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Case Report

Apical Hyperthrophic Cardiomyopathy with Ace of Spades-Form as a Rare Cause of Cardiac Arrest Secondary to Ventricular Fibrillation

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Case Report

A 45-year-old woman with multiple sclerosis was admitted to our hospital after out of hospital cardio-pulmonary resuscitation. The first ECG showed ventricular fibrillation. Following direct current defibrillation and mechanical reanimation, spontaneous circulation was restored and the ECG unremarkable without any signs of ischemia. Coronary angiography showed unobstructed coronary arteries Figure 1, (Panel A). Left ventriculography revealed apical wall obstruction, suggestive of apical aneurysm (Panel B, supplementary videos). Transthoracic echocardiography and Cardiac Magnetic Resonance Imaging (CMR) eventually lead to the diagnosis of a rare case of Apical Hypertrophic Cardiomyopathy (AHC) with ace of spades-form (Panel C,D). The patient underwent Cardioverter-Defibrillator Implantation (ICD) and amiodarone medical therapy for secondary prophylaxis. Patient's family history and screening for HCM were unsuspicious.

AHC is a rare kind of hypertrophic cardiomyopathy with predominantly left ventricular apical wall thickening, prevailing in the Asian and Japanese population [1]. Clinical manifestations include diastolic dysfunction, left atrial enlargement and tachyarrhythmia. Prognosis seems to be benign, despite patients showing typical ECG-changes that mimic acute coronary syndrome such as deep T-wave inversions or other ST-changes. Apical thrombi, ventricular aneurysms and myocardial

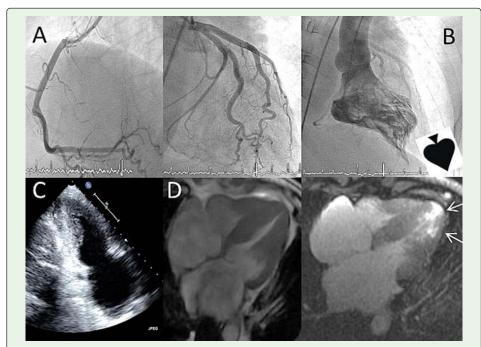


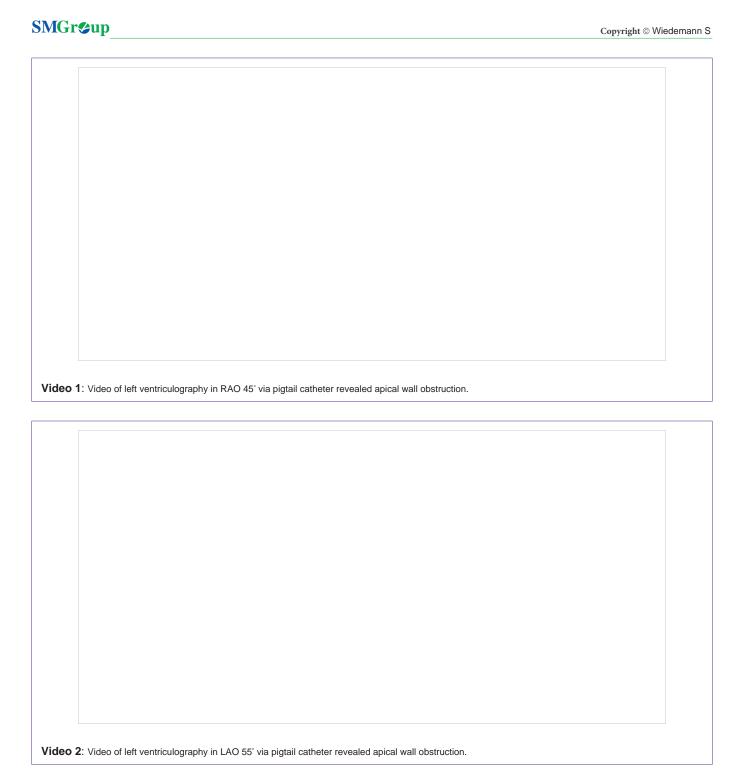
Figure 1: Panel A: Coronary angiography showed unobstructed coronary arteries without signs of atherosclerosis in RAO ap and lateral view.

Panel B: Image of left ventriculography revealed apical wall obstruction.

Panel C: Transthoracic echocardiographic findings with apical hypertrophy up to 18mm and "spade–shaped" configuration of left ventricular end diastolic cavity.

Panel D: CMR in 4-chamber-view demonstrates apical hypertrophic form of HCM in cine sequence (left) and with LGE (arrows) in apical segments (right).

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infarction are associated with poorer prognosis [2]. The characteristic finding is a "spade –shaped" configuration of left ventricular end diastolic cavity in ventriculography. Echocardiography and CMR can differentiate between mid ventricular hypertrophy with apical aneurysm and apical thickening with ace of spades morphology. Mid myocardial Late Gadolinium Enhancement (LGE) in the hypertrophic lesion seems to be a good prognostic parameter for risk stratification to guide therapeutic decision making such as ICD-implantation, medical antiarrhythmic therapies or radiofrequency ablation.

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