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Bland-White-Garland syndrome in an asymptomatic adult individual: the role of imaging diagnostics in establishing a definitive diagnosis and preventing severe cardiac complications

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ABSTRACT

Bland-White-Garland syndrome otherwise known as anomalous left coronary artery from the pulmonary artery is classified as a rare congenital heart defect which can be fatal, if unrecognized and untreated. In most cases, the disease occurs in the neonatal age, while a small percentage of patients remain asymptomatic or sparsely symptomatic for many years, making early diagnosis and treatment difficult and delayed. In the following paper, we present a case of a 39-year-old Caucasian woman with no past medical history or systematic cardiac treatment who was admitted to the cardiology clinic with escalating symptoms of heart failure. Laboratory tests revealed an increase in NT-proBNP without elevation of the myocardial necrosis markers. Echocardiography showed generalized wall hypokinesis, left ventricular enlargement with features of hypertrophy, and additional flow in the pulmonary trunk. The patient was referred for further imaging examination to make a definite diagnosis, with a suspicion of patent ductus arteriosus.

Keywords: ALCAPA; Bland-White-Garland syndrome; coronary computed tomography angiography; sudden cardiac arrest

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Introduction

Bland-White-Garland syndrome (BWG), also known as anomalous left coronary artery from the pulmonary artery (ALCAPA) is a coronary vascular anomaly in which the left coronary artery (LCA) branch out from the pulmonary artery (PA) [1]. It is a rare condition that occurs in 1:300,000 newborns or 0.26–0.5% of infants born with congenital heart disease [1, 2]. The average age of diagnosis is 10.2 months, and the fatality — without corrective surgery — is 90% in the first year of life [3, 4]. In most cases,

it is an isolated defect, although it can coexist with other anomalies such as persistent ductus arteriosus (PDA), ventricular septal defect, tetralogy of Fallot or coarctation of the aorta [1, 5]. Embryologically — BWG, is a consequence of abnormal septation of the common heart trunk, conotruncal malformations and vascular trunk into the aorta and pulmonary arteries or absence of atrophy of the pulmonary collaterals and involution of the aortic collaterals [4]. This syndrome has been divided into 2 types — the adult form and the infantile form [6]. The differences between the two types are summarized in Table 1.

Table 1. Differences between infantile and adult types of ALCAPA

Type Differences	Infantile	Adult
Onset of symptoms	Up to 3 years old	From 20 to 70 years old
Symptoms	Myocardial infarction, heart failure	Chronic myocardial ischemia, arrhythmias, sudden cardiac death (SCD)
ECHO phenomena	Dilated left ventricle, abnormal kinetics of the lateral and anterior wall	Abnormalities in myocardial wall kinetics, mitral regurgitation, RCA enlargement
Presence of collateral circulation from the right coronary artery (RCA) to the left coronary artery (LCA)	None or insignificant	Well-developed collateral circulation with left ventricular reverse perfusion through the RCA

Left-right leakage has a significant role in the pathophysiology of ALCAPA, which disrupts cardiac hemodynamics. Shortly after delivery, with the decrease in vascular resistance in the pulmonary vessels, a left-right leak is formed with reverse flow from the RCA through the LCA to the PA. This leakage leads to the development of coronary steal syndrome and progressive dilatation of the RCA. These changes are directly correlated with clinical symptoms, but also have a preventive effect. Ischemia of the anterior wall of the myocardium is caused by the coronary steal phenomenon, while the development of the collateral circulation and its continuous dilatation, along with the enlargement of the RCA, has a preventive effect on the consequences of ischemia [1, 2]. As a result, patients without surgical correction of this defect can survive up to 70 years of age [1], although 80–90% of patients dies around the age of 35 [5].

Adult patients may be asymptomatic, but over time begin to present symptoms such as chest pain, dyspnea, left ventricular dysfunction, arrhythmias and sudden cardiac death due to left ventricular ischemia [3, 7, 8]. On physical examination, 70% patients present a systolic murmur, which changes its character to continuous as the flow through the peripheral circulation increases [1].

The primary diagnostic test suggesting coronary anomalies is transthoracic Doppler echocardiography, which allows monitoring of permanent left-right leakage and retrograde flow into the pulmonary artery. Additionally, left ventricular dilatation and dysfunction, collateral circulation with dilated RCA, papillary muscle fibrosis and mitral valve prolapse with mitral regurgitation might be detected [5, 9]. Another essential imaging examination that will identify the anomaly definitively and allow a certain diagnosis is computed tomography (CT). It allows accurate localization of the left coronary artery origin, the dilatation of the RCA and the severity of the collateral circulation, which can be considered

as a prognostic factor. Although angiography of the coronary arteries is still the gold standard for diagnosis, non-invasive tests like CT, magnetic resonance (MR) and echocardiography (ECHO) are currently the most commonly used [1].

Case report

A 39-year-old female visited the cardiology outpatient clinic due to decreased exercise tolerance and an intermittent sensation of heart palpitations. The patient denied stenocardial pain and resting dyspnea. Subsequently, she was referred to the cardiology clinic for cardiovascular failure graded NYHA II. Since the age of 9 months, the patient had been under cardiological observations due to dilated cardiomyopathy. However, there is no medical documentation available from that period. The last cardiological follow-up took place in 2013. At that time, a transthoracic echocardiogram (TTE) was performed and a PDA was suspected. The patient has not been under the control of any specialist clinic for the past eight years and is not taking medication on a regular basis. In addition, there is a history of nicotineism.

On admission, an electrocardiogram (ECG) and laboratory tests, including NT-proBNP and myocardial necrosis markers, were performed. A chest X-ray, Holter ECG monitoring and repeat TTE were also ordered.

The initial ECG showed pathological left axis deviation, sinus rhythm at a rate of 78/min, and a QRS duration of 110 ms. Additionally, signs of left ventricular strain and hypertrophy. Myocardial necrosis markers did not indicate an elevation, but the NT-proBNP level was elevated to 672.7 pg/ml. The chest X-ray revealed no lung abnormalities, an enlarged heart, and clear costophrenic angles. The Holter monitoring showed sinus rhythm with sporadic premature atrial and ventricular contractions, with a heart rate ranging from 60/min to 87/min.

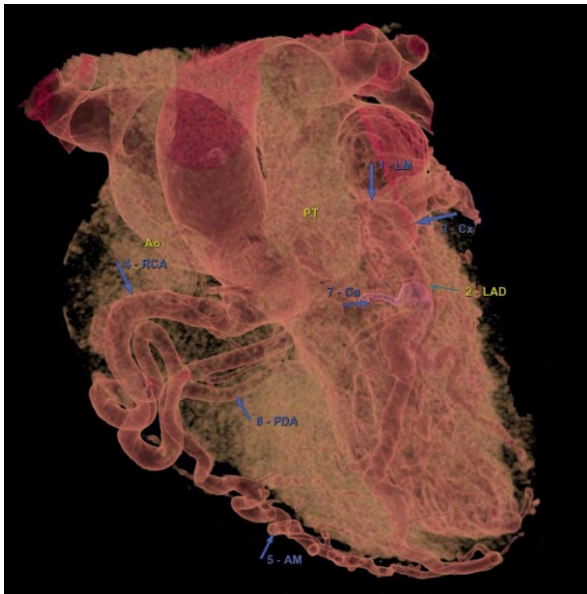


Figure 1. Coronary computed tomography angiography — 3D reconstruction of coronary arteries. Ao — aorta, PT — pulmonary trunk, LM — left main coronary artery, LAD — left anterior descending artery, Cx — circumflex artery, RCA — right coronary artery, AM — marginal artery, PDA — posterior descending artery, Ca — calcifications

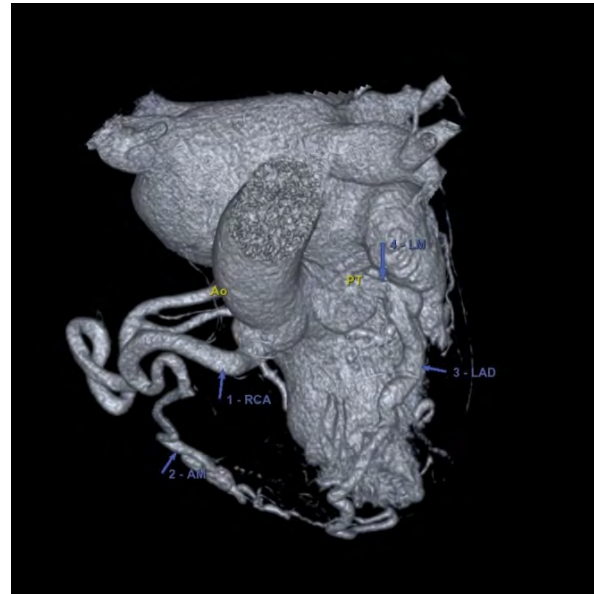


Figure 2. Coronary computed tomography angiography — 3D reconstruction of coronary arteries. Ao — aorta, PT — pulmonary trunk, RCA — right coronary artery, AM — marginal artery, LAD — left anterior descending artery, LM — left main coronary artery

The echocardiogram examination revealed an enlarged left ventricle and left atrium, with features of hypertrophy in the left ventricle. The left ventricular systolic function showed mild global dysfunction and spherical remodeling. Moderate mitral regurgitation was observed. The pulmonary trunk and pulmonary arteries appeared dilated, and an additional shunt in the pulmonary artery raised suspicion of PDA.

To identify vascular anomalies and establish a definitive diagnosis, the patient was referred for coronary computed tomography angiography (CCTA). The CCTA findings are presented in Figure 1, Figure 2, and Figure 3.

The CT scan allowed a certain identification of ALCAPA. LCA originated from the pulmonary trunk. The coronary arteries were dilated and had a tortuous course. RCA measured 8 mm in width (normal: 2–4 mm). Numerous collateral connections between the branches of the right and left coronary arteries were observed. Significant hemodynamically important left-to-right shunting was observed, from the aorta through the RCA and then the LCA to the pulmonary trunk. Following the diagnostic evaluation, establishment of the diagnosis, and pharmacotherapy, the patient was discharged home in a stable condition with recommendations for systematic medication intake and further treatment in a cardiology outpatient clinic. Additionally, the patient was referred to a specialized adult congenital heart disease center.

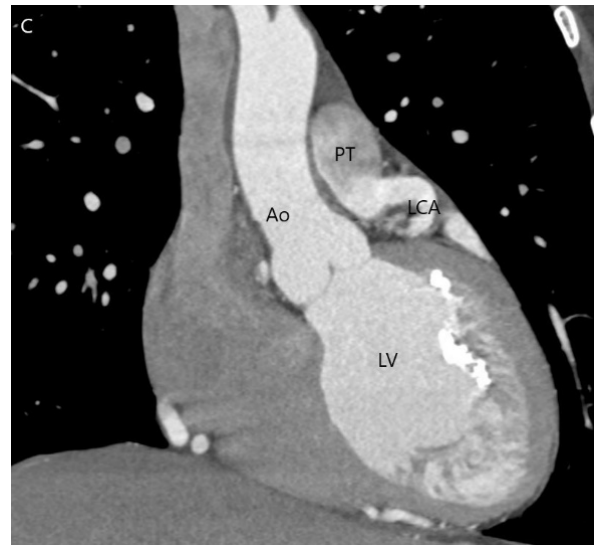


Figure 3. Cardiac angio CT image. Ao — aorta, PT — pulmonary trunk, LV — left ventricle, LCA — left coronary artery

Discussion

An unclear clinical features, enlarged RCA, retrograde flow in the LCA, mitral regurgitation on echocardiography, and mitral valve defect should raise suspicion of ALCAPA [2, 7]. Despite the mild and non-life-threatening course of the disease, this case is significant due

to the correlation between the patient's age, moderate symptoms of heart failure, and surpassing the average survival age of 35 years for affected individuals. During follow-up, the patient reports intermittent fatigue, exertional dyspnea, and episodes of palpitations during physical activity. This case highlights the importance of computed tomography in detecting and diagnosing ALCAPA in patients with mild clinical symptoms and inconclusive results from other diagnostic tests, such as echocardiography. Earlier recognition of ALCAPA could have facilitated an earlier referral to a specialized center and potentially limited the progression of heart failure. It should be noted that currently surgical repair performed in selected centers is the only treatment option. There are two methods of surgical intervention: the rarer one involves creating a tunnel from the aorta to the abnormal left coronary artery and then closing the connection between the left coronary artery and the pulmonary artery (known as Takeuchi procedure), while the most common approach is to transplant the left coronary artery in its normal position on the aorta.

Conclusions

The occurrence of BWG in adulthood is extremely rare, while it can entail very dangerous consequences, which are sudden cardiac death (SCD) and life-threatening arrhythmias. The widespread use of non-invasive tests, such as ECHO and CT scans, makes it possible to diagnose ALCAPA earlier and to care for these patients in order to prevent SCD and to refer patients for corrective surgery, which is the only certain treatment for this disease [2, 8]. The described case confirms that early diagnosis is even more important because, despite the severity of the anomaly, some patients remain sparsely symptomatic and common tests, such as ECHO, are ineffective for BWG and may be insufficient.

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