Isolated Left Ventricular Apical Hypoplasia: Reporting a Case With Mild Manifestations and Different Echocardiography Features

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Abstract

Introduction: Isolated left ventricular apical hypoplasia is an unusual type of cardiomyopathy that presents with different clinical manifestations according to the age of the disease, ranging from no symptoms in children to congestive heart failure, pulmonary edema, or even malignant tachycardia in adults. To our knowledge, only a few cases of isolated left ventricular apical hypoplasia have been reported in Asian adults.

Case Presentation: Herein, we described an adult case of isolated left ventricular apical hypoplasia referred to our heart center in Isfahan, Iran in 2015 with a complaint of mild dyspnea with the absence of obvious fatty tissue in the heart’s apex and an absence of any shunt, which are common findings in patients with this phenomenon.

Conclusions: Patients faced with isolated left ventricular apical hypoplasia should be monitored by echocardiography because of this disease’s possible progressive trend to life-threatening consequences.

Keywords: Electrocardiography, Echocardiography, Ventricle Hypoplasia

1. Introduction

Isolated left ventricular (LV) apical hypoplasia is described as congenital heart disease with an unusual type of cardiomyopathy that was first described in 2004 (1). The typical imaging features of this cardiomyopathy include a spherical, truncated LV with some degree of systolic dysfunction and an elongated, normally functioning right ventricle (RV) that wraps around the distal LV (2). Most reported cases of isolated LV apical hypoplasia have not been accompanied by other abnormalities (3-5). This rare phenomenon frequently presents with different clinical manifestations according to the age of the disease, from no symptoms in children to congestive heart failure, pulmonary edema, or even malignant tachycardia in adults (6).

A 2D echocardiogram and cardiac MRI can successfully indicate different morphological features. The early diagnosis of LV abnormalities, such as LV apical hypoplasia, is particularly important due to the difference in the prognosis and management of LV apical hypoplasia from those of other diseases with symptoms similar to LV apical hypoplasia (7). Various modalities have been introduced to diagnose LV apical hypoplasia with varying diagnostic accuracy. However, echocardiography seems to be the most effective tool for diagnosing LV apical hypoplasia (8). Until now, few cases of isolated LV apical hypoplasia have been reported in Asian adults (9). This report describes a case of isolated LV apical hypoplasia in an adult suffering from mild dyspnea.

2. Case Presentation

A 19-year-old teenager was referred to our Heart Center in 2015 for further meticulous echocardiography after several diagnostic echocardiograms were conducted for his mild dyspnea. No evidence of palpitation, syncope, arrhythmia, or chest pain was described by the patient on physical examination, and there was no family history of premature coronary artery disease, cardiomyopathy, or sudden cardiac death. The patient’s birth and maternal history was unremarkable; the patient was born full term via vaginal delivery.

A twelve-lead electrocardiogram revealed a normal feature without any rhythm abnormalities or ST segment changes. A review of the patient’s previous 2D echocardiograms from three to four years prior revealed a long-lasting spherical LV with no significant deterioration in LV function. Also, evidence of obvious fatty tissue in the apex of the heart or a shunt were not observed (Figure 1). Our patient was managed as a case of mild LV dysfunction with a conservative approach.
3. Discussion

The patient in our case presented with mild dyspnea but without significant deterioration of LV function or the manifestation of heart failure. In the electrocardiography assessment, no arrhythmias were found and the patient was given a favorable outcome. In some affected cases, LV apical hypoplasia has a malignant nature with hemodynamics similar to restrictive cardiomyopathy that may progress to pulmonary hypertension and atrial or ventricular tachyarrhythmias (10). Thus, although our case was able to be controlled with a conservative approach, many affected patients may need invasive procedures and even cardiac transplantation, particularly before the development of irreversible pulmonary hypertension (1).

A review of the literature published within the past ten years showed that most cases of LV apical hypoplasia cases have reported among the European and Caucasian populations and more rarely in Spanish, Mexican, and Middle Eastern nations (5, 10-12). The clinical manifestations of the disease ranged from no symptoms, especially among younger patients; clinical symptoms that included fatigue, shortness of breath, acute chest pain, and palpitations; and, in some cases with more progressive manifestations, abnormal electrocardiography, progressive dyspnea, pulmonary edema, and even sudden death.

Some hypotheses concerning the pathophysiology of this phenomenon have been proposed. One of the proposed mechanisms of LV apical hypoplasia is inadequate LV dilatation during partitioning that results in a spherical LV (1) (Figures 2 and 3).

Patients faced with isolated LV apical hypoplasia should be monitored because of this disease’s possible progressive trend to life-threatening consequences, such as heart failure, pulmonary hypertension, and arrhythmias. However, according to our reported case, isolated LV apical hypoplasia can manifest with mild symptoms without electrocardiography abnormalities or improper outcomes.

Cases of isolated LV apical hypoplasia have only rarely been reported with mild symptoms without serious changes in imaging patterns or poor outcomes. Thus, this case can be practical and educational. However, our data on other modalities for the diagnosis of isolated LV apical hypoplasia, such as MRI, were lacking.

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