Late presentation of ALCAPA syndrome in an elderly Asian lady

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SUMMARY

Coronary artery anomalies are often discovered incidentally during cardiac catheterization or computed tomography coronary angiography and may involve the affected coronary artery origin and its course. Coronary artery anomalies are associated with congenital heart disease. The affected coronary arteries may have an unusual high take off origin, origin from contralateral or non-coronary sinus, origin from the pulmonary artery, single coronary system or coronary artery fistula.

KEY WORDS:

ALCAPA syndrome; Bland-White-Garland syndrome; late presentation; elderly; oldest surviving Asian lady; untreated

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is an extremely rare congenital heart disease, under diagnosed and usually manifests as heart failure in the paediatric age group. The morbidity and mortality rates are very high unless it is diagnosed early and expeditiously treated with surgery. It is even rarer to document a surviving elderly ALCAPA patient.

CASE REPORT

A 71-year-old lady presented with a short history of cough, shortness of breath and fever. She is an ex-smoker and was previously well. She had a low grade fever, mildly raised jugular venous pressure, rhonchi, displaced apex beat at the 6th left intercostal space and a soft systolic murmur over the precordium. Chest X-ray showed features of heart failure. Echocardiography (ECHO) showed global hypokinesia and moderately reduced left ventricular ejection fraction (LVEF) of 35-45%. There was trace tricuspid regurgitation, trace mitral regurgitation and the right ventricular systolic pressure was 30mmHg. She was initially treated as acute lower respiratory tract infection (LRTI) with congestive heart failure. She responded well to antibiotics, nebuliser and heart failure treatment. Coronary angiography showed a large right coronary artery (RCA) with collateral connection to the left coronary system and the left main artery (LM) appeared to drain freely into the presumed cavity of the pulmonary artery (Figure 1). The coronaries were smooth with no evidence of atherosclerotic disease. Coronary computed tomography angiography (CCTA) confirmed the abnormal connection of the LM to the pulmonary trunk. The case was discussed at our Heart Team meeting and we suggested surgical reimplantation of the LM to the aorta. She was not keen for surgery and she opted for medical therapy. She remained well for the past 6 years with good metabolic equivalent of functional capacity, New York Heart Association Class I and LVEF of about 45%.

DISCUSSION

The LM arises from the aortic root and branch into the left anterior descending artery (LAD) and left circumflex artery. When the entire left coronary artery system arises from the pulmonary artery, it is an extremely rare entity called anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) or Bland-White-Garland (BWG) syndrome. It is a congenital anomaly of embryonic division of the truncus arteriosus, resulting in coronary artery abnormality with an incidence of about 1 in 300,000 births.[1] This entity is under diagnosed and is not well understood.

The affected foetus is relatively asymptomatic in utero due to the equivalent blood pressure and oxygen concentration in both left and right systems. The situation changes when the baby is born. The pulmonary pressure and level of blood oxygen immediately decrease. Thus, the LAD starts to receive lower pressure gradient blood flow and oxygen to perfuse the left ventricle. The ischemia stimulates the formation of collateral supply between the RCA and LAD. The RCA supplies collateral blood flow to the left system which subsequently drains into the low pressure pulmonary trunk leading to a coronary steal phenomenon. This so-called single coronary system supplies the whole myocardium. The imbalance between supply and demand places the left ventricle at higher risk of flow ischemia with ventricular systolic dysfunction and possibly ischemic mitral regurgitation causing congestive heart failure usually within the first year of life.

The affected infants have non-specific colicky like crying bouts, failure to thrive, feeding difficulties and heart failure symptoms. They may survive into adulthood with smaller stature, angina like symptoms, reduced effort tolerance, syncope and sudden cardiac death (SCD). Some adult asymptomatic ALCAPA patients may present with just SCD and the diagnosis can only be discovered from post mortem investigation. ALCAPA patients have high mortality rates as more than 90% of them die by one year of life without corrective surgery.[2,3] In other words, less than 10% of

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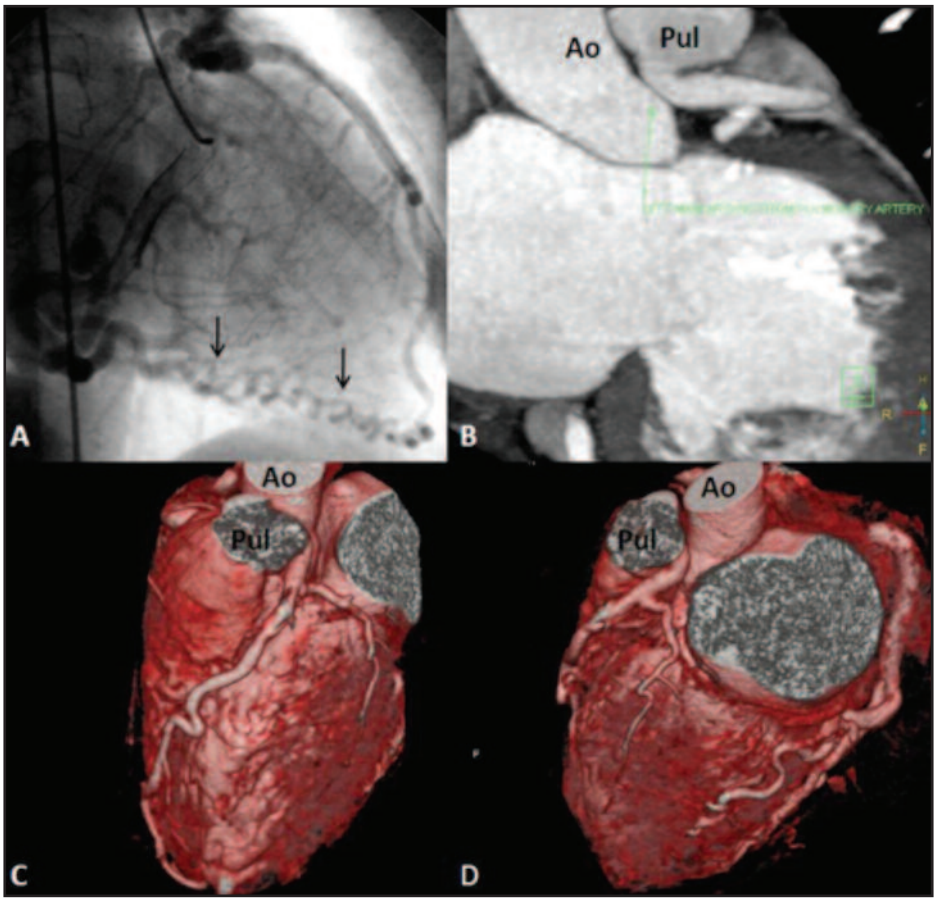

Fig. 1: (A) Coronary angiography showing the collaterals (black arrows) connecting the right coronary artery and the left anterior descending artery.
(B) Maximum intensity projection view of the cardiac computed tomography angiography (CCTA) showing the connection of the left main artery (LM) to the pulmonary artery (Pul) and not to the aorta (Ao).
(C,D) Volume rendering technique view of the CCTA confirming the origin of the LM arising from the pulmonary trunk (Pul) and not from the aorta (Ao).

childhood ALCAPA sufferers survive into adulthood and it is even rarer for this subset of patents to reach an advanced age with no symptoms, minimal symptoms or late onset of symptoms. Hence, it was indeed rare for our patient to have experienced an unremarkable childhood, uneventful spontaneous vaginal delivery episodes, and good functional daily activity.

A retrospective study found an association between significant atherosclerotic coronary artery disease and anomalous coronaries.[5] Her coronaries were almost pristine without any hint of atherosclerotic disease. It was the chronic strain of increased preload on the left ventricular pump with significant left to right shunting of blood from the systemic to the pulmonary circulation which caused the gradual deterioration of the ventricular pump. The reduction in left ventricular systolic function was most probable slow and gradual as she was hitherto asymptomatic. The LRTI coincidently unmasked the dysfunction, precipitating her first episode of heart failure.

Early and appropriate detection of this anomaly requires a high index of suspicion. When assessing an infant with heart failure or a suspected adult, ECHO is an invaluable screening tool. ECHO may reveal increased colour flow doppler signal of the septum and dilatation of the RCA. Invasive cardiac catheterisation and CCTA are the next imaging modalities to be considered if there is ECHO suspicion of a structurally abnormal heart or an unclear cause of heart failure.

Coronary angiography was performed in this patient to look for evidence of atherosclerotic coronary artery disease and an ischemic aetiology for her cardiomyopathy. We were surprised by the absence of the left coronary artery from the aorta, the presence of long collaterals from the RCA which continued and ended at the pulmonary trunk. CCTA is useful to confirm the anomalous course of the affected coronary artery, its drainage and other great vessels. Magnetic resonance angiography is another imaging modality to be considered for the investigation of anomalous coronaries but the current technology limited its usefulness to visualisation of the proximal coronary segments only. Stress perfusion

imaging is useful to demonstrate the areas of ischemia but it is academic and would not change the management in this patient as she had opted for conservative therapy.

The definitive treatment for ALCAPA syndrome is surgery. There are several surgical techniques described such as LM reimplantation, Takeuchi procedure, left coronary artery ligation and left coronary artery ligation with concomitant coronary artery bypass grafting. We believe that surgery is beneficial for paediatric ALCAPA patients, mainly because it would improve the survival rate. When surgery is contemplated for an elderly patient like this lady, one has to consider subjecting her to unnecessary surgical risk without any probable net tangible improvement in the clinical outcome. Contemporary local statistics revealed an average life expectancy of 77 years for females. Perhaps it is prudent to treat her cardiomyopathy medically and conservatively, and she may even survive to become an octogenarian! In conclusion, this is the oldest surviving Asian lady with ALCAPA syndrome without surgical intervention.

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