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A rare reason of dilated cardiomyopathy in the newborn period: ALCAPA syndrome

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Summary

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart defect. ALCAPA is asymptomatic in many cases in the newborn period. It is mostly diagnosed in the first few months. In this case report, we present a newborn with ALCAPA who admitted to our clinic with heart failure which occurred at an earlier age than expected. The electrocardiography showed deep wide Q waves in D1 and aVL and ST elevation in leads V1-V6. Echocardiography revealed a dilated cardiomyopathy and a left main coronary artery originating from the pulmonary artery. Diagnosis was confirmed by coronary angiography. In this report, we emphasized that ALCAPA may cause dilated cardiomyopathy also in the newborn period and we aimed enhanced awareness of this disease. *(Turk Arch Ped 2011; 46: 248-50)*

Key words: Anomalous origin of coronary artery, dilated cardiomyopathy, newborn

Introduction

ALCAPA syndrome which is defined as anomalous origin of the left coronary artery from the pulmonary artery constitutes 0.5% of all congenital cardiac anomalies. The incidence in live newborns is 1/300000 (1). 87% of the children born with this anomaly are symptomatic during the infancy and 65-85% are lost with cardiac failure before the age of one and especially around 2-3 months of age (2).

A diagnosis of ALCAPA syndrome was made as a result of diagnostic angiography and with clinical electrocardiographical and echocardiographical findings in a 28 days old patient with dilated cardiomyopahty (DCM) and congestive cardiac failure. By presenting this case it was aimed to emphasize that ALCAPA syndrome can lead to DCM even during the newborn period and to increase the awarenes of this disease.

Case report

A 28 days old girl was presented to our outpatient clinic with rapid breathing and cyanosis while feeding. It was informed that she had no complaints until before one week ago and her complaints including rapid breathing and cyanosis during feeding increased in the last day. Physical examination revealed the following findings: weight: 3750 g (25-50p), height: 55 cm (50p), head circumference 37 cm (50-75p), apical heart beat: 158/min, blood pressure 67/48 mmHg, respiratory rate: 80/min. The liver was 4 cm palpable and 2/6 systolic murmur was heard in the mesocardium. Telecardiogaphy revealed a cardiothoracic index of 62%. On ECG, deep Q waves were observed in DI and a VL and ST elevation was observed in the other derivations (Figure 1). Complete blood count revealed the following: Hb: 12.4 g/dl, Htc 37%, white cells: 3840/mm^3, platalets: 471000/mm^3. BUN: 5.5 mg/dl, creatinine: 0.3 mg/dl, SGOT: 15 U/L, SGPT: 53 U/L, Na: 138 mEq/L, K+: 4 mEq/L, Ca++: 10 mg/dl. The level of brain natriuretic pepdid (BNP) which was investigated because of cardiac failure was found to be 2422 pg/ml (N≤100). Troponin-I which was investigated because of ischemic findings on ECG was found to be 2.38 ng/ml (N≤0.03). Echocardiography revealed that left cardiac chambers were larger than normal and the left ventricle was spherical. Cardiac contractions were decreased (Shortening

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fraction : 16%). A marked mitral failure and moderate tricuspid failure were found. Right ventricular pressure was found to be 60 mmHg. The right coronary artery was observed to originate from its normal location and not to be enlarged. Since the left coronary artery appeared to originate from the pulmonary artery (figure 2), ALCAPA syndrome was considered in the patient and cardiac catheterization and angiography were decided to be performed. The diagnosis was confirmed, when the left coronary artery was observed to originate from the pulmonary artery during angiography (Figure 3). The patient was referred for operation. Connection of the abnormal coronary artery to the aorta was provided by an intrapulmonary tunnel. However, the patient was lost in the early postoperative period.

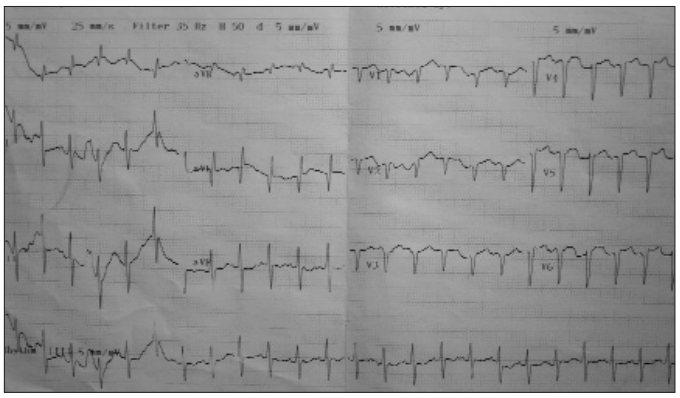

Figure 1: Deep Q waves in DI and aVL and ST elevation in precordial derivations on electrocardiogram are observed.

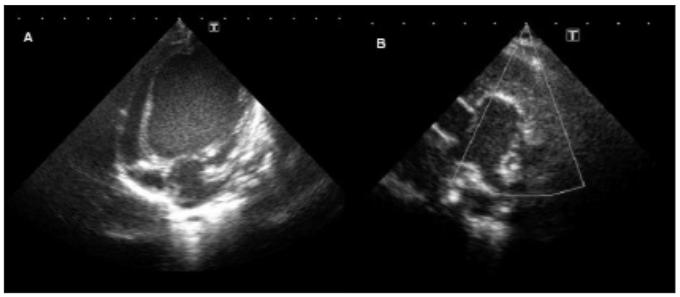

Figure 2: A. Four chambers are seen on echocardiography and the left ventricle is observed to be markedly enlarged. B. On parasternal short axis view on echocardiography, it is observed that the left coronary artery is originating from the pulmonary artery.

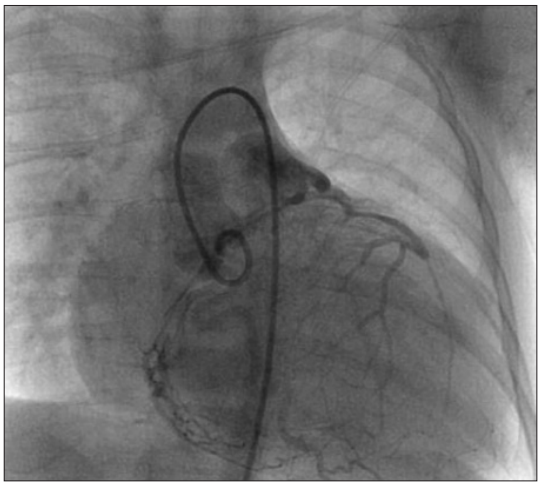

Figure 3: Angiography reveals that following aortic root injection firstly the right coronary artery is filled with contrast material, then the left coronary artery is filled via the collateral vessels and later the pulmonary artery is filled.

Discussion

Although ALCAPA syndrome is observed rarely, if it is not treated, is has a high mortality rate during the first 1-2 years of life. The pathophysiologic and clinical properties of this disease are directly related to the perfusion of the left myocardium. This anomaly may have a good prognosis during fetal period, since both saturations and pressures of the aorta and pulmonary artery are similar. After birth vascular resistance of the pulmonary artery starts to decrease and blood flow from the pulmonary artery into the left coronary artery progressively decreases. The perfusion of the left ventricle depends on the collaterals developed between the right coronary artery and left coronary artery. The number of collaterals or escape syndrome from the left coronary artery to the pulmonary artery with low pressure determine the adequacy of perfusion of the left ventricular myocardium or the degree of ischemia (3,4). Because of this pathophysiology clinical signs occur 2-3 months after birth in 85% of the patients (5). Babies presented to the clinic with a picture of cardiac failure during the first year of life constitute a large portion of the cases. In the remaining 15%, the occurrence of cardiac problems may delay until adult ages (5,6). It is seen more rarely in the newborn period.

In the study performed by Walsh et al.(7), the mean age at diagnosis was found to be 4,5 months in 11 subjects diagnosed during a 5-year period and the age at diagnosis in one subject was found to be 2 weeks. In the study performed by Floren et al (8), 27 subjects were evaluated and the age at diagnosis was found to be between 3 months and 54 months. In this study, no subject diagnosed during the newborn period was reported. In another study, only one subject was diagnosed during the newborn period among a total of 7 subjects diagnosed in a period of 13 years (9).

Diagnosis of ALCAPA which is a treatable condition in children with dilated cardiomyopathy is very important. The subjects frequently have a diagnosis of DCM and are investigated for a long time in terms of metabolic diseases. They are frequently considered to have idiopathic DCM and are lost, since no treatment is performed for the actual cause. In subjects who present with dilated cardiomyopathy, some ECG and ECHO clues have been defined not to miss the diagnosis of ALCAPA. Echocardiographic findings are directly related to the degree of perfusion of the left ventricular myocardium. Main findings on ECG include deep and narrow Q waves in DI, aL and V4-V6, left ventricular hypertrophy and left axis deviation (5). However, these findings are not specific and can be observed in other cardiomyopahties. Main findings on ECHO include enlarged right coronary artery, absence of visualization

of origin of the left coronary artery from the aorta, increase in the echogenicity of papillary muscles and observation of origin of the left coronary artery from the pulmonary artery. False negative results for distinctive findings on echocardiography have been reported in studies performed. In a study, it was reported that the left coronary artery was false negatively observed to be originating from the aorta on the first ECHO examination in 50% of the patients who were diagnosed as ALCAPA (10). In the study performed by Chang and Alada (11), this ratio was found to be 71%. In this study, a measurement system was developed, since negative results can be obtained at least in more than 50% of the patients and this disease can be especially confused with idiopathic DCM. When the findings including QT pattern, a Q wave deeper than 3 mm and negative T wave in aVL on ECG, increase in the ratio of the left coronary artery diameter to the aortic annulus on ECHO (≥0.14), increase in the echogenicity of the papillary muscles and increased flow towards the pulmonary artery are evaluated together, it has been reported that a sensitivity of 100% and a specificty of 91% can be reached for the diagnosis of ALCAPA.

As the level of knowledge and experience increased as a result of the studies performed, the incidence of the disease showed variance. Previously the incidence was reported to be 1/300000. Brotherton et al. (12) reported this figure as 1/4243 in their study they performed in live newborns.

Definite treatment of ALCAPA syndrome is surgery. Medical treatment is mostly performed to increase myocardial contractions. Early diagnosis and treatment provide a more rapid myocardial improvement. While Michielon et al.(13) emphasized young infants had a better potential of improvement for left ventricular functions, Ando et al (14) reported that ventricular function improved in adults after surgical treatment at a degree observed as in children. Although our patient was operated early (late newborn period), left ventricular functions did not improve because of myocardial ischemia and the patient detoriorated in the early postoperative period and was lost.

Consequently, it must be known that surgical treatment is the definite solution for this disease which has a fatal prognosis when untreated. Although the first signs generally occur after the first 2-3 months of life, clinical, telecardiographical and ECG findings should be examined, clues on ECHO related to ALCAPA should be investigated carefully and early treatment should be provided in newborn patients presenting with DCM.

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