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Case Report

A rare presentation of anomalous origin of left coronary artery from the pulmonary artery in a young adult

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ABSTRACT

Anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) is a very rare congenital cardiac disorder with an incidence of about 1 in 300000 live births. The majority of patients are symptomatic in early infancy with symptoms of intractable heart failure. Only very few patients survive to adulthood with minimal symptoms. A young female was referred to us in view of breathlessness, clinical examination didn't show any signs except a grade 2 systolic murmur in the pulmonary area. The patient underwent 2D echocardiography evaluation which revealed suspicion of ALCAPA and was confirmed using coronary angiography. The patient underwent surgical closure of ALCAPA along with left internal mammary artery (LIMA) to left anterior descending (LAD) anastomosis, postoperative period was uneventful, on routine follow-up the patient is doing well with resolution of dyspnea. ALCAPA is a very rare disorder and most of it is diagnosed in infancy and not many surviving beyond infancy, high degree of suspicion of ALCAPA is needed for young patients presenting with symptoms of angina, heart failure.

Keywords: ALCAPA, Left internal mammary artery, Adult presentation

INTRODUCTION

ALCAPA (anomalous origin of left coronary artery from the pulmonary artery) is a very rare congenital cardiac disorder with an incidence of about 1 in 300000 live births.[1] Majority of patients are symptomatic in early infancy with symptoms of intractable heart failure. Only very few patients survive to adulthood with minimal symptoms. These adult patients can present with exercise induced angina, cardiac failure or even sudden cardiac death.[2] Our patient was a 19-year-old female who had been undiagnosed till now and had only New York Heart Association (NYHA) functional class II symptoms, there are many different techniques to correct ALCAPA, our technique was usage of a patch to close ALCAPA orifice with a LIMA to LAD anastomosis.

CASE REPORT

A young girl who was not previously diagnosed to have any cardiac disease was referred to our center for evaluation of exertional dyspnea. General physical examination was unremarkable except for short stature. cardiovascular examination revealed a systolic murmur over the pulmonary area. Chest X-ray showed mild cardiomegaly so patient was planned for further evaluation.

Investigations

Trans thoracic echocardiography evaluation done was suggestive of anomalous origin of left coronary artery from pulmonary artery. A detailed catheterization study showed evidence of left to right shunt with no significant

pulmonary hypertension. Right coronary artery (RCA) angiogram showed hugely dilated RCA, which filled the left anterior descending artery (LAD) and left circumflex artery (LCX) through multiple collaterals (Figure 1). From the left coronary artery, the contrast drained into main pulmonary artery (MPA). A contrast CT angiogram was done which revealed anomalous origin of LCA from posterior facing sinus of MPA (Figure 2). Another abnormality noted was a bovine aortic arch with aberrant origin of right subclavian artery distal to left subclavian artery.

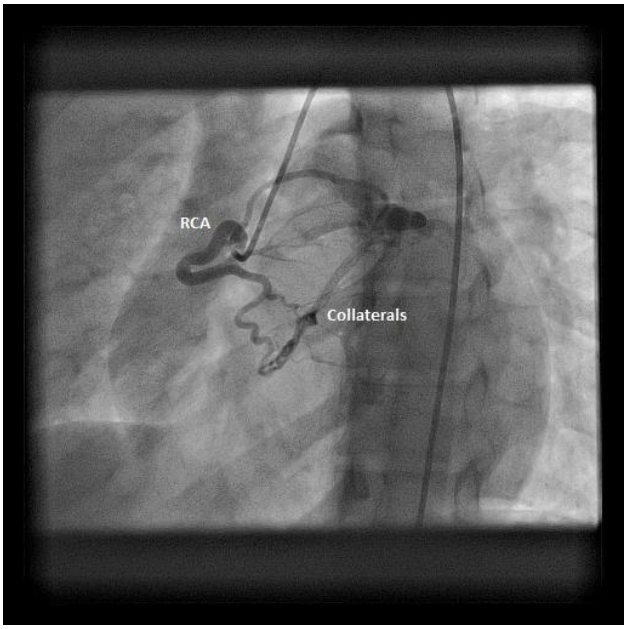

Figure 1: Coronary angiogram showing dilated RCA with collaterals to ALCAPA.

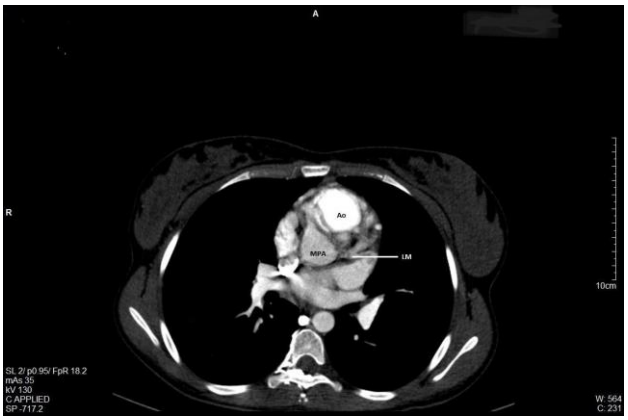

Figure 2: Coronary CT angiogram showing origin of ALCAPA from main pulmonary artery behind the aortal Ao: aorta, MPA: main pulmonary artery, LM – left main coronary artery.

Treatment

The various surgical options were considered and considering that the patient is an adult, it was planned to give an arterial graft to LAD with patch closure of ALCAPA. The presurgical anatomical diagnosis was confirmed. RCA was hugely dilated, giving collaterals to the left system, LCA was arising from the posterior aspect of MPA and trifurcating into LAD, Ramus and LCX, all of which were dilated (Figure 3). The left internal mammary artery was of good quality with excellent flow down its transacted distal end. The distal anastomosis of LIMA to proximal LAD was then constructed using 7/0 prolene with the heart beating. After attaining cardioplegia, MPA was opened just distal to the pulmonary valve. The LCA ostia was identified and further cardioplegia given down it. A piece of bovine pericardium was used for patch closure of the ostium of the LCA, so as to isolate the LCA from MPA and prevent left to right shunt (Figure 4). The pulmonary artery was then closed using 5/0 prolene. On releasing the aortic cross clamp, the heart resumed activity spontaneously in sinus rhythm. The patient was then successfully weaned off from CPB with no rhythm problems.

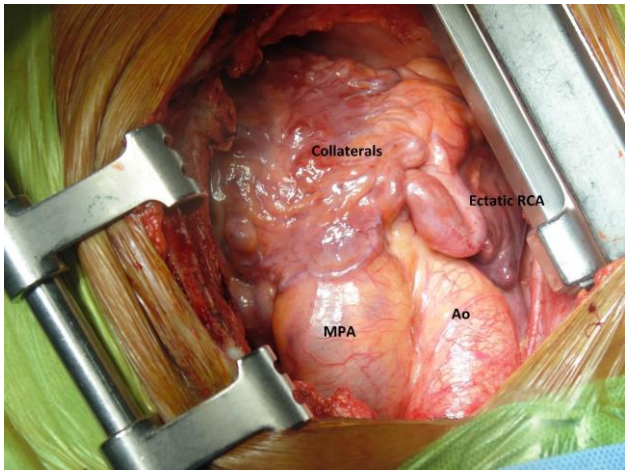

Figure 3: Intraoperative image showing hugely dilated RCA with collaterals.

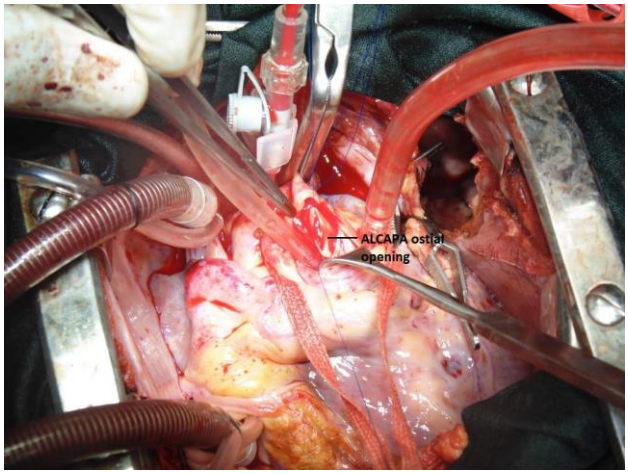

Figure 4: Intraoperative image showing ALCAPA ostium inside the MPA, which was closed with a pericardial patch.

Outcome and follow up

Post operative recovery was good and patient was discharged uneventfully on 10th day. On follow-up visit at the end of 1 year the patient was doing well with no complaints.

DISCUSSION

ALCAPA is a very rare congenital cardiac disorder with an incidence of about 1 in 300000 live births.[1] Almost 90% of patients die within 1 year of life, very rarely they survive beyond infancy. There are two types of ALCAPA the infant type and adult type, adult type generally is associated with development of good collaterals between RCA and LCA and because of the development of these collateral vessels the prognosis in adult type is better than the infant type.[3]

ALCAPA is usually diagnosed in childhood as child presents with symptoms related to angina and cardiac failure. (electrocardiography) ECG in children gives evidence of anterolateral infarct. Echocardiography with Doppler color flow mapping has replaced cardiac catheterization as the standard method of diagnosis. 2D ECHO with color Doppler shows multiple collaterals between RCA and ALCAPA, these collaterals show systolic and diastolic flow which is suggestive of ALCAPA left coronary artery draining to MPA.[4] The other evidences which support ALCAPA include dilated RCA, prominent septal collaterals, dilated left ventricle with systolic dysfunction, regional wall motion abnormalities, mitral regurgitation and fibrosed papillary muscle. Our patient had a dilated RCA and prominent septal collaterals but no other supportive ECHO findings. Color Doppler flow of LCA to MPA could not be clearly visualized since the patient was an adult. This made it mandatory to have further imaging studies which included a cardiac catheterization study and computed tomography.

Various surgical options were considered for this patient. Ligation of the origin of the left coronary artery and reconstitution of flow through it with a subclavian arterial or saphenous venous graft has been successful, grafts using the internal mammary artery have a longer survival and may be desirable in adults.[5]

Direct reimplantation of the origin of the LCA into the aorta (with a button of pulmonary artery around the origin) has been proven successful and is considered the standard approach in many centers.[6] An alternative approach is the Takeuchi procedure, in which an aortopulmonary window is created and a tunnel is fashioned to direct blood from the aorta to the LCA ostium.[7] In this patient, considering the fact that the patient was an adult and had early bifurcation of LCA, the procedure of choice will be an internal mammary graft to LAD and closure of ALCAPA using a Patch within the MPA.

CONCLUSION

ALCAPA should be in the list of differential diagnosis when a young patient presents with myocardial infarction or cardiac failure, even though it is rare for it to present after infancy. There are two types of ALCAPA the adult type and the infant type. If ALCAPA is diagnosed in adults it usually carries a better prognosis than the infantile type. Conventional coronary Angiogram is the confirmatory test to diagnose ALCAPA. LIMA to LAD anastomosis and patch closure of ALCAPA is an alternative to the standard Takeuchi procedure and reimplantation of LAD.

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