Anomalous origin of the right subclavian artery from main pulmonary artery

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**Learning Objectives**

To describe a rare anomalous origin of right subclavian artery from main pulmonary artery in a patient with normal left aortic arch.

In this poster we demonstrate the contribution of multidetector computerized tomography (MDCT) in providing assessment of an extremely rare congenital cardiovascular anomaly.

**Background**

Aberrant right subclavian artery as the last brachiocephalic branch of the aortic arch is not infrequent, and in fact, is the most common congenital anomaly of left aortic arch[1,2]. (Fig.1 and 2) There were various reports on abnormal origins of subclavian artery from the aortic arch[1-10]. However, abnormal origin of subclavian artery from main pulmonary artery is extremely rare[8].

Anomalous origin of subclavian artery which loses its connection with the aorta and arises from pulmonary artery is an uncommon anomaly of the aortic arch and is called isolation of a subclavian artery. The isolated subclavian artery connects to pulmonary artery via an ipsilateral ductus arteriosus which may be closed or patent.

Isolation of subclavian artery more commonly involves left subcalvain artery and usually associates with intracardiac or aortic arch anomaly [3]. Anomalous origin of right subclavian artery from pulmonary artery is an extremely rare type of aortic arch malformation and in most cases are associated with intracardiac or aortic arch anomaly [3,4,5,6,7]. Our case is a case of anomalous origin of right subclavian artery from main pulmonary artery with left aortic arch.

**Images for this section:**
**Fig. 1:** Left aortic arch with aberrant right subclavian artery in an 8-day-old infant. Volume-rendered image from a CT angiogram seen from the front shows right carotid artery (1), left carotid artery (2), left subclavian artery (3), and aberrant right subclavian artery (4) as the last branch of the aortic arch.
Fig. 2: Left aortic arch with aberrant right subclavian artery in the same patient in fig.1. Volume-rendered image from a CT angiogram seen from the back shows right carotid artery (1), left carotid artery (2), left subclavian artery (3), and aberrant right subclavian artery (4)
Imaging Findings OR Procedure Details

We present MDCT angiography in an 18-month-old child with anomalous origin of right subclavian artery from main pulmonary artery. Related literature was also reviewed.

Case Report

An 18-month-old child presented with symptoms of congestive heart failure and persistent continuous murmur at left parasternal border. Echocardiography revealed continuous turbulent flow in main pulmonary artery, suspicious for abnormal vascular connection causing left to right shunt. The patient was then referred for cardiac CT angiography.

Electrocardiographic (ECG) gating cardiac CT angiography shows left aortic arch which has normal connection with left ventricle. The aortic arch gave rise to three major branches: right carotid artery, left carotid artery, and left subclavian artery. (Fig.3) No connection between right subclavian artery and the aortic arch was seen. There was abnormal connection of right subclavian artery to distal main pulmonary artery just before its bifurcation. (Fig.4 and 5) Right subclavian artery gave rise to right vertebral artery, right thyrocervical trunk, right costocervical trunk and right internal mammary artery before it continued to supply right upper extremity. Multiple collateral vessels were seen connecting these branches with enlarged and tortuous upper right intercostal arteries which arose from the descending aorta. (Fig.6) It was also noted that the right subclavian artery has higher density than the main pulmonary artery and was isodense with the aortic arch. (Fig. 5) Therefore, reversal flow within the right subclavian artery was suspected. (Fig.7)

In our review of related literature, hemodynamic manifestations of subclavian artery isolation depend on the state of associated ductus arteriosus, extent of associated intracardiac anomalies and pulmonary vascular resistance. In cases with obliterated ductus arteriosus, the isolated subclavian artery is supplied by collaterals commonly from the contralateral subclavian artery and ipsilateral vertebral artery which is called subclavian steal phenomenon [4]. In cases with patent ductus arteriosus or patent connection of subclavian artery to pulmonary artery, a left to right shunt could be demonstrated as pulmonary vascular resistance is normally lower than systemic vascular resistance. This shunt would accentuate the degree of steal from vertebral artery. On the other hand, when pulmonary vascular resistance is high, usually secondary to associated lesions, there may be antegrade flow from pulmonary artery to subclavian artery (right to left shunt). In a case with right to left shunt, selective cyanosis of the arm may be demonstrated on physical examination [4,6,10]. In our case we found patent connection between right subclavian artery and main pulmonary artery which poses possibility of
left to right shunt. Isodense appearance of right subclavian artery with the aortic arch on cardiac CT angiogram also supports this suspicion. Hence, we performed Doppler sonography on this patient to confirm reversal flow in right subclavian artery.

Doppler sonography demonstrated complete retrograde flow in right vertebral artery and proximal part of right subclavian artery toward main pulmonary artery, suggestive of a pulmonary steal phenomenon [5,7]. (Fig. 8 and 9) This retrograde filling of main pulmonary artery from anomalous connection of right subclavian artery represented left to right shunt. Because of this steal phenomenon, the patient has a potential for pulmonary overcirculation, vertebrobasilar insufficiency and weakness of right upper limb. Measurement of both upper extremities showed relatively smaller size of right upper limb but the patient did not have symptoms of vertebrobasilar insufficiency which rarely occurs in young patients.

With CT angiography and Doppler sonography findings, the patient was appointed for elective angiography about 2 months later. Angiography revealed pulmonary steal phenomenon through the isolated right subclavian artery.(Fig.10) Retrograde opacification of right vertebral artery from left vertebral artery across the vertebrobasilar junction was observed during the early phase of selective left vertebral angiography. Retrograde flow from right vertebral artery into intrathoracic portion of right subclavian artery and into main pulmonary artery was shown on slightly later phase of the angiography. (Video) No flow into the right subclavian artery distal to the origin of right vertebral artery was seen. Transcatheter vascular occlusion was then performed and the connection between the isolated right subclavian artery and the main pulmonary artery was successfully disconnected, obviating the need for surgical treatment.

Images for this section:
**Fig. 3:** Volume-rendered image from a CT angiogram seen from the front shows normal left aortic arch with right carotid artery as the first arch vessel (1). Left carotid artery (2) is the second arch vessel and left subclavian artery (3) is the third.
Fig. 4: Volume-rendered image from a CT angiogram seen from the back shows right subclavian artery (S) with no connection to the aortic arch but is contiguous with main pulmonary artery instead. 1, right carotid artery. 2, left carotid artery. 3, left subclavian artery.
**Fig. 5:** Maximal intensity projection (MIP) image from a CT angiogram shows connection of right subclavian artery (S) to main pulmonary artery (PA). The right subclavian artery, however, shows higher density than the pulmonary artery and is isodense with the aortic arch (A). Right vertebral artery (V) is also shown.
Fig. 6: Maximal intensity projection (MIP) image from a CT angiogram shows enlarged and tortuous upper right intercostal arteries and cervical artery (arrow) which are thought to give collateral blood flow to right subclavian artery.
Fig. 7: Diagram showing possible hemodynamic manifestation of the abnormality. Right subclavian artery and right vertebral artery are suspected to have retrograde flow filling into main pulmonary artery. RPA, right pulmonary artery. RSCA, right subclavian artery. RVA, right vertebral artery. RCA, right carotid artery. LCA, left carotid artery. LSCA, left subclavian artery. LPA, left pulmonary artery. LV, left ventricle. RV, right ventricle.
Fig. 8: Pulsed wave Doppler image of right vertebral artery shows complete reversed flow.
**Fig. 9:** Color Doppler ultrasound at proximal part of right subclavian artery shows flow direction toward the pulmonary artery.
**Fig. 10:** Image from selective left vertebral angiography shows retrograde opacification of right vertebral artery (arrowhead) from left vertebral artery (open arrow) across the vertebrobasilar junction and into main pulmonary artery (arrow).
Fig. 11: Video from selective left vertebral angiography shows retrograde opacification of right vertebral artery from left vertebral artery across the vertebrobasilar junction and into main pulmonary artery.
Conclusion

Anomalous origin of right subclavian artery from main pulmonary artery is extremely rare. This anomaly can cause congestive heart failure and subclavian steal syndrome. MDCT angiography is relatively noninvasive for young children and can provide adequate anatomical details for surgical treatment planning. It is also useful in predicting hemodynamic manifestations of cardiovascular anomalies using its multiplanar and three-dimensional capabilities. Moreover, it is a better and quicker examination for infants and young children, especially those who have a limited ability to hold still and those who need close monitoring. Compared with other available imaging modalities, better global assessment of intrathoracic structures is also obtained by MDCT.

Personal Information

References

References: