Correspondence

Left-dominant Arrhythmogenic Cardiomyopathy: A Case Misdiagnosed as Pseudoaneurysm

Chao Yu¹, Tian-Gang Zhu¹, Wen-Ling Liu¹, Wen-Feng Huang²

¹Department of Cardiology, Peking University People's Hospital, Beijing 100044, China
²Department of Emergency Medicine, Peking University People's Hospital, Beijing 100044, China

To the Editor: A 60-year-old man was admitted to our emergency department with a new-onset sudden palpitation and precordial discomfort that occurred several times a day. Sustained monomorphic ventricular tachycardia (VT) of a right bundle-branch block (RBBB) pattern (245 beats/min) was documented, which was reversed into a sinus rhythm after synchronized cardioversion. Subsequent 12-lead electrocardiogram revealed a normal sinus rhythm with T-wave inversion in leads V4 to V6 and ST segment elevation in leads V1 to V3 (78 beats/min). He had been diagnosed with diabetes about 10 years ago which was well treated. There was no family history of arrhythmogenic cardiomyopathy or other signs or symptoms of heart disease. Myocardial enzyme levels, including troponin I, creatine kinase isoenzyme MB, and myoglobin, were within normal level. The level of serum C-reactive protein and D-dimer was normal. Transthoracic two-dimensional echocardiography presented a normal left ventricular (LV) global systolic function (an end diastolic/systolic volume of 150/66 ml, an ejection fraction of 57%), an LV pseudoaneurysm (apex, 4.2 cm × 2.6 cm) [Figure 1], and normal morphological right ventricle (RV). Coronary angiography showed normal coronary arteries.

The patient received surgical aneurysm resection and LV restoration under cardiopulmonary bypass finally. Exploratory operation presented that there is an aneurysm in the LV apex, instead of pseudoaneurysm (multiple prominent trabeculations in the apex and poor acoustic window provided a misleading echo picture of the situation) and several emboli were taken out from the aneurysm. RV showed no abnormalities. Ventricular apical myocardial biopsy specimens report demonstrated classic features of left-dominant arrhythmogenic cardiomyopathy (LDAC) – a replacement of the ventricular myocardium was present with fibrous and adipose tissue. Genetic sequencing showed that no known mutation of cardiomyopathy was detected, but he had three mutant heterozygotes in TGFBR2, SMAD9, TRPM4 gene which did not find a link with this disease. Based on these findings, the clinical diagnosis of LDAC was made.

The patient was adequately informed about the risk of life-threatening ventricular arrhythmias and about the efficacy on preventing sudden death and the possible complications of both implantable cardioverter-defibrillator and radiofrequency catheter ablation.

Figure 1: Apical 4-chamber, two-dimensional echocardiography demonstrates an apical pseudoaneurysm during cardiac systole (yellow arrows). Color Doppler showed multicolored shunt flow signal through ventricular apical outlet (white arrows). LV: Left ventricle; LA: Left atrium.

but he refused any more invasive therapeutic intervention and was discharged to treatment with amiodarone.

During his 6-month follow-up, he remained asymptomatic and did not have a recurrence of the VT. Repeated echocardiography revealed normal global systolic function (an end diastolic/systolic volume of 187/94 ml and an ejection fraction of 60%).

LDAC is rare and is primarily associated with LV aneurysms that are not explained by vascular events. It usually presents from adolescence to old age, typically with palpitations and/or symptoms of impaired consciousness, with the major complication being sudden cardiac death. The clinical suspicion of LDAC is alerted in patients who present ventricular arrhythmias of RBBB and left bundle-branch block type and unexplained T-wave inversion in inferolateral leads. Regional wall motion abnormalities or aneurysm detected by echocardiography or cardiac magnetic

Received: 07-04-2016  Edited by: Yuan-Yuan Ji
How to cite this article: Yu C, Zhu TG, Liu WL, Huang WF. Left-dominant Arrhythmogenic Cardiomyopathy: A Case Misdiagnosed as Pseudoaneurysm. Chin Med J 2016;129:1763-4.

Address for correspondence: Dr. Wen-Ling Liu,
Department of Cardiology, Peking University People's Hospital,
Beijing 100044, China
E-Mail: liuwenling@medmail.com.cn

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

© 2016 Chinese Medical Journal | Produced by Wolters Kluwer - Medknow
resonance (CMR) without any pattern of coronary artery disease should raise suspicion of underlying myocardial disease due to fibrous or fibro-fatty replacement.\textsuperscript{1,\textsuperscript{2}} This case illustrated a misleading clinical feature of LDAC indicating that LV aneurysm without coronary diseases accompanied by ventricular arrhythmia might be a morbid association underlying LDAC. The limitation of this case was that the area of affected myocardium was determined by echocardiography instead of CMR, extent of aneurysm resection may not be as accurate as possible, a risk of arrhythmia was still left.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. Sen-Chowdhry S, Syrris P, Prasad SK, Hughes SE, Merrifield R, Ward D, \textit{et al.} Left-dominant arrhythmogenic cardiomyopathy: An under-recognized clinical entity. \textit{J Am Coll Cardiol} 2008;52:2175-87. doi: 10.1146/annurev.med.052208.130419.
2. Szymanski P, Klisiewicz A, Spiewak M, Szumowski L, Walczak F, Hoffman P. Left dominant arrhythmogenic cardiomyopathy – A newly defined clinical entity. \textit{Int J Cardiol} 2012;156:e60-1. doi: 10.1016/j.ijcard.2011.08.073.