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Case Series

Computed tomography angiography in anomalous left coronary artery from pulmonary artery: a case series

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ABSTRACT

Anomalous left coronary artery from pulmonary artery (ALCAPA) is a rare congenital anomaly with a high rate of mortality in first year of life. It is one of the most common causes of cardiomyopathy and myocardial ischemia in children. In the present scenario, computed tomography (CT) coronary angiography is sufficient for the diagnosis of ALCAPA, eliminating the need of invasive angiography. CT coronary angiography performed in infants with suspicion of ALCAPA is technically challenging in terms of acquisition techniques, radiation protection and diagnostic interpretation. In this case series, we present five cases of ALCAPA diagnosed on CT coronary angiography. CT coronary angiography was able to delineate the anomalous origin of left coronary artery (LCA) from pulmonary artery in all cases. Anomalous LCA was noted to arise from posterior pulmonary sinus of pulmonary trunk in four cases and main pulmonary artery in one case. CT angiography also identified the extent of inter-coronary collateral vessels, left ventricular dysfunction and signs of pulmonary hypertension.

Keywords: ALCAPA, Coronary angiography, Congenital, Coronary anomalies, Paediatric

INTRODUCTION

The incidence of ALCAPA is widely recognized as 1 in 300,000 newborns. High mortality is associated with this anomaly, with 90% of patients not surviving beyond 1st year of life.[1] As the pulmonary pressure decreases after 8th week of life, the flow in LCA reverses into the pulmonary artery leading to formation of left to right shunt. This is also known as coronary steal phenomenon. The prognosis depends on formation of collateral circulation between RCA and LCA which determines the extent of myocardial ischemia.[2]

ECG and transthoracic echocardiography are the initial investigations performed which raises the suspicion of ALCAPA. Definitive diagnosis traditionally required catheter angiography with many of the cases being diagnosed on autopsy. With the advent of ECG gated multi slice computed tomography for coronary angiography, the definitive diagnosis and surgical planning can be carried out without the need of invasive catheter angiography.[3] In this case series, we describe cases of five infants diagnosed with ALCAPA on CT coronary angiography from January to December 2021.

CASE SERIES

Computed tomography angiography was performed in six infants with echocardiography findings suspicious of ALCAPA over a period of six months. Informed consent was taken from parents of all patients. ECG gated Ultrahigh pitch prospective ECG gated CT angiography was acquired on 128 slice dual source (Siemens Somatom flash) CT. Iodinated contrast-2 ml/kg (Iobitridol- 350

mgI/100 ml) at 1.2 ml/s was injected via 24G cannula in right antecubital vein using pressure injector followed by saline flush. Manual bolus tracking was used for triggering the scan to ensure adequate contrast opacification of both ventricles in single phase. It is important to note that while LCA arises from pulmonary artery in these patients, pulmonary angiography study is often unable to delineate the pathology because of left to right shunt which develops after formation of inter coronary collaterals.

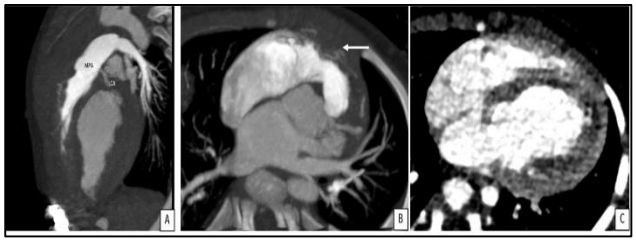

Figure 1 (A-C): Oblique sagittal MIP CT angiogram image showing LCA arising from posterior aspect of main pulmonary artery. Axial MIP CT angiogram image showing multiple tortuous collateral arteries (←) in anterior aspect coursing from right coronary artery to branches of LCA. Axial CT image-shows subendocardial ischemia along left ventricular wall in form of hypo-enhancement.

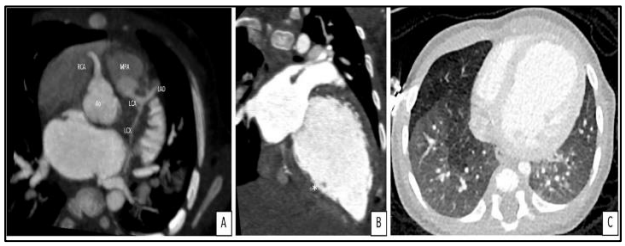

Figure 2 (A-C): Oblique axial MPR image showing LCA arising from posterior sinus of main pulmonary artery and right coronary artery (RCA) arising from right coronary sinus of aorta. Oblique MPR CT image showing dilation of left ventricle and left atrium with decreased enhancement in subendocardial aspect of left ventricular wall (*). Axial CT image in lung window-shows mosaic attenuation in bilateral lower lobes of lungs.

Adequate quality of computed tomography images was obtained in all cases. Multiplanar reformatted (MPR) images and maximum intensity projection (MIP) images were reviewed in all cases and evaluation of origin of RCA, LCA, inter coronary collaterals, ventricular size and function, ischemic changes and signs of pulmonary hypertension was carried out.

ALCAPA identified in 6 patients on CT angiography. The mean age of patients was 5.8 months, with five patients under the age of 12 months. Five patients were male while 1 was female. Clinical presentation was similar in all patients, with failure to thrive being the most common presenting complaint. Decreased left ventricular ejection fraction noted in all cases.

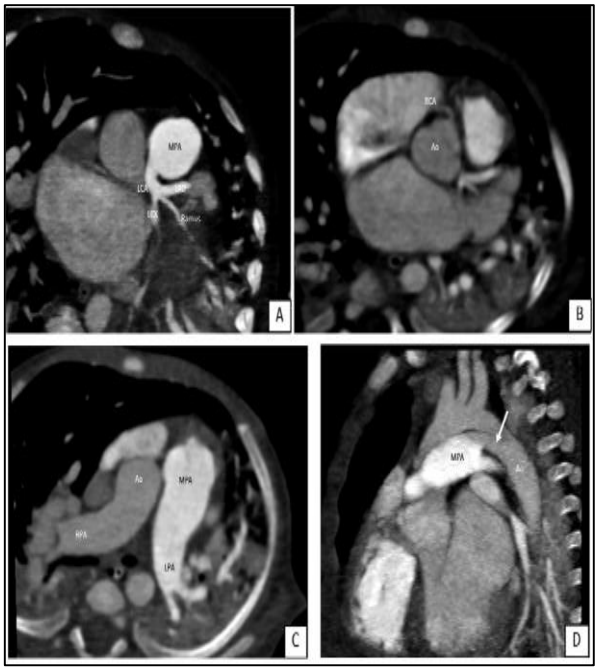

Figure 3 (A-D): Oblique axial MPR image showing LCA arising from right posterolateral aspect of main pulmonary artery, which is trifurcating into LAD, LCX and ramus intermedius. Oblique axial MPR image showing right coronary artery arising from right anterior coronary sinus of Aorta (attenuated in caliber). Oblique axial MPR image showing anomalous right pulmonary artery arising from aorta, and left pulmonary artery arising from MPA. Oblique sagittal MIP image showing good sized tubular patent ductus arteriosus between MPA and descending aorta.

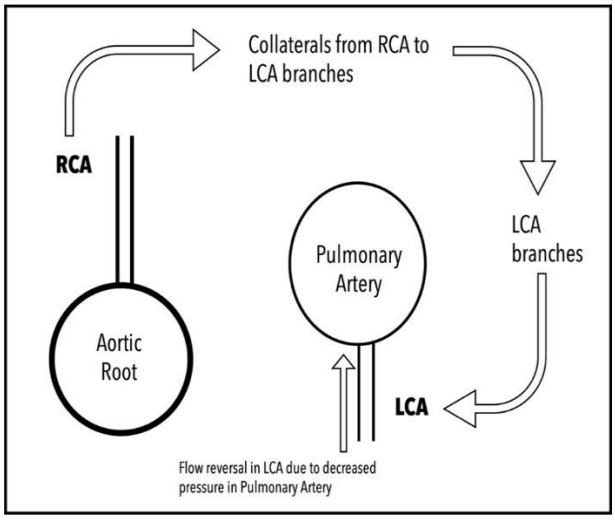

Figure 4: Pathogenesis in ALCAPA syndrome (RCA-right coronary artery, LCA-Left coronary artery).

Anomalous LCA was noted to arise from posterior sinus of main pulmonary artery in five cases (Figure 1 and 2) while in one case, it was noted to arise from right posterolateral aspect of main pulmonary artery above the sino-tubular junction. This case also revealed anomalous right pulmonary artery arising from aorta along with tubular patent ductus arteriosus (Figure 3).

Right coronary artery showed normal origin from right coronary sinus of aorta in all cases, and revealed normal caliber in four cases. Attenuated caliber of RCA was noted in one case while another revealed ectatic caliber.

Significant intercoronary collaterals were noted in the case with ectatic RCA (Figure 2), while others revealed minimal intercoronary collaterals.

Table 1: Patient characteristics and imaging findings.

Age (months)	Sex	Clinical presentation	LVEF (%)	LCA origin	RCA origin, course	Coronary dominance	Inter coronary collaterals	LV cavity size (cm)	Pulmonary hyper-tension	Associated abnormalities
7	M	Failure to thrive	40	Posterior sinus of MPA	Normal origin, ectatic	Right	Significant inter coronary collaterals	2.1	No	None
5	M	Poor feeding, failure to thrive	20	Posterior sinus of MPA	Normal origin, course	Right	No	3.1	Yes	None
13	M	Failure to thrive	30	Posterior sinus of MPA	Normal origin, course	Right	Minimal	4.3	Yes	None
3	F	Poor feeding	15	Posterior sinus of MPA	Normal origin, course	Right	Minimal	3.0	Yes	None
6	M	Recurrent pneumonia	30	Right posterolateral aspect of MPA	Normal origin, attenuated	Left	No	2.8	Yes	Anomalous right pulmonary artery from aorta, tubular PDA
1	F	Failure to thrive	40	Posterior sinus of MPA	Normal origin, course	Right	Minimal	2.1	No	None

Coronary circulation was right dominant in five cases, and left dominant in one case.

Signs of pulmonary hypertension were identified in four cases in form of dilated pulmonary trunk (MPA: aorta>1) and mosaic attenuation in bilateral lungs.

Dilated caliber of left ventricle was noted in all cases with decreased left ventricular ejection fraction. Subendocardial ischemia was noted in five cases (Figure 1 and 2).

DISCUSSION

ALCAPA syndrome also known as Bland-Garland-White syndrome, is a rare congenital anomaly of the coronary arteries. Only 10-15% patients survive beyond childhood.[1] The pathogenesis of ALCAPA involves left to right shunt developed after pulmonary pressure decreased in neonatal period of life. Blood flows from LCA to pulmonary artery leading to reduced perfusion of left ventricle. This is known as "coronary steal" phenomenon (Figure 4). Consequently, arterial collateral vessels develop in response to left ventricular ischemia from right coronary artery to branches of LCA. Two type of the disease process have been described based on the collateral vessels-the Infant type in which the poor development of collateral vessels lead to ischemic cardiomyopathy of left ventricle and high rate of mortality, and the adult type in which there are multiple collateral vessels between RCA and LCA branches, leading to a better outcome. Though there is increase rate of survival in adult type ALCAPA, the risk of sudden cardiac death due to chronic myocardial ischemia and dysrhythmias is very high. Dilated and tortuous coronary arteries are noted in the adult type ALCAPA syndrome.[4]

In cases of infants with suspicion of ALCAPA syndrome, CT coronary angiography should be performed with ECG gating and proper sedation of the infant. It should be ensured that both aorta and pulmonary arteries are opacified on the CT scan, unlike in conventional CT coronary angiography where the contrast opacification of aortic root is the main aim.[6] CT scan acquired in pulmonary angiography phase does not provide adequate visualization of the LCA arising from pulmonary artery since there is reversal of flow in LCA because of pressure gradient.

Anomalous LCA was noted to arise from posterior pulmonary sinus of pulmonary trunk in four cases and main pulmonary artery in one case.

Among our cases, the first case had good development of arterial collateral vessels between RCA and branches of LCA. Consequently, the extent of left ventricular dysfunction was less compared to the other cases. This is an indicator of relatively better prognosis and post-operative outcome.

As a consequence of the left to right shunt, pulmonary hypertension can develop in the setting of ALCAPA syndrome. It usually develops when there are abundant inter coronary collateral arteries.[7] Signs of pulmonary hypertension seen on CT include dilated pulmonary trunk, mosaic attenuation in lung fields and dilated right atrium and ventricle.[8] We observed signs of pulmonary hypertension in three cases. However, we observed that pulmonary hypertension developed in both type of cases with good as well as poor inter coronary collaterals.

We found one patient with associated anomalous origin of right pulmonary artery from aorta and patent ductus arteriosus.

The standard treatment for ALCAPA syndrome is surgical correction with the aim of the surgery to restore a two-coronary-artery circulation system. In infants, early surgical correction should be prioritized to reduce irreversible changes in left ventricular function.[9,10]

CONCLUSION

CT coronary angiography can provide accurate diagnosis with remarkable anatomical detail, reducing the requirement of invasive diagnostic procedures. It can also characterize left ventricular function and identify inter coronary collateral arteries, pulmonary hypertension, and other associated congenital anomalies.

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