

A Rare Case of ALCAPA and Rheumatic Mitral Valve Regurgitation in an Adult Patient

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ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary artery defect leading to sudden cardiac death. Diagnosis is made after the onset of symptoms, mainly in the pediatric population. We describe an uncommon presentation of ALCAPA and rheumatic mitral valve regurgitation, diagnosed by a coronary 64-CT scan performed before a planned mitral valve repair operation.

INTRODUCTION

ALCAPA was first described in 1933 as Bland-White-Garland Syndrome. It represents between 0.24% and 0.46% of all congenital cardiac anomalies and affects 1 of every 300,000 live births [Kim 2006]. If not corrected, up to 90% of patients die in the first year of life. When the right to left collateral blood flow is sufficient, symptoms can be absent or relatively fleeting, allowing patients to grow into adulthood. In these cases, ALCAPA syndrome may cause myocardial infarction, left ventricular dysfunction, mitral regurgitation, silent myocardial infarction, and sudden cardiac death.

CASE REPORT

A 30-year-old woman was referred for rheumatic mitral valve disease with severe regurgitation and normal ejection fraction. In her past history, she reported rheumatic disease at the age of 10, and caesarean childbirth at the age of 28. She was symptomatic only for dyspnea after moderate exercises.

Electrocardiogram and biochemical laboratory investigations were normal. Dilatation of left atrium and ventricle were present at echocardiographic examination, together

with severe pulmonary artery hypertension and mild tricuspid regurgitation. Moreover, an anomalous blood flow in the pulmonary artery was noticed, raising the suspicion of ALCAPA. She underwent CT coronary angiography, which confirmed the diagnosis; moreover a very tortuous right coronary artery arising from the aorta was shown, as well as dilated right and left coronary arteries (Figure 1). Coronary angiography confirmed the presence of ALCAPA, and early signs of anterior medio-ventricular wall hypokinesia were recorded by magnetic resonance examination.

Cardiopulmonary bypass was instituted through aorta and selected cava veins cannulation. The left main (LM) coronary artery was long, arising posteriorly from the pulmonary artery (Figure 2). A dilated left anterior descending artery (LAD) originated from the LM coronary artery.

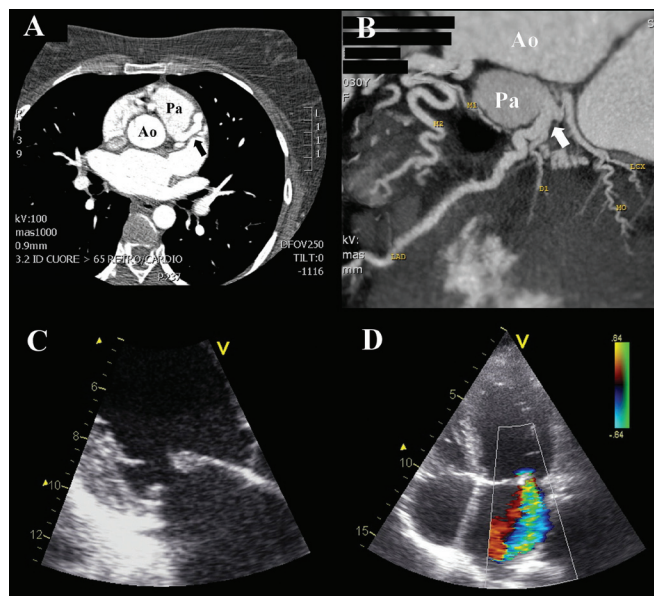


Figure 1. A, CT scan revealed the presence of ALCAPA. The left main coronary artery (arrow) originated from the pulmonary artery (axial view). B, Bidimensional curved multiplanar reconstructions showed the anatomical anomalies of the coronary arteries. C, D, Bidimensional transthoracic echocardiographic images revealed a severe regurgitation in the rheumatic mitral valve; thickening of the leaflets (mainly the anterior leaflet) and retraction of the posterior leaflet can be observed.

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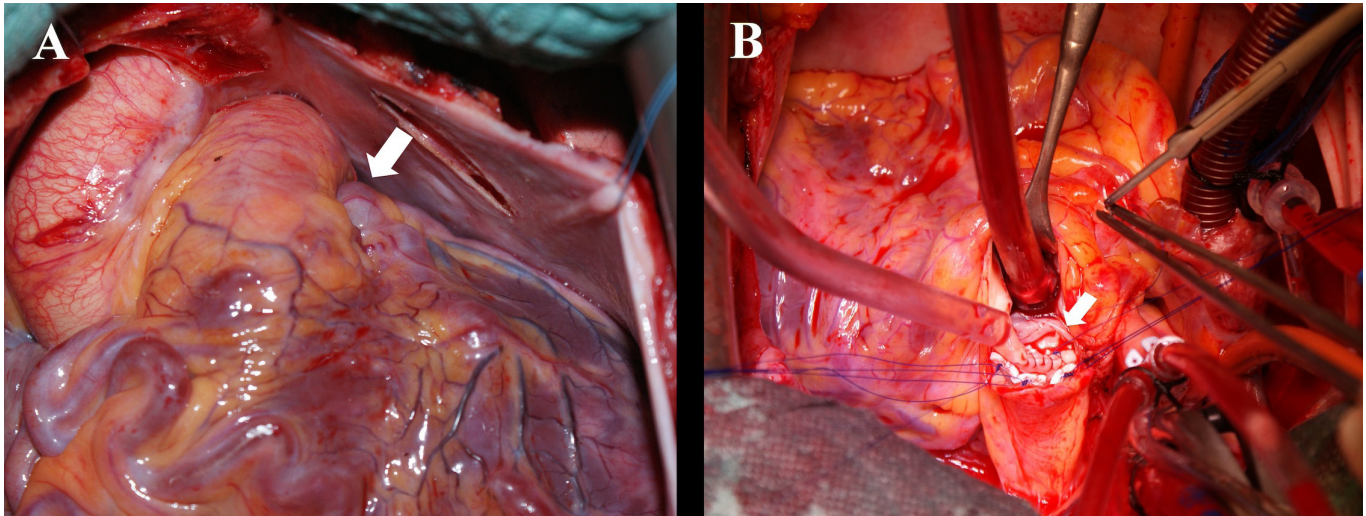


Figure 2. A, At the pericardial opening, the right coronary appeared dilated, while the left main coronary artery (arrow) was visible next to the pulmonary artery. B, After longitudinal incision of the pulmonary artery, the left main ostium was sutured.

Direct examination of the mitral valve from left atriotomy confirmed a posterior leaflet retraction with small and stiff middle (P2) and posteromedial (P3) scallops. Both mitral leaflets were thickened, and thickening was prevalent along the free margin of the anterior leaflet as observed in trans-thoracic echocardiogram (Fig 1, C). Both commissures were free from fusion or calcification and no chordal fusion or shortening was observed. The anatomical abnormalities of the mitral valve were clearly caused by rheumatic disease and the valve was repaired by a posterior leaflet elongation with an autologous pericardial patch plus a 30-mm mitral ring (Carpentier-Edwards Physio II, Edwards Lifesciences Corp, Irvine, CA). The pulmonary artery main trunk was opened longitudinally. The LM ostium was located in the posterior wall of the pulmonary artery trunk and directly closed in 4 layers with 4-0 Prolene with pledgets. The LIMA was grafted with the LAD artery close to the bifurcation with the circumflex artery, using 8-0 Prolene. The left atrium and pulmonary artery were sutured and the patient was weaned off bypass. The patient was extubated after 6 hours. The first postoperative echocardiography exam showed decreased pulmonary artery pressure, absent mitral regurgitation, and a normal ejection fraction. She had an uneventful recovery and was discharged on the postoperative day 5.

At 18-months follow-up, the patient was asymptomatic and no regurgitation of the mitral valve was present.

DISCUSSION

ALCAPA is a rare congenital heart disease. After the second month of life, the vascular resistance of the pulmonary bed starts to decrease and the pressure in the pulmonary artery system drops, resulting in a decrease of left coronary

blood flow in ALCAPA patients. If blood flow from collaterals is sufficient, symptoms can be absent or relatively minor, allowing growth into adulthood [Cowles 2007]. The few subjects who remain asymptomatic until adulthood often show subclinical myocardial ischemia [Moodie 1983]. Without surgical intervention, they are at increased risk of malignant ventricular arrhythmias and sudden cardiac death. Global cardiomyopathy is very common among those patients, reaching an estimated incidence of 80% to 90% at mean age of 35 years [Alexi-Meskishvili 1995].

In the present case, we report the diagnosis of ALCAPA at adult age associated with a non-related, non-congenital concomitant cardiac pathology such as mitral regurgitation. Usually, mitral regurgitation is present in ALCAPA patients because of myocardial ischemia [Moodie 1983; Alexi-Meskishvili 1995]. The only case describing the association between ALCAPA and non-ischemic mitral regurgitation was reported in a 5-month-old female patient with concomitant mitral valve congenital disease (anomalous mitral arcade) and ALCAPA [Su 2011].

Our patient remained undiagnosed until the age of 28, when the diagnoses of mitral regurgitation and increased pulmonary pressure were made during her first pregnancy. However, the first echocardiographic examination did not reveal the presence of ALCAPA, possibly because ALCAPA's peculiar echocardiographic abnormalities were not investigated. She underwent caesarean-section delivery because of the valve disease; the procedure was uneventful despite the severe risk for myocardial infarction related to ALCAPA.

When she was referred to our unit for mitral valve surgery, her symptoms were compatible with the mitral defect, and no clinical sign was suggestive of ALCAPA. In the literature, bidimensional echocardiography is the

first diagnostic test to evaluate patients with suspected ALCAPA. When clinical signs are not present, echocardiographic findings can be effectively confirmed with a non-invasive imaging technique such as CT scan, before proceeding to coronary angiography.

Surgical options for the treatment of adult ALCAPA patients are mainly represented by 1) the transposition of the LM ostium from the pulmonary artery to the aorta; 2) the coronary artery bypass grafting with direct closure of the LM; 3) the Takeuchi technique. We chose the second option due to the posterior origin of the LM ostium, which represented a relative contraindication for its transposition to the anterior wall of the ascending aorta.

In non-ischemic adult patients, the diagnosis of ALCAPA is challenging. In this case, a concomitant cardiac disease such as mitral rheumatic regurgitation led to the execution of further diagnostic tests, which revealed the presence of ALCAPA. Mitral valve disease led the obstetrician to choose a cesarean partum rather than natural childbirth, minimizing the risk. In this case, the presence of a rheumatic mitral

insufficiency may have reduced the risks of a natural childbirth due to undiagnosed ALCAPA.

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