subclavian artery arising from the single pulmonary artery with left arch and left superior vena cava. Patient underwent single lung repair with total transatrial approach with reimplantation of the right subclavian artery into the right carotid/ innominate artery (first branch of the ascending aorta). Early intervention with complete surgical repair normalised the patient's circulation with resolution of both the pulmonary and subclavian steal phenomenon.

Keywords: Right Subclavian Artery (RSCA); Tetralogy of Fallot (TOF); Pulmonary Artery(PA).

Introduction

AnIsolated subclavian artery is the rarest described arch anomaly [1] in which the subclavian artery is separated from the aortic arch and arising from the same sided pulmonary artery either connected through a ductus or ligamentum arteriosum. They are commonly associated with DiGeorge syndrome [2] or intracardiac anomalies [3] especially conotruncal defects (VSD, TOF, DORV) or aortic arch anomalies (Right or Interrupted Aortic Arch or Bilateral Ductus). There is no vascular ring [1] as the ductus is ipsilateral to the isolated subclavian artery and is not attached to the aorta. We present a similar case report.

Case Report

We report a case of 6 years old female child had come with complaints of increased incidence of Respiratory tract infection frequently since she was 3 years old and breathlessness on exertion with no history of cyanosis. Grade 2 clubbing present but no cyanosis. Saturation in the right upper limb and other limbs were the same approximately 96% with normal development of all the four limbs. BP in right upper limb was less than the other limbs. There were no any positive family history for congenital heart disease. On examination, she had single 2nd heart sound with a systolic thrill and murmur heard in the pulmonary area. She was investigated and found to have on Echo and CT angio as Tetralogy of Fallot with single pulmonary (right) artery with no left pulmonary artery with an anomalous Right Subclavian artery arising from the single pulmonary artery with left arch and an LSVC. Vertebral artery was arising from the right subclavian artery. Neck vessel angio showed patent cicle of Willis. CXR showed increased Qp more on the right side as compared to the left side.

Patient underwent single lung repair with total transatrial approach with reimplantation of the right

subclavian artery into the right carotid/innominate artery (first branch of the ascending aorta). Single Lung Repair consisted of Dacron Patch Closure of the VSD with Infundibular and Valvular Resection. Pulmonary Annulus was within normal limits and hence did not require the need for Transannular Augmentation. Patient recovering was stable in the postoperative period with 100% saturation in the right upper limb and was thus discharged.

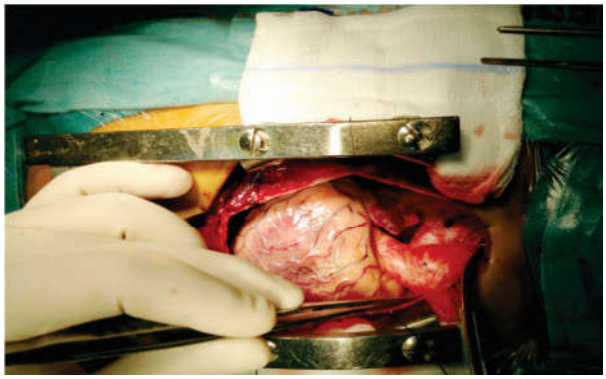

Fig. 1:

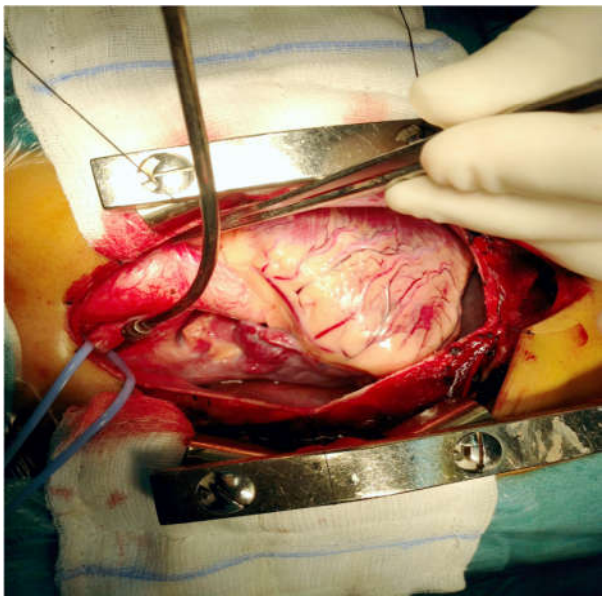

Fig. 2:

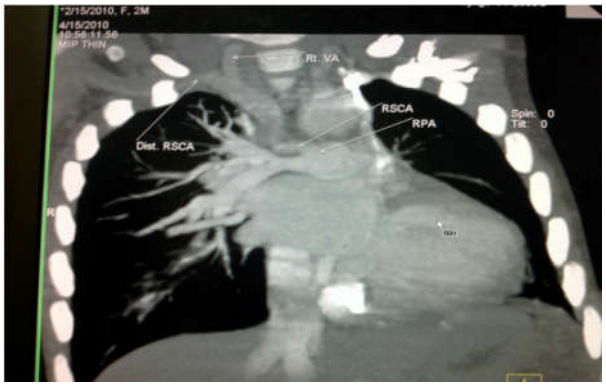

Fig. 3:

The reimplantation was done off CPB with a clamp placed across the first branch of the ascending aorta higher up and right subclavian artery divided flush at its origin from the pulmonary artery and anastomosed with the first arch vessel in an end to end fashion as illustrated in the following figures.

Discussion

Complete loss of continuity of subclavian artery from the aortic arch was named **Isolation** by Stewart and colleagues [4] in 1964. It is an uncommon malformation of the aortic arch system defined as a complete disconnection between one subclavian artery and aorta with persistent connection to the ipsilateral pulmonary artery through a patent or non-patent ductus arteriosus. It is always observed opposite to the side of the aortic arch and does not form a vascular ring as the ductus and the isolated subclavian artery are lying on the same side. The left subclavian artery is more frequently involved than the right one (4:1) and more commonly associated with intracardiac anomalies(VSD, TOF, d-TGA) or a syndrome. It is of peculiar importance in patients with decreased pulmonary blood flow as in TOF to identify this anomaly before considering a Blalock-Taussing Shunt as the anastomosis is usually performed on the opposite side of the aortic arch where this anomaly may cause hinderance in performing the shunt surgery [5].

Embryologically the distal part of the RSCA originates from the 7th intersegmental artery while its proximal part is derived from the right 4th aortic arch and the proximal right dorsal aorta (Fig 1). The distal part of the right dorsal aorta normally undergoes involution. The Isolated subclavian artery can be understood using the EDWARD'S hypothetical arch plan [6] which describes interruption at two levels of the aortic arch: 1. The right 4th arch between the aortic sac and the right dorsal aorta and 2. The right dorsal aorta proximal to the right ductus arteriosus (6th arch)(Fig 2). When the right ductus arteriosus remains patent , it connects the right PA to the RSCA and when the right ductus arteriosus regresses, it results in Isolation of the RSCA.

Clinically Isolation of the RSCA is usually aymptomatic, recognised incidentally during evaluation of an associated cardiac lesion or it may also have diminished pulses and blood pressure in the involved extremity. There may be occurrence of "Subclavian steal syndrome" in isolated RSCA, as the subclavian fills from collaterals of the vertebra-

basilar system. Similarly it can also result in "Pulmonary steal phenomenon" when the right ductus arteriosus is patent where the PA fills retrogradely from the vertebro-basilar system stealing from the circle of Willis owing to lower pulmonary vascular resistance and such patients are at high risk of pulmonary overcirculation which was quite seen in our patient. Nath [7] described a similar patient presenting at 18 months with secondary pulmonary hypertension who underwent repair but died at 3 years with autopsy showing grade IV pulmonary vascular disease. In patients with high pulmonary vascular resistance, there may be antegrade flow from the PA to the RSCA resulting in differential cyanosis with diminished oxygen saturation in the right upper extremity [8].

Diagnosis of Isolated RSCA from PA is very difficult as these patients are mostly asymptomatic and because of the development of good amount of collaterals from the contralateral subclavian artery through the vertebral arteries. However with the help of imaging techniques like Echocardiography and angiography diagnosis can be made. Angiography is diagnostic showing delayed opacification of the RSCA or the PA (if ductus is open). Selective angiography of the head and neck vessels delineates the collateral circulation which was done in our patient. MRI and Digital subtraction angiography can be used for rapid accurate non-invasive assessment of the arch anomalies [8].

Indications for Intervention [2]:

1. In patients with "Pulmonary steal phenomenon" who are prone for pulmonary overcirculation leading to features of pulmonary vascular obstructive disease.
2. To prevent Subclavian steal syndrome in adult age.
3. When associated with other intracardiac anomalies requiring correction (VSD, TOF, D-TGA).

Treatment Includes [8,10]

1. Surgical re-implantation of the Isolated RSCA to either the carotid artery or the aortic arch directly or with bypass graft.
2. Ligation of the RSCA or the connecting artery to the right PA can also be done to prevent subclvian steal syndrome later on.
3. Transcatheter coil occlusion of the patent ductus arteriosus
4. Close observation can also be appropriate in patients with no symptoms and in the absence of any additional cardiac anomalies requiring intervention because of the development of adequate collateral circulation as described by Madan et al [9]. However they may require surgical intervention at any time of the development of symptoms of subclavian steal syndrome.

Conclusion

Isolated RSCA from the right pulmonary artery is a very rare vascular arch anomaly and difficult to diagnose clinically with needs of high grade of suspicion while investigating for the same. Early intervention is advocated with complete surgical repair so as to normalise the patient's circulation with resolution of both the pulmonary and subclavian steal phenomenon.

References

1. Stewart JR, Kincaid OW, Titus JL. Right aortic arch: plain film diagnosis and significance. Am J Roentgenol Radium Ther Nucl Med 1966; 97:377-89.
2. Stephen G. Miller, Michael J. Campbell, Piers C.A. Barker, Kevin D. Hill. Isolated right subclavian artery arising from the right pulmonary artery via a right sided ductus arteriosus with associated pulmonary steal phenomenon. Cardiology in the Young 2012; 22:216-8.
3. Eugene Baudet, Xavier F. Roques, Jean-Philippe Guibaud, Nadine Laborde, Alain Choussat. Isolation of the Right Subclavian Artery. The Society of Thoracic Surgeons 1992; 53:501-3.
4. Stewart JR, Kincaid OW, Edwards JE. Malformations with left aortic arch (group II). "Isolation" of the right subclavian artery from aorta. In: An Atlas of vascular rings and related malformations of the aortic arch system. 1st ed. Springfield, IL:Charles C. Thomas, 1964:76-9.
5. Lansing AM, Murphy J. Origin of the left subclavian artery from the pulmonary artery with congenital subclavian steal. Surgical implications in cyantic patients. Ann Thorac Surg 1968; 5:146-52.
6. Edwards JE. Anomalies of the derivatives of the aortic arch system. Med Clin North Am 1948; 33: 925-49.
7. Nath H. Isolation of the right subclavian artery. Am J Roentgenol 1989; 152:430-1.
8. Jackmerry Mosieri, Kavitha Chintala, Ralph E.

Delius, Henry Walters, Mehdi Hakimi. Abnormal Origin of the Right Subclavian artery from the Right Pulmonary artery in a patient with D-Transposition of the Great Vessels and Left Juxtaposition of the Right Atrial Appendage: An unusual anatomical variant. J Card Surg 2004; 19:41-4.

9. Madan N, Schneider DJ, Jacobs ML. Right aortic arch, isolated left subclavian artery and ductus arteriosus with normal intracardiac anatomy: Rare manifestation of chromosome 22q11 deletion. Pediatr Cardiol 2006; 27:781-3.

10. Matthew A. Crystal, Shannon M. Rivenes, Frank F. Ing. Unmasking of an Isolated Right Subclavian artery from the Pulmonary Artery after device occlusion of a Patent Arterial Duct. Catheterization and Cardiovascular Interventions 2013; 82:581-4.