AN UNUSUAL CAUSE OF STROKE WITH AN ABNORMAL ECG SPOILER

Diego H. González-Bravo, MD; Jose Escabí-Mendoza, MD, FACC; Liliana Llopart, MD
Cardiovascular Division, Department of Medicine
VA Caribbean Healthcare System, San Juan, Puerto Rico.
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INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is the most common genetic heart disease characterized by a diverse clinical and phenotypic spectrum. Apical HCM (A-HCM) was initially reported in Japan, by Sakamoto in 1976 and subsequently by Yamaguchi et al in 1979. In epidemiological studies it seems to be particularly more common in East Asian populations, reporting to represent approximately 25% of Japanese patients while in more western regions as the United States only reporting 2% of HCM patients. For A-HCM the diagnosis is made by the presence of an asymmetrical left ventricular hypertrophy (LVH) predominantly in the ventricular apex, with an apical/posterior wall thickness ratio of 1.5 or more, as determined by echocardiography and/or cardiac magnetic resonance (CMR) imaging. The electrocardiogram in A-HCM typically shows repolarization changes with "giant" T-wave inversions > 1mV in the precordial leads and commonly associated to voltage criteria for LVH, that not infrequently hints the physician in suspecting this condition.

CASE PRESENTATION

76-year-old male with type-2 diabetes mellitus and hypertension who presented with ataxia, apraxia, and diplopia since 3 days prior to admission. Physical examination revealed left monocular diplopia without other focal neurological deficits. No heart murmurs or carotid bruits were detected. His electrocardiogram showed a normal sinus rhythm, LVH and marked T-wave inversions in V3-V6. Brain CT scan and a subsequent MRI confirmed an acute non-hemorrhagic right parieto-occipital cortical infarct with findings suggestive of microembolism. In our search for a thromboembolic source, a carotid Doppler revealed no hemodynamically significant disease and a transthoracic echocardiogram was performed. It showed preserved LV systolic function with mid-apical hypertrophy (2cm) and nearly complete obliteration of the apical chamber. Echocardiography with contrast disclosed distal apical dyskinesia with a localized intracavitary pressure gradient, causing the formation of a small apical aneurysm (<3cm), that was likely the source of thromboembolism despite no evident thrombi in it. These findings together with the initial electrocardiographic changes were diagnostic of A-HCM and chronic anticoagulation was recommended.

ELECTROCARDIOGRAPHY

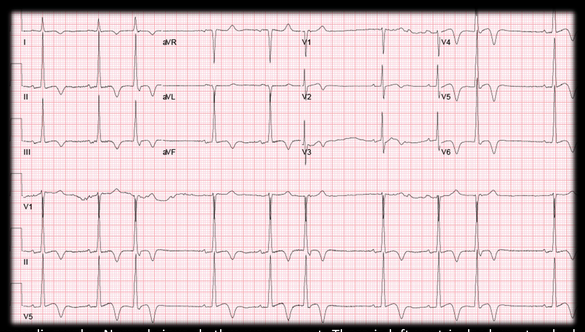

Figure 1: Electrocardiography. Normal sinus rhythm was present. There is left ventricular hypertrophy by Sokolow-Lyon criteria (R in V5 + S in V1 >35mm) with associated T wave inversions from V3-V6 suggestive of apical hypertrophic cardiomyopathy.

BRAIN MAGNETIC RESONACE IMAGING (MRI)

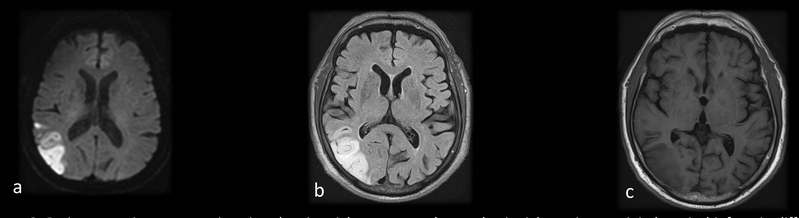

Figure 2: Brain magnetic resonance imaging showing right acute non-hemorrhagic right parieto-occipital cortical infarct in diffusion-weighted imaging (a), fluid attenuated inversion recovery (b) and T1-weighted sequences (c).

TRANSTHORACIC ECHOCARDIOGRAPHY (TTE)

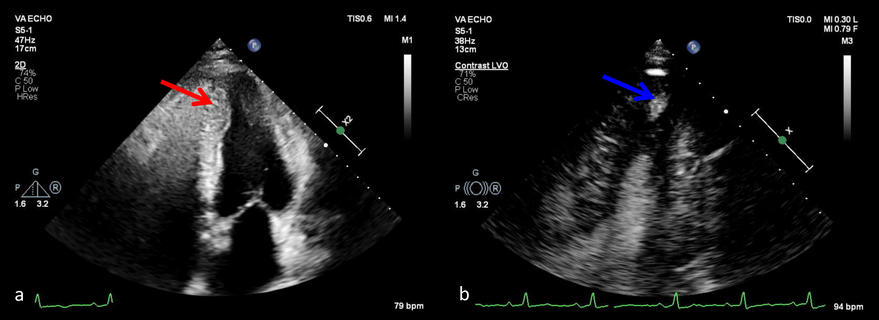

Figure 3: Transthoracic echocardiogram 4-chamber view showing marked apical left ventricular hypertrophy (red arrow) in diastole (a). During systole (b), there is apical dyskinesia with the formation of a small (<3cm) apical aneurysm (blue arrow) that can be visualized after contrast administration.

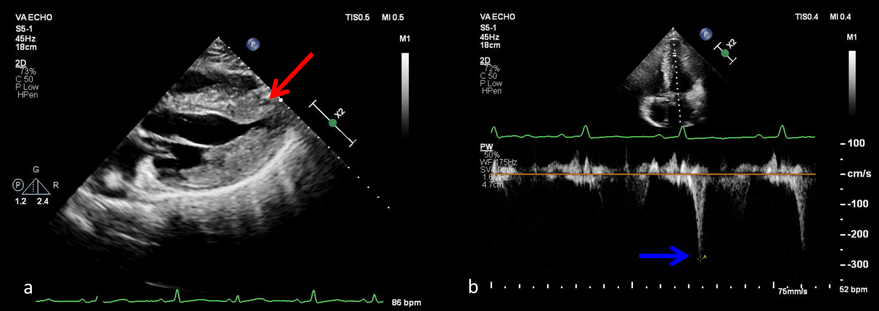

Figure 4: Transthoracic echocardiogram subcostal window (a) showing left ventricle with predominant apical hypertrophy (red arrow). Spectral pulse wave Doppler (b) showing an apical intracavitary gradient of 30mmHg (blue arrow).

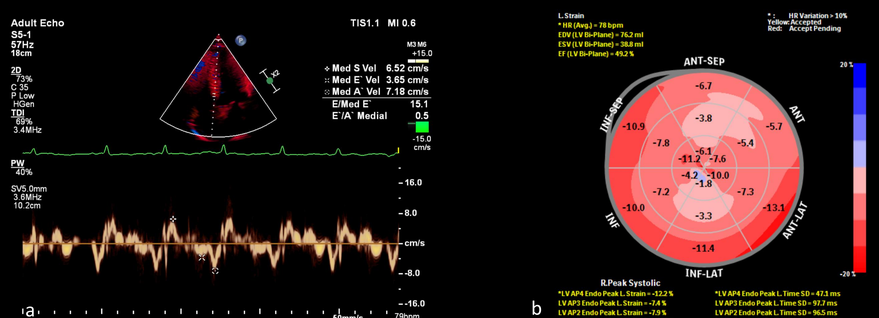

Figure 5: Spectral tissue Doppler imaging (a) showing reduced e' (3.65cm/s) as expected with myopathogenic diseases such as hypertrophic cardiomyopathy. Longitudinal strain bull's eye plot showing reduced longitudinal strain predominantly in the mid and apical areas of the left ventricle suggestive of apical hypertrophic cardiomyopathy.

DISCUSSION

Echocardiography will sometimes miss the diagnosis of apical hypertrophy if the apex is not clearly seen, which may be significantly enhanced with the use of contrast echocardiography or by the use of CMR imaging. A-HCM is generally more benign, not associated to LVOT obstructive problems and has a very low risk of SCD. However, on long term prognosis, up to 30% of individuals developed significant late cardiovascular events, including atrial fibrillation, myocardial infarction, congestive heart failure, transient ischemic attack, stroke, and ventricular arrhythmias. The progression or development of apical aneurysm in A-HCM is an uncommon feature with a reported incidence of approximately 5% during the clinical course of the disease (6). This feature is associated to a higher risk for arrhythmic sudden death or thromboembolic events, with a 3-fold increase of HCM-related deaths (or aborted events). Non-anticoagulated patients experienced non-fatal thromboembolic events (1.1%/year) compared to no embolic events in patients with chronic anticoagulation. For this reason, the presence of HCM with apical aneurysm with or without a prior thromboembolic event should be considered for chronic anticoagulation treatment.

CONCLUSION

Progression of apical hypertrophic cardiomyopathy can result in the formation of an apical aneurysm in 5% of cases and result in thromboembolic events. Such diagnosis needs to be considered in patient with an ischemic stroke in which the source of thromboembolism remains unknown after excluding more common causes such as atrial fibrillation and carotid artery disease. Echocardiography is of vital importance for diagnosis, and in case where the apex is not clearly seen, contrast echocardiography or MRI should be considered instead. Chronic anticoagulation in these patient should be highly considered, especially if there is already history of thromboembolic events.

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AUTHOR DISCLOSURES: NONE