ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (ALCAPA)

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ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), is also known as the Bland-White-Garland Syndrome. It is a rare congenital heart lesion mostly seen in infants aged one and rarely seen in adults. A 60 year old female, suffered from chest tightness, shortness of breath, dizziness, palpitations, discomfort accompanied by light headiness especially during walking for a short distance, whilst walking upstairs starting in late February . She had no history of diabetes or hypertension. An Electrocardiogram showed atrial fibrillation and left ventricular hypertrophy. Echocardiography revealed LVD 61mm, LA 41mm, EF 63%, possible coronary pulmonary artery fistula, moderate aortic regurgitation, mild mitral regurgitation and for further treatment she was sent to our hospital. She was advised to undergo multi detector computed tomography examination (MDCT) and was diagnosed with ALCAPA and she was scheduled for a surgery.

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), is also known as the Bland-White-Garland Syndrome. It is a rare congenital heart lesion mostly seen in infants aged one and rarely seen in adults. If it is left untreated and may result in a high mortality rate at the early age.[1] The first known case was of a 2 year old baby reported by Konstantinowitsch in 1906.[2] The incidence of ALCAPA is 1/300 000 live births comprising between 0.24% and 0.46% of total congenital heart disease. It shows typical signs of an antero-lateral myocardial infarction, characterized by abnormal Q waves in I, avL, V5, and V6, leads and there is transient change in ST segment. Around 20-45% of such patients do not show abnormal Q waves, if there is an abnormal R wave progression in the chest leads than ALCAPA should be suspected. Surgical procedures depend on the anatomical anomaly presented in the patient's. The procedure includes re-implantation of the anomalous coronary Ostia, use of extra cardiac arterial blood supply, or the Takeuchi procedure.

Case Detail

A 60 year old female, complained of chest tightness, palpitation, dizziness and breathlessness especially while walking for a short distance and sudden onset while walking upstairs. Her symptoms had been getting worst for a few months and no family history of cardiac problems. Trans thoracic echocardiography done at Suzhou hospital showed possible coronary pulmonary artery fistula measuring around 12 mm (Fig. 1b), moderate aortic regurgitation (Fig. 1a) and increased size of RCA 8 mm (Fig. 1c), abnormal signal in right ventricle and similar results were observed in our hospital (Fig. 1d). The patient's physical

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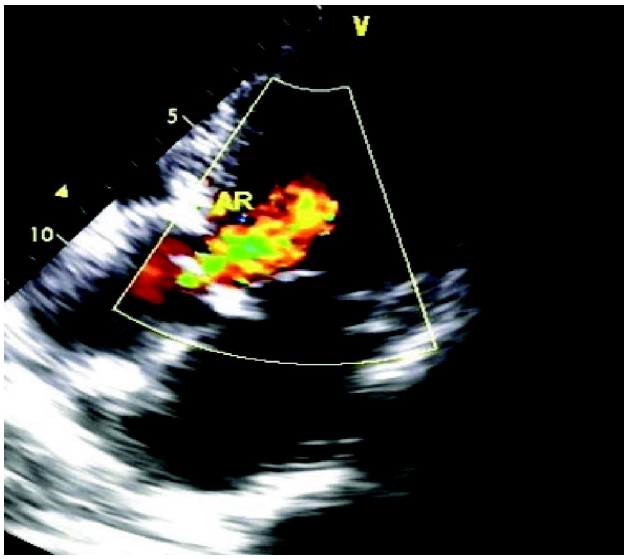

Figure 1A: Mild Aortic Regurgitation

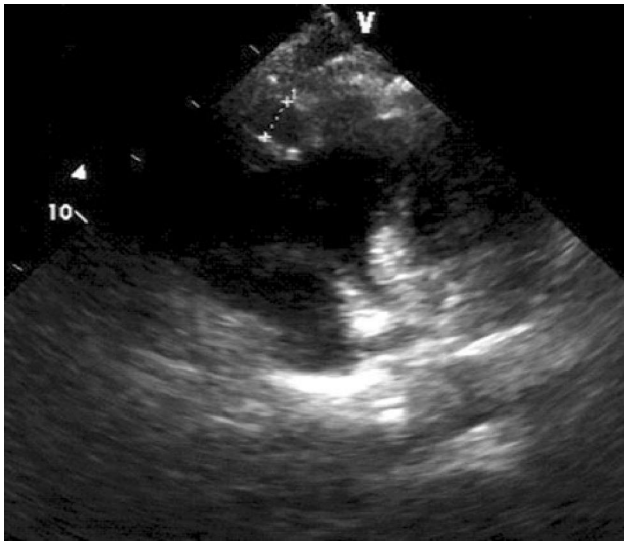

Figure 1B: Blue dots showing abnormal origination of LAD (14 mm)

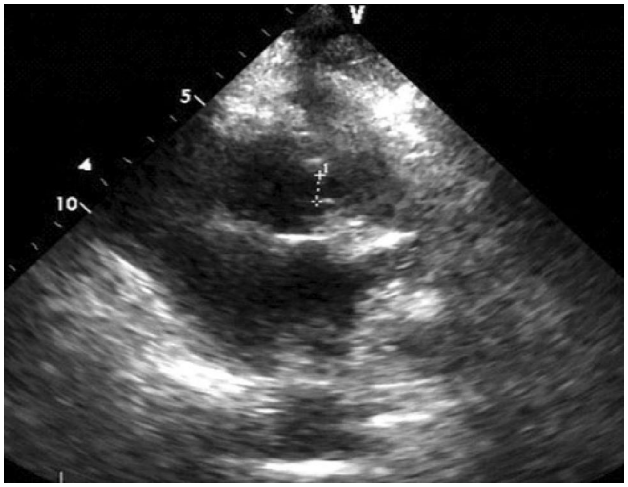

Figure 1C: Increased RCA size measured around 8 mm

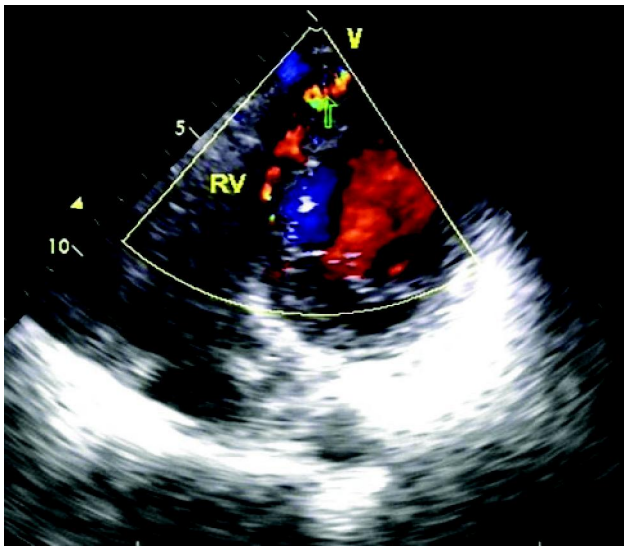

Figure 1D: Abnormal signal seen in the right ventricle

examination showed body temperature was 36.5 degrees Celsius, 110/70 mmhg, and pulse was 70 beats/ min and breathing 18 / mins. An E.C.G showed atrial fibrillation and left ventricular hypertrophy. Auscultation of the chest revealed wet rales and diastolic murmur. Her x-ray showed severe cardio-megaly (Fig. 2) lung congestion. CT angiography was performed using dual Ct and the angiography

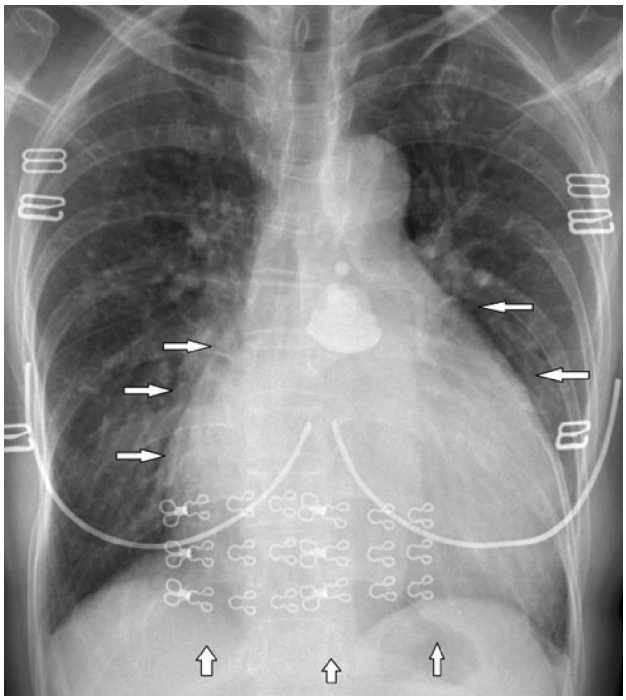

Figure 2: PA chest shows cardiomegaly (arrows) and increased lung markings

showed a large heart window (Fig. 3). 3D (Dimensional) with curved multiplanar reformations (cMPR) images showed single abnormal opening in the pulmonary artery measuring nearly 12 mm. Tortuous and dilated right coronary artery with 8 mm diameter and several collateral vessels formation were observed. (Fig. 4) and the patient was diagnosed with ALCAPA.

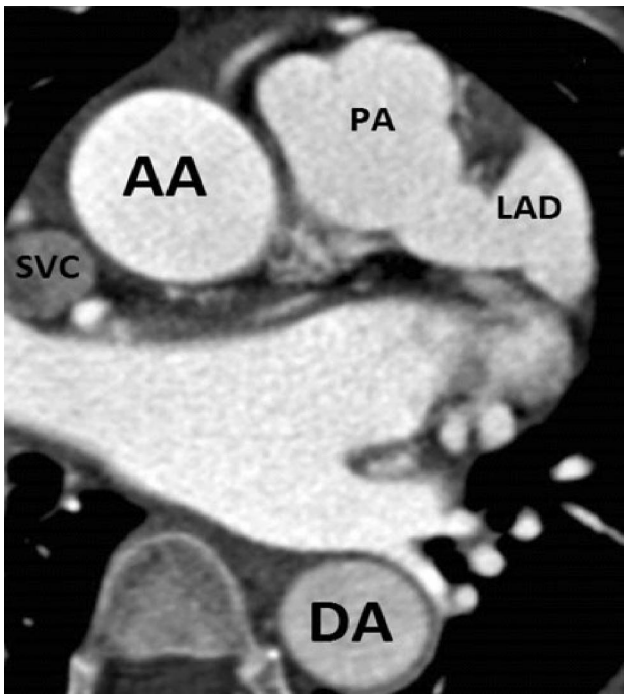

Figure 3: CT angiography multiplanar reformatted image shows abnormal originating of left coronary artery (LAD) from pulmonary artery (PA)

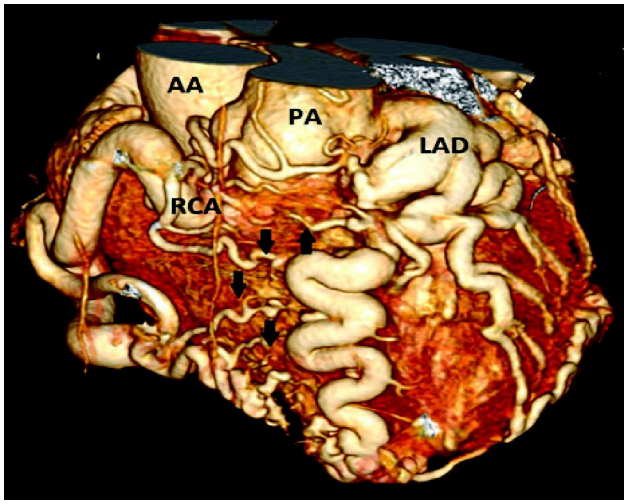

Figure 4: CT angiography volume rendered reformation image shows abnormal ordination of CAD from pulmonary artery,dilated and torturous RCA and increased collateral blood vessels (arrows) connecting the RCA and LAD

Discussion

ALCAPA is a rare heart disease with occurrence of 1 in 300,000 live births which carries a high mortality rate in the first year of life. 90% of infants die without treatment in the first year of life. The main causes of death are myocardial infarction, circulatory insufficiency, left ventricle dysfunction, myocardial ischemia and heart failure because LAD arises from pulmonary artery.[3,4,5] This is interesting case of ALCAPA with late presentation. This patient survived infancy to adulthood. Few cases survive early in the life and make to the adulthood with changes in the heart like cardiomegaly (Fig. 2), collateral vessels development and increased in RCA (Fig 4, 1c) vessel size due to compensating mecha-nism and similar finding are seen in this case with several collateral development and increased diameter of RCA ≥ 8 mm (Fig. 1c) changes. Older ALCAPA pati-ents usually have exertional angina, syncope and dyspnea as their presenting complaints.[6] This case had complains of chest tightness, shortness of breathe, dizziness, palpitations.

Echocardiography is useful in visualizing the ejection fraction, regurgitation and vavular defect however, it's difficult to notice anomaly like abnormal origins of an artery due to poor resolution which are the important findings seen in ALCAPA patients.

Recently conventional coronary angiography(CAG) was the diagnostic method of choice for detecting coronary anomalies. However, CAG may carry some complication due to the invasive procedure.

The advancement in non invasive technologies such as multi-detector computed tomography (MDCT) angiography and MR imaging are very useful in diagnosing the anomalies with high accuracy and are non invasive procedures. MDCT is a noninvasive imaging technique anomalies can be diagnosed easily as the MDCT has good spatial resolution and less time consuming. The advantages of MDCT over CAG is that it takes less time, has a good resolution and does not require hospital stay but it has more dosages as compare to CAG. MRI has long scan time and less spatial resolution compared with MDCT. Patient underwent surgery and the LAD was ligated and grafting was used to re-establish the perfusion between LAD and RCA. Patient was kept in Intensive care unit for 3 weeks and later discharged.

CONCLUSION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). It is a rare congenital heart lesion most seen in first year of life, and rarely seen in adults. MDCT and electrocardiography are important, non invasive procedures for diagnosing ALCAPA. MDCT has advantages such as less procedure time, no hospital stay and good spatial resolution.

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