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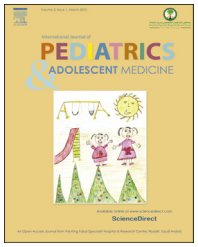

INSTRUCTIVE CASE

Near missed reversible cardiomyopathy: The value of the electrocardiogram

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Abstract Dilated cardiomyopathy is a devastating disease affecting the myocardium that is characterized by cardiac chamber dilatation with contractile function impairment in the absence of structural heart disease. The majority of cases are idiopathic; these patients have a poor outcome. However, identifying a reversible etiology and instituting appropriate intervention could reverse the disease process and result in complete recovery. An electrocardiogram is a simple, non-invasive and quick test that should be performed in each patient presenting with dilated cardiomyopathy. Failure to perform this test or misinterpreting its result could result in a tragic misdiagnosis of idiopathic cardiomyopathy, depriving the patient of a potentially curative intervention. Here, we report two cases of dilated cardiomyopathy caused by clinical conditions with recognizable ECGs. In both cases, the diagnosis was missed initially, delaying corrective interventions. These cases draw attention to the importance of performing and correctly interpreting ECGs in patients with dilated cardiomyopathy.
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1. Introduction

Dilated cardiomyopathy (DCM) refers to a large group of heterogeneous myocardial disorders that are characterized by ventricular dilation and depressed myocardial contractility in the absence of abnormal loading conditions, such as hypertension or valvular disease [1]. The Pediatric Cardiomyopathy Registry in North America reported the annual incidence of DCM to be 0.57 per 100,000 children per year [2]. Almost half of these patients die or undergo a heart transplant within 2 years of diagnosis [2,3]. In addition,

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two-thirds of patients with DCM are thought to have DCM that is idiopathic in origin. Such a high frequency of idiopathic DCM may lead the practitioner to pay less attention during the search for an etiology of a newly diagnosed case, thereby committing the patient to a poor prognosis with high mortality.

However, finding and appropriately treating the primary cause of DCM may result in the total restoration of cardiac function and reversal of its sequela.

One of the routine and important tests in investigating DCM is an electrocardiogram (ECG). Unlike blood tests that produce quantitative results, the ECG result is based on physician interpretation that requires prior knowledge and experience. Failure to recognize the cardiac etiology of DCMs that manifest on an ECG can result in a delayed or missed diagnosis with potentially morbid or even fatal outcomes.

We report two patients who, on initial assessment, were misdiagnosed as idiopathic DCM or DCM following presumed myocarditis. Twelve months later, after careful interpretation of their ECGs, these patients were appropriately diagnosed and treated. Both underwent corrective intervention. One had complete recovery of cardiac function with reverse remodeling; the other patient is still undergoing follow-up treatment and is expected to improve.

2. Case 1

The first patient is a 2-year-old boy born at 36 weeks of gestation with a birth weight of 2.6 kg. As a one year old, he was admitted for two weeks with a history of tachypnea and poor weight gain. His weight at the time was 5.7 kg, and he had a height of 62 cm and a head circumference of 44 cm (all below the 5th percentile).

As part of the initial work-up, a chest X-ray and ECG were performed. The chest X-ray showed cardiomegaly, while the ECG showed sinus tachycardia, with a deep Q wave in leads I and aVL and left ventricular hypertrophy with a strain pattern in leads V5 and V6 (Fig. 1). These findings were not reported at the time.

The patient was referred for cardiac evaluation, and the diagnosis of idiopathic DCM was made after the echocardiogram showed a dilated left ventricle with an ejection fraction (EF) of 35% and mild mitral regurgitation. No comment was made about the origin of the left coronary artery.

The patient was started on anti-failure medication (furosemide, captopril and digoxin) and was discharged home. One year later, the patient was seen by a different cardiologist who spotted the ECG abnormalities and referred the patient to our center with a suspected anomalous left coronary artery from the pulmonary artery (ALCAPA). The clinical evaluation at our center was not different from the referring site; however, echocardiography showed the right coronary artery (RCA) to be dilated with multiple collaterals, while the left coronary artery (LCA) origin was not clearly observed. With color Doppler, the flow in the LCA was retrograde, which is highly suspicious for ALCAPA. To confirm the diagnosis, cardiac catheterization was performed. A selective angiogram showed a dilated RCA with multiple collaterals filling the LCA that originated from the pulmonary artery instead of the aorta (Fig. 2). The patient was taken for surgery, and the LCA was found to originate high on the MPA at the takeoff of the right pulmonary artery (Fig. 3). Such a high LCA takeoff is unusual for cases of ALCAPA, which explains the difficulty in visualizing its origin on echocardiography. The LCA was implanted to the aorta, and after a smooth post-operative course, the patient was discharged home on anti-failure medications four days after surgery. One month after surgery, he continues to be on medical therapy for his heart failure with the hope that cardiac function will improve with time.

3. Case 2

A 3-year-old boy presented for the first time to a local hospital with a one month history of shortness of breath, lethargy, and weight loss. These symptoms were preceded

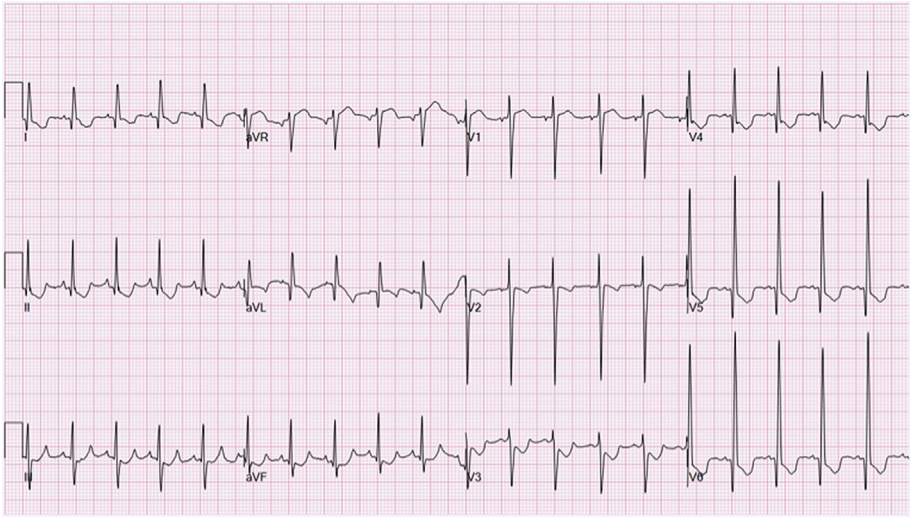

Figure 1 ECG of case 1. There is sinus tachycardia with deep Q wave in leads I and aVL strongly suggesting ALCAPA. There is also LVH with ST segment depression and T wave inversion most evident in V5 and V6.

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