Asymptomatic Adult Type ALCAPA Syndrome Coexisting with Bicuspid Aortic Valve- A Case Report

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), also known as Bland-White-Garland syndrome (BWGS) is a rare variant of the congenital anomalies of the coronary arteries. Two forms, the infantile (most common) and the adult types (rare) of these syndromic manifestations have been reported in the literature. Affected infants present with myocardial infarction and congestive heart failure. Few of them survive the first year of life, without appropriate surgical intervention. Adult type ALCAPA syndrome is one of the important causes of malignant cardiac arrhythmias, acute coronary syndromes, valvular pathologies and sudden cardiac death in young adults and adults. Asymptomatic adult type ALCAPA is uncommon, much more so its coexistence with bicuspid aortic valve (BAV). We present a 65-year-old patient with ALCAPA and BAV, who was largely asymptomatic. She had ligation of the ALCAPA, and aortic valve replacement, with a very good outcome.

Keywords: Asymptomatic adult type ALCAPA syndrome; Bicuspid aortic valve; Coexistence.

Introduction

The presence of an anomaly of the coronary arteries is nearly 1% in the general population [1]. These anomalies may be related to the number, origin, route, termination, structure or the size of the coronary arteries. While some of the coronary artery anomalies are associated with clinical manifestations such as sudden cardiac death, persistent arrhythmias, myocardial ischemia (persistence or paroxysmal), congestive heart failure, others are discovered incidentally [2]. Classic description of the anomalous origin of the left coronary artery from the pulmonary...
artery was first described in 1882 by Brooks [3] and it took nearly half a century before Bland, White and Garland reported clinical symptoms associated with ALCAPA [4], now known as Bland-White-Garland syndrome.

Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital anomaly occurring in 1 of 300,000 live births and only about 15% of these subjects survive through to adult life [5]. In such cases, the single right coronary artery, through a good collateral circulation is able to perfuse both ventricles, in addition to blood lost through retrograde flow from the left coronary artery into the pulmonary artery, called the coronary steal phenomenon.

About 90% of the long-term survivors of ALCAPA die at an average age of 35 years [6], as a consequence of imbalance between myocardial oxygen demand and supply. This may explain why for more than a century the long-term survivors over 50 years is about 50 cases in the literature (only 26 cases as at 1995) [7].

Approximately 1% of the general population is born with bicuspid aortic valve (BAV), which becomes stenotic in the 4th through 6th decade of life with hemodynamic abnormalities.

The coexistence of ALCAPA and BAV is extremely rare. We present the case of ALCAPA with BAV in a 65-year-old patient. She was managed by aortic valve replacement and ligation of the ALCAPA with a good outcome. The clinical significance of this case is firstly the uncommon nature of adult type ALCAPA, secondly its coexistence with the BAV, and thirdly the largely asymptomatic nature in this patient until the age of 65 years.

Case Report

This 65-year-old female patient complained of mild palpitations and exertional dyspnea (NYHA I). She had no past history of tobacco use, and no known family history of coronary artery disease. She had two uneventful term deliveries. Being an uncommon condition, diagnosing this patient was difficult. We had to reschedule our procedure on two occasions as there was initially no consensus between the transthoracic echo and the CT scan, in detecting the ostium of the LCA by echocardiography. But the contrast-enhanced computed tomography with 3D reconstruction was very specific (Figures 1 and 2).

Coronary angiographic imaging studies was also helpful (Figure 3). She was then prepared for surgery. Following median sternotomy, to ascertain the hemodynamic state and adequate perfusion with a single right coronary artery, we occluded the root of the anomalous LCA and monitored with transesophageal echocardiography and the arterial line BP half an hour before the ligation and resection of the ALCAPA.

Cardiopulmonary bypass was instituted and the aortic valve was replaced with a mechanical valve. The pulmonary artery was patched with a Dacron graft. She made an uneventful recovery and was discharged 7 days after the surgery.
Figure 1: Computed tomography (CT) showing the Aorta (Ao), calcification of the aortic valve, and (ALCAPA) left coronary artery (LCA) taking off from the pulmonary artery (PA). Right coronary artery not in view in this section.

Figure 2: Posterio-anterior 3D reconstructed contrast CT showing, dilated and tortuous right coronary artery (RCA) with extensive collaterals feeding the left coronary artery (LCA) a typical “one for all configuration” and the LCA sprouting from the right side of the pulmonary artery (PA).
Discussion

Bicuspid aortic valve is the most common congenital cardiac malformation, with an incidence of 1-2% in the general population [8]. A significant number of individuals with BAV develop aortic stenosis and complications requiring treatment. The literature suggests a possible relationship between BAV and coronary artery anomalies as evidenced in this patient (Figure 4).

This relationship may be genetic and is best explained by the hypothesis that the cause is abnormal septation of the conotruncus into the aorta and pulmonary artery, or the persistence of the pulmonary buds together with involution of the aortic buds that

Figure 3: Reconstructive image of the intra-aortic navigation view of the aortic valve showing bicuspid aortic valve. AoW denotes aortic wall, AV is the aortic valves showing two cusps with calcification.

Figure 4: Coronary angiogram of anterior posterior view, the aortic root (red labeled AoR) is seen with single right coronary artery (RCA labeled in red) which large and massively tortous.
eventually form the coronary arteries. The coronary anomalies are classified in terms of origin, number, course, termination and the structure, in patients with bicuspid aortic valves undergoing cardiac catheterization [9]. The importance of this is planning the surgical treatment and also avoiding injury to the anomalous vessels (Figure 5) at surgery.

**Figure 5**: The aorta (Ao) – cannulated, with the large and tortuous right coronary artery (RCA) and the lying freely in the pericardium. LCA is the left coronary artery shooting off from the pulmonary artery.

This 65-year-old patient presenting with ALCAPA is uncommon, much more its coexistence with the BAV stenosis, two uneventful term deliveries, no typical clinical presentation such as reported among long term survivors of the ALCAPA syndrome. Patients older than 50 years are but a handful, and the presentation may be in the form of a new-onset exertional angina, dyspnea, or syncope [10,11]. She was largely asymptomatic. Surviving with a functionally single right coronary artery requires an elaborate collateral circulation between the right and the left coronary arteries to prevent myocardial ischemia owing to blood lost through retrograde flow from the left coronary artery into the pulmonary artery [12]. Some reports mentioned cases where individuals above the age of 50 with adult type ALCAPA did well without surgery [11]. These may be cases without comorbid cardiac conditions requiring surgery. However, this is not the recommended modality of management. The three commonly used surgical approaches for treating ALCAPA are reimplantation, simple ligation, or ligation with saphenous vein bypass grafting (SVG), of which none is of significant surgical advantage [13].

The anomalous LCA was ligated and resected. Simple ligation is an adequate treatment. An important determinant of myocardial ischemia is the presence of coronary steal with left to right shunting from the LCA to the pulmonary artery. Patients have immediate improvement after the surgery [14,15], and still doing well, the patient was last seen at outpatient surgical clinic before COVID-19 pandemic.
Conclusion
Asymptomatic adult type ALCAPA is uncommon, much more so its coexistence with BAV. We have presented a 65-year-old patient with ALCAPA and BAV, who was largely asymptomatic. She had ligation of the ALCAPA, and aortic valve replacement, with a very good 6 years outcome.

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