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Late presentation of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA): How can clinicians diagnose?

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SUMMARY

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare disease characterized by myocardial ischemia which becomes symptomatic shortly after birth leading to left heart failure and death. Survival to adulthood have been reported in some cases. Recognition and diagnosis of ALCAPA syndrome is important due to its potentially life-threatening complications. This case report emphasizes the importance of diagnosing late presentation of the ALCAPA syndrome.

Key words: *Coronary artery anomaly, late presentation, pulmonary artery*

ÖZET

Pulmoner arterden köken alan sol koroner arter anomalisinin (ALCAPA) geç ortaya çıkışı: klinisyenler nasıl tanı koyabilir?

Pulmoner arterden köken alan sol koroner arter anomalisi (ALCAPA), doğumdan kısa süre sonra semptomatik olan, sol kalp yetmezliği ve ölüme yol açan miyokardiyal iskemi ile karakterize nadir görülen bir hastalıktır. Erişkinliğe kadar sağkalım birkaç vakada bildirilmiştir. ALCAPA sendromunun fark edilmesi ve tanı konulması, olası hayatı tehdit eden komplikasyonları nedeniyle önemlidir. Bu olgu sunumu, geç ortaya çıkan ALCAPA sendromunun tanısının önemini vurgulamaktadır.

Anahtar kelimeler: *Koroner arter anomalisi, geç ortaya çıkış, pulmoner arter*

Introduction

Congenital coronary anomalies are seen in less than 1-2% of infants and approximately 1% of adults undergoing coronary angiography. Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) is an extremely rare anomaly and survival to adulthood is exceptional (1-3). We herein report a case of a young adult with ALCAPA syndrome.

Case Report

A 21-year-old male was admitted with the complaints of fatigue and exertional dyspnea to our hospital. His previous cardiac history was unrevealing. Physical examination revealed a grade 2/6 apical systolic murmur. ECG depicted nonspecific T wave changes in anterior derivations. Cardiothoracic index was found to be slightly increased in chest X-ray. Transthoracic echocardiography (TTE) revealed mild mitral regurgitation with an ejection fraction of 60%. Doppler investigation showed turbulent diastolic flow within the interventricular septum and inferior wall (Figure 1). In addition, 2D and Doppler investigations demonstrated that the left coronary artery (LCA) was originating from the main pulmonary artery (Figure 2).

Cardiac catheterization was performed. The LCA was not visualized at the left sinus valsalva. The right coronary artery (RCA) was dilated and tortuous. LCA, which was filling retrogradely via collateral vessels from the RCA appeared during right coronary injection. It was originating from the main pulmonary artery, which also visualized during the RCA angiography (Figure 3). Surgical correction of the coronary anatomy was planned.

Discussion

Anomalous origin of the LCA from the pulmonary artery is a rare anomaly, accounting for 0.24% of all congenital heart anomalies. This anomaly, which is also called “Bland-White-Garland Syndrome” is seen

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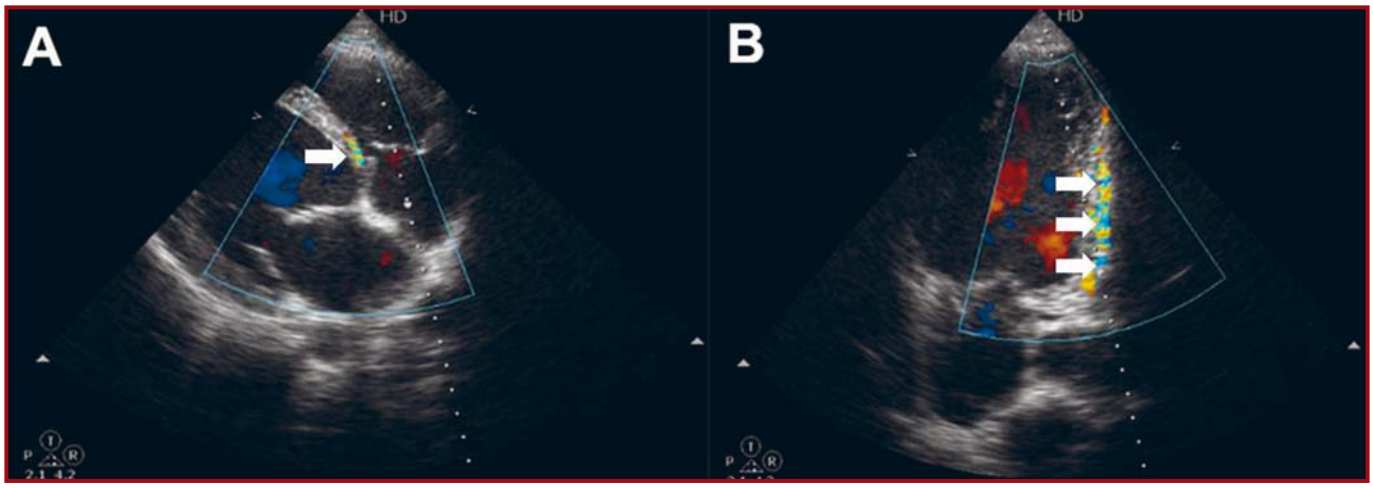

Figure 1. Doppler imaging showing turbulent diastolic flow within the interventricular septum (A) and inferior (B) wall

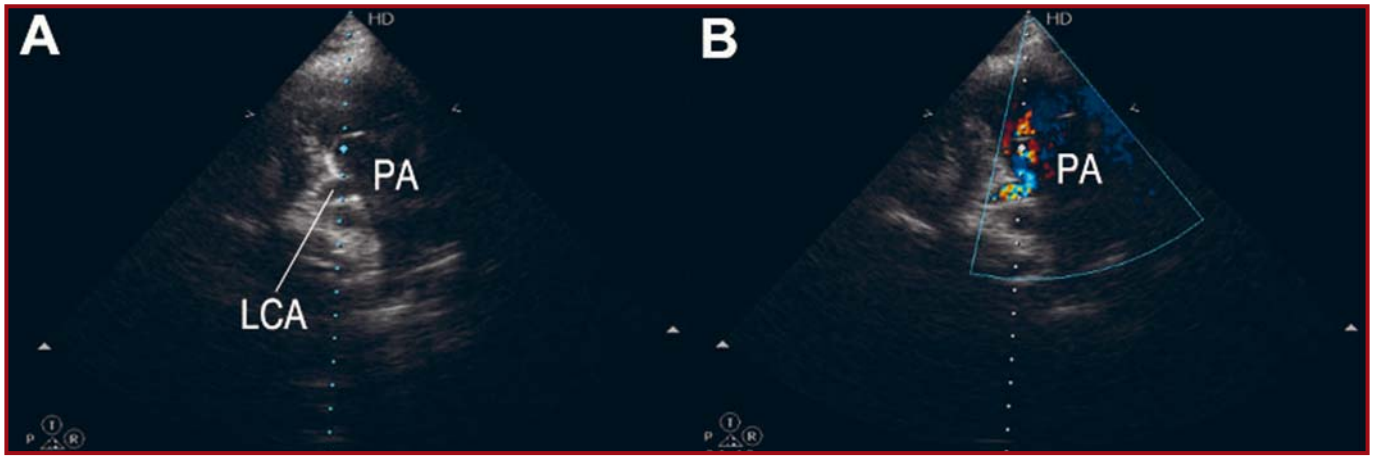

Figure 2. Two-dimensional (A) and Doppler (B) investigation showing that the left coronary artery (LCA) was originating from the main pulmonary artery (PA)

in approximately 18% of the children with congestive heart failure under 2 years old (2,4). The left anterior descending artery, left circumflex coronary artery and RCA may have individual origins from the pulmonary artery, with similar pathophysiologic and clinical picture. Individual origins of the coronary arteries have been thought to be benign (2). On the other hand, when the LCA arises from the pulmonary artery, this anomaly can cause myocardial ischemia that may be progressive and lead to left ventricular dysfunction, mitral insufficiency, congestive heart failure and sudden cardiac death. Asymptomatic presentation after infancy and survival into adulthood is extremely rare (3,4). The clinical presentation is related with the coronary artery involved and its myocardial distribution, the pulmonary vascular resistance, and the number and size of collateral vessels. Collateral circulation between RCA and LCA is the major determiner for the extent of the myocardial ischemia. When the collateral circulation is sufficient, symptoms may be absent or unremarkable and these patients are classified as adult type. Without significant collateral circulation, severe myocardial ischemia deteriorates myocardial function with the associated signs and symptoms of heart failure. These patients are classified as the infantile type (2-5).

In adults with ALCAPA RCA is dilated and tortuous because of the collateral vessels that this vessel gives to LCA, as in our case (Figure 1).

Myocardium can tolerate low oxygen concentration as in cyanotic heart disease but it is vulnerable in low perfusion pressure. After the birth, as the pulmonary artery pressure falls, left ventricular function begins to disturb because of both low perfusion and low oxygen pressure in patients with ALCAPA. Fortunately, this fall in pulmonary artery pressure is usually gradual as left ventricular ischemia and dysfunction tend to increase the left ventricular end-diastolic and left atrial pressure which in turn retard the fall in pulmonary artery pressure. This process may slow the deterioration of the left ventricular function. But, pulmonary artery pressure and left

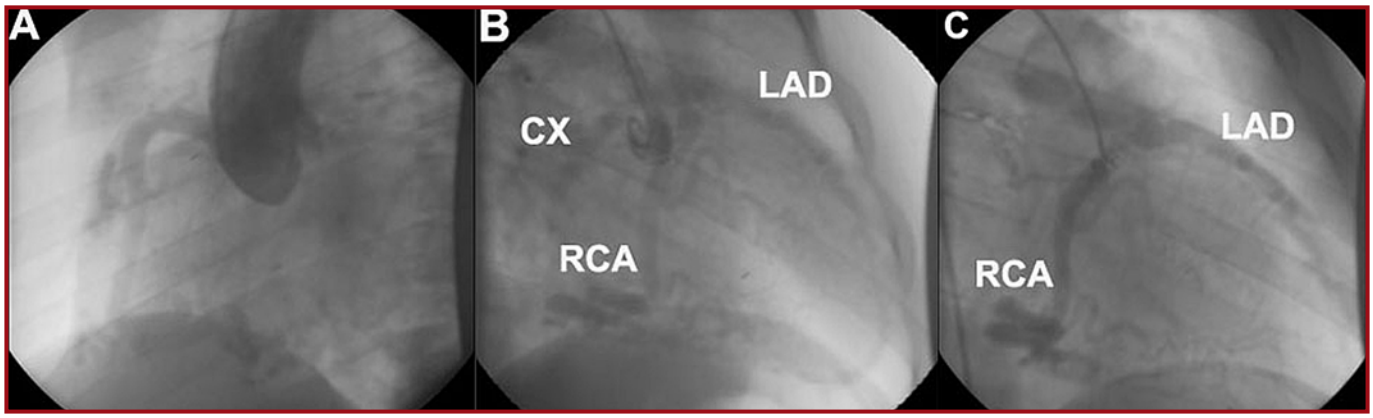

Figure 3. Aortagraphy showing that no artery was originating from the left sinus valsalva (A). Coronary angiography showing ectatic and tortuos right coronary artery (RCA) filling left coronary artery (LCA) via collateral vessels and draining into the main pulmonary artery (B, C). LAD: left anterior descending artery, CFX: left circumflex coronary artery

ventricular perfusion pressure decrease to critical levels between the 6th and 8th weeks of life and patients may be symptomatic in this period. On the other hand, in some patients with ALCAPA, a rich network of collateral vessels from the RCA may improve and increase the perfusion and O_2 pressure in the LCA. In this group, LCA drains into relatively low-pressured pulmonary artery. Although this may cause a "pulmonary steal phenomenon", these patients survive to adulthood and constitute the adult type of ALCAPA syndrome.

In patients with ALCAPA, ischemic injury usually causes left ventricular dilatation and mitral regurgitation due to papillary muscle dysfunction. In the present case, left ventricular function was in normal limits with only a mild dilatation, and only a mild mitral regurgitation was observed. The main echocardiographic finding related to ALCAPA was diastolic turbulent flow in the left ventricle due to collateral flow from the giant RCA.

The prognosis in patients with adult type ALCAPA is poor. Sudden cardiac death is common and mortality until the age of 35 may be as high as 90% (2). Therefore surgical correction of the anomaly is recommended in all patients as we planned in the present case.

In conclusion, although extremely rare, late presentation of the ALCAPA syndrome is possible and may be suspected from the echocardiographic findings. Regardless of the presence of the symptoms, surgery is recommended as soon as possible due to the risk of sudden cardiac death.

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