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DUAL LEFT ANTERIOR DESCENDING ARTERY WITH ANOMALOUS ACCESSORY VESSEL FROM PULMONARY ARTERY

Radiodiagnosis

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ABSTRACT

The incidence of the congenital coronary anomalies is quite uncommon. We report a case of dual LAD variant with anomalous origin of accessory LAD from pulmonary artery detected with the help of multidetector CT coronary angiography. It is associated with coronary steal phenomenon and pulmonary stenosis. To our knowledge, this case report is first of its kind making it unique and different from dual LAD variants described till now and not reported yet in the literature.

KEYWORDS

anomalous origin, dual LAD, Pulmonary artery stenosis, CT angiography, coronary steal

An 8 year old male child presented to us with recurrent episodes of respiratory tract infections, effort intolerance and diaphoresis on minimal exertion since birth.

Physical examination revealed mild tachycardia and ejection systolic murmur in the pulmonary area. Electrocardiogram revealed findings of right ventricular hypertrophy.

On transthoracic echocardiography (TTE) there was severe valvular pulmonary stenosis (74 mm Hg pressure gradient across pulmonary valve) with mild pulmonary hypertension. It revealed multiplecoronary fistulae; from LAD to right ventricularoutflow tract (RVOT), pulmonary artery (PA) and right ventricle (RV) cavity, dilated left main coronary artery and short LAD feeding the fistula

The child was referred for CT Coronary angiography to delineate coronary anatomy. It revealed a short Left main coronary artery (LMCA) arising normally from left coronary sinus and bifurcating almost immediately into LAD and left circumflex artery (LCX) (Figure.1). LAD was very short and coursed in proximal anterior interventricular sulcus terminating after giving rise to ventricular branches and few collaterals to an anomalous vessel arising from pulmonary artery.

An anomalous vessel was seen arising from the main pulmonary artery from its anterior wall just above the valve. It was dilated, tortuous and continued distally in the anterior interventricular sulcus (Figure.2) giving branches to both the ventricles. It also received collaterals from the small main LAD (Figure.3). The density of contrastin accessory LADwas higher than that in the PA with a jet of high density contrast seen near its origin at the pulmonary artery suggesting flow reversal or coronary steal phenomenon (Figure.4).

Thus the final diagnosis of dual LAD with anomalous accessory LAD originating from PA with coronary steal phenomenon and associated pulmonary stenosis was made.

In our case, balloon pulmonary valvulolasty was done for pulmonary stenosis in the hope that elevated pulmonary artery pressures would reduce the steal phenomenon. Post valvuloplasty, pressure gradient across pulmonary valve dropped down from 74 mm Hg to 18 mm Hg. Since this Dual LAD shows ALCAPA morphology, hence surgical bypass from aorta to this vessel using LIMA/RIMA or saphenous vein, after ligation of its ostium was planned as definitive treatment.

DISCUSSION:

Congenital coronary artery abnormalities are rare, occurring in only 1% to 2% of the population. Variations in origin, course, and distribution of the left anterior descending coronary artery (LAD) are rarer, though such variations are fairly common in the right coronary artery.

Duplication of the LAD artery in otherwise normal hearts has been reported to occur in 0.13%–1% of the general population(1). Duplication of the LAD artery consists of a short LAD artery, which courses and terminates in the anterior interventricular sulcus(AIVS) without reaching the apex, and a long LAD artery, which originates from either the LAD artery proper or the RCA, then enters the distal anterior interventricular sulcus and courses to the apex(1).Spindola-Franco and co-authorsangiographically describedfour important variants of dual LAD (2)andin none of these types second LAD arises from PA. Newer variants have also been described based on CT angiography in literature other than those mentioned in classification described by Spindola et al (3).

Anomalous origin of the coronary artery from the pulmonary artery (ALCAPA) has an estimated prevalence of one in 300,000 live births(4). Approximately 90% of untreated infants die in the 1st year of life, and only a few patients survive to adulthood (5). Its most common type is the one where the left main coronary artery (LMCA) arises from the PA and right coronary artery (RCA)arises normally from the aorta and is called Bland-White-Garland syndrome(6). Conventional or CT Coronary angiography usually helps confirm the diagnosis of ALCAPA by demonstrating collateral circulation between the RCA and LCA and a coronary "steal" into the PA(7).There are two types of ALCAPA syndrome: the infantand the adult type, each having different manifestations and outcomes. In infantile type there are no collaterals between RCA and LCA and both are normal in calibre. In adult type multiple collaterals develop between RCA and LCA and both are dilated owing to increased volume. ALCAPA syndrome results in the "coronary steal" phenomenon, in which a left-to-right shunt leads to abnormal left ventricular perfusion. The presence of extensive intercoronary collaterals, systemic collateral supply or restrictive opening between the LCA and pulmonary trunk is believed to permit some ALCAPA affected persons to remain asymptomatic (8)

The pitfall in the earlier transthoracic echocardiography (TTE) diagnosis of coronary fistulae was due to visualization of normal origin of LMCA from left coronary sinus and its bifurcation into LAD and LCX. However this anomalous vessel is not a coronary fistulous tract because of its typical long and complete course from PA to the apex of heart in the anterior interventricular sulcus similar to that of LAD. Moreover, had it been fistula from LAD to PA the normal LAD would have been dilated which is not seen in our case. So the possibility of coronary fistula was ruled out.

Another differential is ALCAPA. Hallmark of ALCAPA is direct visualization of LMCA from PA and dividing into LCX and LAD, coronary steal phenomenon. Isolated origin of LAD from PA with LCX originating from left coronary sinus has also been described in literature. There is also a case report of anomalous isolated origin of LAD from pulmonary artery (ALADAPA)(9). However in that report the vessel arising from left coronary ostium was LCX morphologically.

The artery arising from the PA was considered to be accessory LAD because it follows the course of LAD despite the presence of normal however short LAD arising from LMCA. However it behaves like an ALCAPA as it shows coronary steal phenomenon.

CONCLUSION:

Our case is unique in that it describes, in an 8 year old child, a rare variant of dual LAD with ananomalous accessory LAD from pulmonary artery. The importance of this variant is that it can be misdiagnosed or misinterpreted as coronary fistulae on imaging and it can cause coronary steal phenomenon and associated with pulmonary stenosis as seen in our case which can be lethal if undetected on imaging. To our knowledge such case is not reported in literature so far.

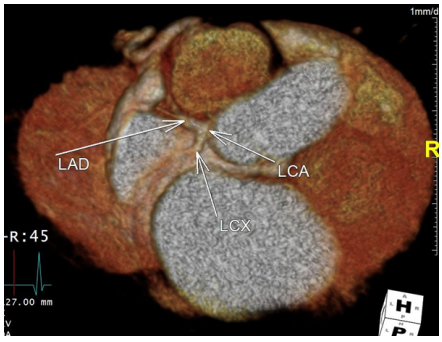


FIGURE 1- MDCT image showing Left main coronary artery (LMCA) arising normally from left coronary sinus and bifurcating immediately into left anterior descending artery (LAD) and left circumflex artery (LCX) shown by white arrows.

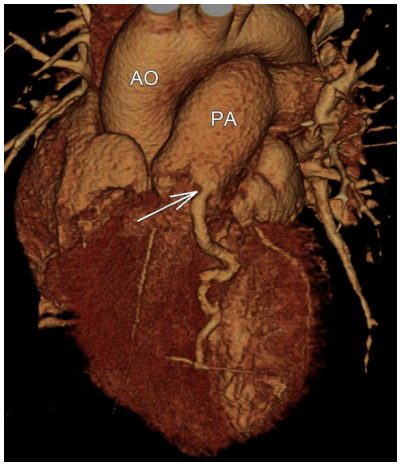


FIGURE 2- MDCT image showing origin of anomalous accessory LAD (shown by white arrow) from pulmonary artery coursing in anterior interventricular sulcus

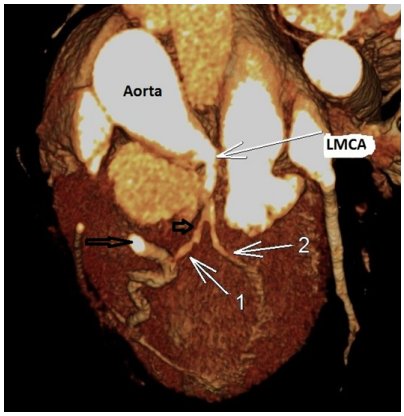
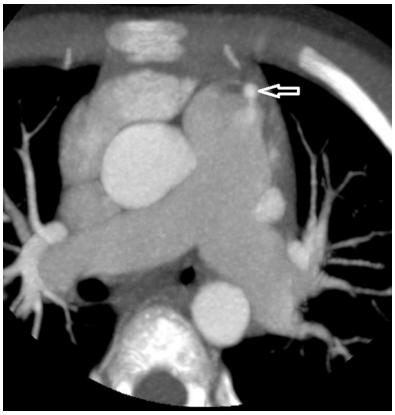


FIGURE 3- MDCT image showing collateral communication between main and accessory anomalous LAD (white arrow 1), white arrow 2 showing left circumflex artery, long open black arrow showing anomalous accessory LAD and short open black arrow showing main LAD

FIGURE 4-MDCT image showing jet of high density contrast (open white arrow) seen at the origin of anomalous LAD from pulmonary artery suggesting flow reversal or coronary steal phenomenon



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